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Noon Chai and gastric cancer

Imtiaz Wani, Fazl Q Parray, Rauf A Wani, Sameer H Naqash, Khursheed A Wani, Ajaz A Malik, Nisar A Choudri, Majeed A Wani, Nawab A Khan, Tariq A Sheikh

ABSTRACT

Gastric cancer remains a global disease worldwide. This is a partly preventable public health problem. Several factors are suspected to play a role in gastric carcinogenesis. High salt intake and gastric cancer are closely related. Dietary sodium chloride has been identified both epidemiologically and experimentally to be capable of increasing the risk of gastric cancer. Kashmir is a high prevalence zone of gastric cancer. The consumption of Noon Chai is considered one of the factors contributing to gastric cancer in Kashmir. Noon Chai is a traditional salted tea beverage made in Kashmir. It is taken in morning and afternoon, almost by everyone, every day, irrespective of gender or age. It is made from special tea leaves, milk, salt and sodium bicarbonate. Salted tea has high methylyating activity and leads to exposure to some potent nitrosamines or their precursors which are suspected carcinogens. A review on Noon Chai and its relationship with gastric cancer in Kashmir is presented.

Keywords: Noon Chai, Salted tea, Gastric cancer

INTRODUCTION

Gastric cancer continues to be a major health problem throughout the globe with approximately 989,600 new cases and 738,000 deaths per year, accounting for about 8% of new cancer cases [1, 2]. In India, gastric cancer is the fifth most common cancer among males and seventh most common cancer among females [3]. The incidence of gastric cancer in India is high in certain geographical areas (southern part and north-eastern states of the country) where the incidence is comparable to high-incidence areas of the world [4]. There has been a decline in the incidence of gastric cancer by this declining trend has not been seen in certain parts of India [5]. Kashmir is a high prevalence zone of gastric cancer [6–12]. Incidence of gastric cancer in Kashmir has been reported to exceed 40% of all cancers, and the incidence is three to six times higher than that at various metropolis cancer registries in India [6]. Cancer of the stomach is amongst the first five cancers in the Kashmir valley, with a M:F ratio of 3:17:1 [12].

Dietary factors are thought to contribute to the large international variation in gastric cancer rates [13]. There is marked variation in the incidence of gastric cancer among different ethnic groups residing in the same geographical area, pointing to host genetic factors or socio-environmental factors peculiar to a particular racial group. The association between few epidemiological factors and gastric cancer incidence may be attributed to some extent, to some unique dietary habits [14–16]. It is estimated that the global burden of gastric cancer could be reduced by up to 50%
by dietary changes [17]. Epidemiological studies conducted mainly in Asian countries indicate that high intake of salted food increases the risk of gastric cancer [18, 19]. The current literature suggests that salt preference has a marginal positive association with the risk of gastric cancer [20, 21]. The peculiar geography, genetics and some special dietary habits with a possible familial predisposition may have a bearing on the high risk of gastric cancer in the Kashmir [6, 7]. The excess intake of hot salted alkaline tea (Noon Chai), consumption of a dried leafy vegetable Brassica oleracea (Haak), pickled vegetables, dried smoked fish, dried raw food, spice cakes (Wur), use of red chillies and spices are some of the distinct dietary habits for increased risk of gastric cancer in Kashmir [7, 9].

Noon Chai: Noon Chai is a traditional pink colored salted tea beverage made in Kashmir. This beverage (salted tea) in Kashmir is referred to as Noon Chai, whereas ethnic Kashmiri population uses the Central Asian term Sheer Chai. Noon Chai is salty in taste and called Namkeen (salty) Chai or also called Pink Chai or Pink Tea. Noon Chai is a prime traditional salted cuisine in Kashmir and almost everyone takes this daily [6–12]. This is the most popular and common drink of Kashmir. The custom of drinking Noon Chai is very old and practiced from generations in almost every family of Kashmir. Often, this tea is served in a flask or large samovar (brass utensil) which keeps tea warm by burning coal inside.

This peculiar beverage has unique method of preparation which is practiced exclusively in the Kashmir valley. Green tea leaves are brewed in sodium bicarbonate until a thick red-brown color extract is obtained which is called 'tueth'. Correct measurement of baking soda is the key for getting nice pink color of tea. Excess sodium bicarbonate will turn tea dark and less soda will not turn tea pink. The formation of tueth takes about 45 minutes to 2 hours till brown color tueth is achieved. This tueth is then diluted with water and then salt and milk are added. The tea is served hot and may be garnished with cream on top depending on the personal liking. The cream is taken from the top layer that forms on boiled milk after the milk is cooled. The formed salted tea may be boiled again before drinking. Noon Chai is first beverage taken in the morning and later in the afternoon with the Naan (a type of bread, also called tchot or telvur) brought fresh from the bakers. A typical naan recipe involves mixing white flour with salt, baking soda and enough yogurt to make a smooth, elastic dough which is cooked in a tandoor (a clay oven) in which burning wood is used for cooking. Salt and baking soda added in all of these native naan recipes, contributes to further salt ingestion along with the Noon Chai.

The per capita daily consumption of Noon Chai ranges from 200 to 2500 mL and most of the population likes to take it at high temperature particularly during winters [11]. The practice of Noon Chai intake is slightly more prevalent in rural regions where agriculture is main profession. In these areas besides morning and afternoon, it is also often taken during working hours. Noon Chai intake increases in winter particularly in rural regions for giving warmth and indoor enjoyment. There is a belief in the Kashmiri folklore that Noon Chai is refreshing in the heat and resists the cold in winters. It is believed to have digestive properties, possibly because of soda bicarbonate content.

Salt intake in Noon Chai: A critical issue in interpreting relation of salt intake (in Noon Chai) with stomach cancer risk, is variation in salt consumption levels across the population in Kashmir valley. The amount of salt added to Noon Chai depends on subject’s perception for preference for salt use. Amount of salt intake with Noon Chai varies with amount of salt added and number of cups taken daily. A high consumption of salted tea (>4 cups a day) is independently associated with high risk for gastric cancer [8]. Also, the naan which is served with Noon Chai has added salt which contributes to salt intake.

Salt intake and gastric cancer in other parts of world: High sodium intake appears to be responsible for the high rate of gastric cancer in cultures where processed salted foods are consumed frequently. Excessive salt intake has been identified as a possible risk factor for gastric cancer in many correlation studies and case-control studies [22–25]. There is a graded positive association between salt consumption and incidence of gastric cancer. A progressive significant increase in risk of gastric cancer is observed from moderately high to high salt intake in comparison with low salt consumption [24].

An epidemiological survey of the Japanese nationals who migrated from overseas and who changed their habit of salted food had a sharp decline in the incidence of gastric cancer with time [25]. The role of diet in incidence of cancer was seen in Hawaii where gastric cancer was found to be associated with the consumption of salted fish [26]. The consumption of salted mushrooms had been found to increase the risk of gastric cancer in Lithuania [27]. High intake of salty meals was associated with higher risk of gastric cancer in Serbia [28]. Salted black beans are the staple food of most Costa Ricans which has one of the highest stomach cancer levels on record in international literature [29].

Thermal effects of Noon Chai: Tea consumed at high temperature may cause thermal injury to gastric mucosa [30–32]. Noon Chai if taken at high temperature, may cause thermal injury to the gastric epithelium [6, 11]. Inflammatory response then leads to inflammation and generation of free radicals of oxygen and nitrogen that promote carcinogenesis [33]. A large case-control study in Mongolia on healthy controls reported almost three times increased risk for gastric cancer with drinking hot tea [34]. People who preferred drinking of strong and hot tea were at higher risk of gastric cancer than those who did not [35, 36].

Carcinogens in Noon Chai: The frequent consumption of hot salted tea is shown to result in exceptionally high exposure to methylamine, ethylamine, diethylamine, pyrroldine, and methylenzylamine, an animal carcinogen [10, 37] besides the presence of
preformed N-nitrosodimethylamine (NDMA), a considerable amounts of N-nitrosoproline (NPRO) (360 μg/kg) and N-nitroso piperolic acid (NPIC) (5870 μg/kg) along with three yet unidentified non-volatile N-nitroso compounds are formed on preparing salted tea by local methods in Kashmir [10]. Nitrosamines have been shown to act as potent carcinogens in a wide variety of animal species, and there is no reason to assume that humans are resistant [38]. N-nitroso compounds, their possible endogenous formation due to high consumption of salted tea may be a critical risk factor for the high occurrence of gastric cancer in Kashmir [6, 7, 10, 37].

Tannins isolated from salted tea have been found to give a positive result in ribosomal degradation tests and extract showed genotoxicity to rat hepatocytes in alkaline elucidative assays [10]. Tannins may also be a risk factor causing gastric cancer in Kashmir.

**Action of salt on stomach:** Salt exerts an enhancing effect at both initiation and promotion steps within the two stage model of gastric carcinogenesis [39]. A temporal corollary of precancerous changes that eventually leads to the development of gastric cancer involving a high salt diet results in chronic active gastritis [40, 41]. In some cases, this may lead to atrophic gastritis with loss of glandular tissue followed by intestinal metaplasia, dysplasia, early gastric cancer and eventually advanced gastric cancer.

Chronic hypergastrinemia by high salt intake can synergize with helicobacter infection and lead to eventual parietal cell loss and progression to gastric cancer [42]. A high-salt intake strips the lining of the stomach and may make infection with *H. pylori* more likely or may exacerbate the infection. On molecular level, high dietary salt intake may potentiate CagA (*H. pylori* gene) expression and enhance the ability of CagA to translocate into gastric epithelial cells and enhance the ability of *H. pylori* to alter gastric epithelial cell function [43].

**CONCLUSION**

Noon Chai is related to the risk of gastric cancer in Kashmir. A dietary modification involving less Noon Chai intake could be practical strategy to decrease gastric cancer in Kashmir.

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**Author Contributions**

Imtiaz Wani – Substantial conception and design, Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published

Fazl Q Parray – Substantial conception and design, Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published

Rauf A Wani – Substantial conception and design, Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published

Sameer H Naqash – Substantial conception and design, Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published

Khursheed A Wani – Analysis and interpretation of data, Final approval of the version to be published

Ajaz A Malik – Analysis and interpretation of data, Final approval of the version to be published

Nisar A Choudri – Analysis and interpretation of data, Final approval of the version to be published

Majeed A Wani – Analysis and interpretation of data, Final approval of the version to be published

Nawab A Khan – Acquisition of data, Critical revision of manuscript, Final approval of the version to be published

Tariq A Sheikh – Acquisition of data, Critical revision of manuscript, Final approval of the version to be published

**Guarantor**

The corresponding author is the guarantor of submission.

**Conflict of Interest**

Authors declare no conflict of interest.

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Combination therapy with vitamin D₃, progesterone, omega-3 fatty acids and glutamine reverses coma and improves clinical outcomes in patients with severe traumatic brain injuries: A case series


ABSTRACT

Introduction: Traumatic brain injury (TBI) is a major public health problem and a leading cause of death and disability in the United States. Management of patients with TBI has changed very little over the last 20 years. Case Series: A case series of three patients with severe TBIs who were aggressively treated with vitamin D₃, progesterone, omega 3-fatty acids, and enteral glutamine for six weeks, termed neurocritical augmentation for traumatic brain injury (NATBI), with very favorable outcomes. Conclusion: A large clinical study trial using these four supplements (NATBI) together is warranted.

Keywords: Traumatic brain injury, Vitamin D₃, Omega-3 fatty acids, Loveza, Progesterone, Cerebral edema, Glutamine

INTRODUCTION

Traumatic brain injury (TBI) is a major public health problem. According to CDC it affects over 1.7 million people annually in U.S. with 275,000 hospitalizations and 52,000 deaths [1]. The medical cost for treating TBI patients in the United States in 2010 was $76.5 billion and rising annually [1]. Primary causes for TBI include the following: motor vehicle crashes, falls, assaults and sports or recreation-related injuries (concussions). Finding the right treatment to reduce mortality rates and improve the clinical outcomes in TBI patients has been elusive.

Management of patients with TBI has changed very little over the last 20 years. Advancements in the treatment of TBI requires great understanding of the biochemical mechanisms of the brain during a normal resting state as well as the metabolism after a severe traumatic event. Brain metabolism is markedly altered during TBI. After the initial insult to the brain, the brain’s metabolism is altered and can increase up to 140% of its normal metabolism.

Vitamin D (a steroid hormone) and omega-3 fatty acids (an essential fatty acid) are both very powerful anti-inflammatory agents that reduce cerebral edema.
and swelling. Glutamine becomes an essential amino acid during stress and produces the extra glucose (via the Cori cycle) that is used by the injured brain and the extra glucose used by the immune response system to fight off infection during stress. Progesterone (also a steroid hormone) is a neuroprotector of injured brain cells and potentiates the effect of vitamin D.

These agents are all immune modulators which work synergistically to prevent secondary brain injury by limiting or decreasing inflammation; an increasing well-recognized cause of ongoing brain swelling after a primary injury. They are also neuroprotectors that makes the neurons more resistant to stress, ischemia, hypothermia, hyperthermia, hypoglycemia, hyperglycemia, hypotension, and hypertension. Immune modulation with nutritional supplements is a rapidly advancing field with a very promising future in treating TBI as well as other critically injured/ill patients.

We present a case series of three patients with severe TBIs who were aggressively treated with vitamin D₃, progesterone, omega-3 fatty acids and enteral glutamine for up to six weeks, termed neuroprotective augmentation for traumatic brain injury (NATBI), with very favorable outcomes [2–5]. Patients in a coma with severe TBI (Glasgow Coma Score <8) who were admitted to a Level I trauma center were evaluated in a prospective observational study. Patients were treated with a neuroprotective combination of vitamin D₃, omega-3 fatty acids, progesterone, and glutamine initially via a nasogastric tube and later orally for six weeks. Primary outcomes were mortality rate and return to recovery which was defined as a Glasgow Coma Score (GCS) of 10 or greater. NATBI protocol works on multiple levels and neuroprotective pathways in TBI patients by down regulating cytokine production, preventing oxidative stress (free radical oxygen formation), decreasing cerebral edema, and inflammation, thus limiting secondary brain injury in contradistinction to progesterone therapy alone (Protect III study) [3–5]. In addition, our NATBI regimen is relatively inexpensive, safe, and very effective at reducing brain and systemic inflammation post-injury.

**CASE REPORT**

**Case 1:** Patient 1 was a 17-year-old female restrained driver, who was involved in a single car, multiple rollover motor vehicle crash with a 10 foot ejection. She presented to the Emergency Department intubated and unresponsive with a GCS of five out of fifteen. Her physical exam was notable for a blood pressure of 105/56 mmHg, pulse of 87 beats/min, temperature of 37.7 °C, respiratory rate of 20/min, and oxygen saturation of 100% on the ventilator. Her secondary survey revealed unequal pupils with discordant reactivity. Her right pupil was 8 mm and non-reactive to light and her left pupil was 3 mm and reactive to light. Ominously, she was noted to have decerebrate posturing of both the upper and lower extremities bilaterally. On further examination, a 5 cm laceration to the right lower anterior thigh was identified and repaired. Her Focused Assessment Sonogram for Trauma (FAST) exam was negative.

The initial computed tomography (CT) scan of her head revealed multifocal, punctuate brain hemorrhages, consistent with a diffuse axonal injury (DAI) (Figure 1). CT scans of the cervical spine, chest, abdomen, and pelvis revealed bilateral spinous process fractures of C7, T1, and T2, a mid sternal body fracture, bilateral pulmonary contusions, and a distal right clavicle fracture. She also sustained a cardiac contusion associated post-injury arrhythmias, which were treated conservatively. An external ventricular drainage device was placed by neurosurgery to help monitor and manage her intracranial pressure and maintain her cerebral perfusion pressure within acceptable limits.

Upon her admission to the surgical intensive care unit (SICU), she was started on a regimen of vitamin D₃ 50,000 IU, progesterone 20 mg, omega-3 fatty acids 2 grams (Loveza), and enteral glutamine 20 grams via her nasogastric tube (NGT). Her decerebrate posturing resolved in less than 24 hours. By hospital day 3, she was able to follow simple commands while off sedation.

Her GCS and clinical status continued to improve and she was able to be extubated on hospital day-9. She was discharged to inpatient rehabilitation on hospital.
day-18. Although her GCS improved to 12 prior to rehab transfer, some residual right sided weakness remained. She rapidly progressed to a GCS of 15 during her recovery and was discharged home from inpatient rehab doing well after one month. In less than three months after her initial insult, she has returned to school full time and is completing her senior year of high school with her right sided weakness essentially resolved.

**Case 2:** Patient 2 was a 31-year-old male who was brought to the Emergency Room by ambulance due to altered mental status (AMS) and a witnessed seizure following an assault with suspected head trauma. He suffered blunt force trauma to his head secondary to being struck with a brick. His primary and secondary examinations were unremarkable with the exception of a GCS of 9. His vital signs were within normal limits and his hemodynamics were stable.

His initial CT scan of the head revealed a bilateral frontal intraparenchymal hemorrhage, with left frontal, parietal, temporal subdural hematomas (SDH), a left frontal subarachnoid hemorrhage (SAH) with a 7 mm right to left midline shift, cerebral edema and effacement of left frontal horn and right temporal hematoma (Figure 2), computed tomography scan of the cervical spine, chest, abdomen and pelvis were unremarkable.

On tertiary survey, his past medical history was noted to be significant for human immunodeficiency virus (HIV positive) infection, hepatitis A, syphilis, shingles, and alcohol abuse. His CD4 T-cell count on admissions was 46 mm$^3$ (normal mm$^3$$>500$), consistent with a diagnosis of acquired immunodeficiency syndrome.

His mental status declined rapidly during his initial evaluation and management period in the ED, and he was taken to the operating room by neurosurgery for an emergent depressive hemicraniectomy. Due to the severity of his head injury and his multiple comorbidities, the patient’s prognosis was deemed to be very poor by the neurosurgery service. Upon admission to the SICU, his GCS was 3T out of 15. He was immediately started on our NATBI protocol, with a regimen of vitamin D3, progesterone, loveza, and glutamine via orogastric tube.

His postoperative course was complicated by acute respiratory distress syndrome (ARDS), ventilator associated pneumonia (VAP) and acute sepsis. However, his condition improved with intravenous antibiotics, ventilatory management, and nutritional support. His GCS continually improved over the course of his ICU stay and he was able to be discharged to a long-term rehabilitation facility on hospital day-18 with a GCS score of 11T breathing spontaneously via his tracheostomy. One month after discharge, he was evaluated in the trauma clinic and was noted to have a GCS of 15. He was tolerating a regular diet, was evaluated without assistance, and adjusting very well to home life. The only deficits reported were some memory loss, which he noted was improving on a daily basis.

**Case 3:** Patient 3 was a 23-year-old female who was an unrestrained passenger involved in a single car motor vehicle collision (hit a tree) with a fatality at the scene. Patient had a GCS of 3 out of 15 in the field with decerebrate posturing, according to emergency medical personnel. She was immediately intubated by paramedics at the scene.

Physical examination in the emergency department showed that the patient was hemodynamically unstable with a blood pressure of 90/60 mmHg, pulse of 128 min, respiratory rate of 18 min, and oxygen saturation of 100%. She required four units of packed red blood cells plus three liters of isotonic crystalloid to become hemodynamically stable.

Her pupils were 5 mm and sluggishly reactive to light bilaterally. She had massive facial edema and swelling. Her endotracheal tube was intact and in good position and confirmed with positive end-tidal CO2.

Computed tomography scans of the head, face and cervical spine revealed the following: diffuse SAH over the left frontoparietal lobes, cerebral edema, SDH, transtentorial herniation, tonsillar herniation (Figure 3); fracture of the right mandibular angle, body, and parasymphyse; but there was no cervical spine fracture or dislocation.

Computed tomography scans of the chest, abdomen, and pelvis showed the following: a mid sternal fracture,
bilateral, multiple rib fractures, left pulmonary contusion, Grade 1 splenic laceration and a left acetabular fracture. X-rays of the lower extremities revealed a right, distal tibia/fibula fracture, which was stabilized by orthopedic surgery.

An external ventricular drainage (EVD) monitor was placed and revealed that the patient had an opening cerebral perfusion pressure of 27 cm H2O. Standard protocol for patients with elevated intracranial pressure (ICP) was initiated. Patient was admitted to the SICU and started on our NATBI protocol consisting of vitamin D3, progesterone, omega-3 fatty acid, and glutamine.

She had a prolonged hospital course which was complicated by refractory elevation of her ICP, prolonged coma with a depressed GCS of 4T to 8T, VAP, urinary tract infection (UTI) with urosepsis, candidemia with fungal sepsis and acute renal insufficiency.

Patient was transferred on hospital day –109 with a GCS of 8T and breathing spontaneously on tracheostomy collar to a long-term rehab facility. She continued to make satisfactory improvement during rehab, and was discharged from the long-term rehabilitation facility to home six weeks later with a GCS of 12 to 13, talking, following commands, and eating with assistance. Patient was not ambulating when discharged and will require extensive ongoing physical therapy. She was lost to follow after discharge from the rehabilitation center.

All three patients, who had a very poor prognosis, survived their severe TBI and had a return of recovery to a GCS of 15 out of 15. Six months follow up revealed that all three patients, short-term memory lost had resolved.

**DISCUSSION**

Emerging understanding of NATBI has a very promising future in the treatment of TBI. Vitamin D (classified as a vitamin) is actually a steroid hormone with pleiotropic effects, which includes its action as immune modulator [6]. Of note, receptors for vitamin D are located on every cell and tissue of the human body including brain tissue. Vitamin D has been discovered to be very important in immunomodulation, regulation of inflammation and cytokines, such as IL-1 beta and tumor necrosis factor-alpha (which increases brain cell edema), cell proliferation, cell differentiation, apoptosis, and angiogenesis, in addition to the traditional calcium, magnesium, phosphate homeostasis and bone formation. Consequently, deficiency of vitamin D affects more than 70% of the United States general population, and has been found to be associated with worsening of many inflammatory conditions [7–9].

Even more important to brain health, vitamin D binds receptors in brain cells and helps to produce heat shock proteins (HSP), which act as chaperone proteins that make the cells more resistant to stress [10, 11]. Heat shock proteins help brain cell proteins maintain their 3-D shape/conformation during stress, ischemia, hyperthermia, hypothermia, hyperglycemia, hypoglycemia, hypertension, and hypotension. Loss of 3-D protein shape by neuronal cells results in loss of cellular function, which predisposes the brain cells to apoptosis and cell death [12]. All cells in the human body are capable of producing HSP. Therefore, low vitamin D levels may be associated with lower levels of HSP. Thus, vitamin D deficient neuronal cells are less likely to survive a stressful event, such as trauma or ischemia to the brain. HSP has anti-apoptotic (prevents programmed brain cell death) and anti-inflammatory properties, which also decreases cerebral edema [13, 14]. Heat shock protein plays a very important and central role in brain cell survival and resiliency after a traumatic brain injury.

Recent research has shown that progesterone is a neuroprotectant that works synergistically with vitamin D in protecting the nerve cells from injury. Progesterone has been shown to protect the brain from traumatic injury and is now in Phase III clinical trials. However, studies have shown that progesterone's beneficial effects can be ameliorated in vitamin D deficient patients. Vitamin D can modulate neuronal apoptosis, trophic
factors, inflammation, oxidative stress, excitotoxicity, and myelin and axon repair. Low dose vitamin D hormone plus progesterone has been demonstrated by Hua et al. to improve performance in acquisition, more effectively than progesterone alone, suggesting that a lower dose of activated vitamin D may be optimal for combination therapy. Their data support that the combination of progesterone and vitamin D is more effective than progesterone alone in preserving spatial and reference memory [15].

According to the CDC, up to 80% of the United States population is omega-3 fatty acid deficient. As 30% of human brain tissue is made up of omega-3 fatty acid, emerging evidence suggests that supplementing TBI patients with omega-3 fatty acid may help the injured brain to repair itself. This makes omega-3 fatty acid a very essential adjunct in the treatment of severe TBI. A broken brick wall is repaired with bricks and not straw. The same analogy applies to the brain. It needs omega-3 fatty acid to heal properly. Also, omega-3 fatty acids are anti-inflammatory and works very well with vitamin D3 in down-regulating inflammation, which counteracts cerebral edema and swelling.

Thus, vitamin D deficiency and omega-3 fatty acids deficiency may work synergistically to worsen outcome in patients with TBI. As both are very prevalent in the general US population and even more pronounced in critically ill patients with TBI, in our opinion, at-risk patients should be routinely supplemented with vitamin D and omega-3 fatty acids. In fact, vitamin D levels less than 17.8 ng/mL is shown to be associated with a 28% increased all-causes risk of premature death [6]. Equally important, omega-3 fatty acid deficiency is associated with over 96,000 preventable deaths in the US annually according to a recent report from the CDC. Therefore, nutritional deficiencies of these two supplements can potentially have a grave impact on the clinical outcomes of TBI patients.

On the other hand, glutamine is a non-essential amino acid that becomes a conditionally essential amino acid during periods of stress. Glutamine is the most abundant amino acid and in human skeletal muscle. During stress, glutamine is used as the primary energy source for rapidly dividing cells and is used by the liver to make glucose via gluconeogenesis to supply glucose to the brain, red blood cells, enterocytes of the small bowel, and the cell of the immune system. Glutamine also works synergistically with vitamin D3 to increase HSP70 [16, 17]. Thus, adequate levels of glutamine and vitamin D3 are needed to produce a optimal concentration of HSP70 in brain cells, which potentially work together to protect the injured brain from ongoing insult or injury.

Of note, there were no side effects or complications from treating these three patients with our NATBI therapeutic regimen using the combination of supplements as noted above. Increasing data suggests that each supplement in the NATBI protocol is essential to obtaining optimal clinical outcomes in severe TBI patients. This novel approach (NATBI protocol) to treating TBI patients works by down-regulating multiple inflammatory response pathways which produces cerebral edema, upregulating HSP which helps injured brain cells survive stress of any kind, and by helping the brain to repair itself with omega-3 fatty acid.

We have reported a case series of three patients with very severe TBI’s who were managed with vitamin D3, progesterone, omega-3 fatty acids, and glutamine. All three patients were presented in a coma, and had very poor and grave prognosis based on their CT scan and neurosurgical consultation/recommendations. They are now well adjusted and have returned to their mental baselines with minimal long-term affects of TBI, other than short-term memory loss which is rapidly improving.

CONCLUSION

Our group is the first to report in literature the multi-component therapeutic regimen of vitamin D3, progesterone, omega-3 fatty acids, and glutamine as a combination therapy for moderate and severe traumatic brain injury treatment, which we have termed neurocetal augmentation for TBI (NATBI). The potential for improving clinical outcomes and potentially decreasing healthcare costs associated with TBI patients is limitless. A large clinical study trial using these four supplements together is warranted.

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Author Contributions

Leslie R Matthews – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Omar K Danner – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Y A Ahmed – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Diane M Dennis-Griggs – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Alexis Frederick – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Clarence Clark – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Ronald Moore – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Wilson DuMormay – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Ed W Childs – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
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ABSTRACT

Introduction: Although manufacturing defects related to airway equipment are not so common in practice, it risks the safety of the airway. Prompt recognition of equipment malfunction can prevent life threatening complications. Case Series: Case 1: A 25-year-old patient had anesthesia induction for septorhinoplasty surgery. After the patient was ventilated via mask and intubated without problem, peak and plateau pressures increased above 35 cm H₂O on controlled ventilation. It was noticed that the anesthetic reservoir bag did not maintain adequate compliance in a setting of high peak and plateau pressure and was thought to be due to a faulty endotracheal tube (ETT) connector. We noticed that lumen of ET tube connector was narrow and connector has been changed. After this change the relaxation of the balloon and a decrease in the ventilator pressures were noted. Case 2: A 45-year-old female patient had anesthesia induction for hysterectomy. After induction it was noted that there was a leak in the circuit, which was traced to the connection between the ETT and the inflation tube. An opening at the point of the connection of the EET with the inflation tube was detected. ETT was changed and anesthesia was maintained with no further problems. Conclusion: Difficult ventilation after successful endotracheal intubation can be due to equipment failure such as faulty ETT connector and faulty inflation tube.

Keywords: Endotracheal tube, Manufacturing defect, Leakage, Increased pressure

INTRODUCTION

Endotracheal (ET) tubes are used for facilitating ventilation during various surgical procedures performed under general anesthesia. It is a known fact that there can be manufacturing defects of ET tubes used during anesthesia application [1–3]. Equipment should be checked for defects prior to anesthetic induction. If not recognized, they can cause life-threatening problems during anesthesia application. We present two cases to remind of ET tubes’ manufacturing defects which adversely affected ventilation.

CASE SERIES

Case 1: A 25-year-old patient for planned septorhinoplasty was evaluated as American Society of Anesthesiologists grade I on preoperative examination. The patient was taken to the operating theater and vascular access was obtained. His heart rate, peripheral
oxygen saturation and blood pressure were monitored non-invasively. Anesthesia was induced with routine doses of propofol, vecuronium and fentanyl and the patient was ventilated via mask without problem. He was intubated with an ET tube with transparent and spirals (size II/7.5 mm). After it was observed that both lungs were equally ventilated, the ET tube cuff was inflated and ventilation was changed to control mode from manual mode. As peak and plateau pressures increased above 35 cm H2O during controlled ventilation, the ventilation was changed back to manual mode. The lungs were auscultated again and normal lung sounds were heard. It was detected that the breathing bag did not inflate well during expiration along with inspiratory difficulty. The anesthesia devices were checked by detaching the ET tube from the anesthesia machine, and no problem was detected. Simultaneously, the patient was ventilated via an ambu-bag. Upon inspecting the ET tube, it was noticed that the lumen of ET tube connector was narrow and connecter was immediately changed (Figure 1). As soon as it was changed, the relaxation of the breathing bag and a decrease in the peak and plateau pressures were noted. Difficulty with insertion of sylent was incidentally noted prior to intubation. In this process, there wasn’t decrease in the patient’s SpO2 level, its level continued to be 98–99%.

**Case 2:** A 45-year-old female patient in ASA II with a planned hysterectomy had anesthesia induction by routine doses of propofol, vecuronium and fentanyl for her surgery. She was intubated with a transparent ET tube of 7.5 mm (size 30/7.5). On hearing the leaking air after the patient was intubated and the cuff was inflated, the system and ET tube was checked. No problems in the system were detected. Localization of air leak was found by palpating and by listening to leakage flow. A hole at the point of the connection of the ET tube with the inflation tube was detected (Figure 2). ETT was changed and anesthesia was maintained with no further problems. In this process, there was no decrease in the patient’s SpO2 level.

**DISCUSSION**

Improperly checking anesthesia equipment and instruments prior to use can lead to patient injury. It is also associated with an increased risk of severe postoperative morbidity and mortality. Therefore, in 2008, American Society of Anesthesiologists (ASA) recommendations for equipment checkout are recommends by Sub-Committee of ASA Committee on Equipment and Facilities. According to this guide, the following steps should be verified on a daily basis: 1) auxiliary oxygen cylinder and self-inflating manual ventilation device are available and functioning, 2) patient suction is adequate to clear the airway, 3) turn on anesthesia delivery system and confirm that AC power is available, 4) availability of required monitors and check alarms, 5) pressure is adequate on the spare oxygen cylinder mounted on the anesthesia machine, 6) piped gas pressures are ≥50 psig, 7) vaporizers are adequately filled and, if applicable, that the filler ports are tightly closed, 8) there are no leaks in the gas supply lines between the flowmeters and the common gas outlet, 9) test scavenging system is function, 10) calibrate the oxygen monitor and check the low oxygen alarm, 11) carbon dioxide absorbent is not exhausted, 12) breathing system pressure and leak testing, 13) gas flows properly through the breathing circuit during both inspiration and exhalation, 14) document completion of checkout procedures, and 15) confirm ventilator settings and evaluate readiness to deliver anesthesia care. Step 2, 4, 7, 11, 12, 13, 14 and 15 should be done prior to each procedure. In our department, anesthesia equipment are prepared in accordance with ASA recommendation [4].

![Figure 1: Narrow orifice of tube connector (above) as compared to other tube of same size (below).](image1.png)

![Figure 2: An opening at the point of the connection of the EET with the inflation tube.](image2.png)
For a safe airway and ventilation, not only anesthesia machine and equipment, but also the equipment used to provide the airway should be checked. There have been case reports of various equipment failures with respect to face masks and double lumen endotracheal tubes [1, 2, 5]. Endotracheal tube manufacturing defects are usually noticed during the preoperative preparation period. In preoperative preparation period, ET tube, particularly its cuff and lumen, should be checked. If manufacturing defect is not noticed in preoperative period, it can result in serious airway obstructions [1, 3, 6–9] or air leakage during anesthesia [10–12]. If manufacturing defect of ET tube is suspected, then the equipment must be immediately changed.

An airway obstruction should be kept in mind if the airway and breathing bag pressures are increasing or if there is ventilation difficulty before enough tidal volume is reached. When this happens one should switch to manual mode from controlled mode and the anesthesia system should be checked. If there is no problem in the system, the ET tube should be checked. Herniation of the ET cuff, kinking of the tube, intraluminal plastic film or obstruction by meniscus and obliteration of the ETT connector are the known manufacturing defects that can result in difficulty in ventilation [1, 3, 6–9]. In case 1, there was a narrowing of the tube connector making ventilation difficult. There was a reported difficulty in inserting the stylet inside the tube which makes us think that this may have happened due to narrow lumen itself or total or near total occlusion or narrowing of the tube which may have been diluted by inserting the stylet.

Some manufacturing defects may cause air leakage and it may cause adverse effects on the ventilation [10–12]. When this happens, one can hear the leakage sound, the tidal volume is not enough and balloon and airway pressures decrease. Two cases reported in the literature were similar to our second case where they reported air leakage from the point of connection of inflation tube and air leakage due to an asymmetric cuff [10–12].

CONCLUSION

Although manufacturing defects are not so common, they risk the safety of airway. Therefore, it is important to check anesthesia equipment against a possible manufacturing defect as well as anesthesia machine in preoperative period. When ventilation problems happen after a successful intubation, ventilator, anesthesia instruments (ETT, mask, LMA), devices and lungs should be checked. Problem should be identified with a systematic approach and fixed. Prompt recognition of equipment malfunction can prevent life-threatening complications.

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Editor’s Note

Name of the manufacturing companies and LOT number has not been rewarded in the article.

Author Contributions

Ayse Belin Ozer – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content; Final approval of the version to be published

Omer Lutfi Erhan – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES

Clear cell adenocarcinoma of vagina associated with uterine prolapse: A rare entity

Rekha Sachan, Pooja Gupta, Patel ML

ABSTRACT

Introduction: Clear cell adenocarcinoma usually affects the cells of the female genital tract. In this cancer, cells become vacuolated and filled with glycogen hence it is called clear cell. This cancer is insidious and slow growing. Here we report a case of uterine prolapse with polypoidal growth in the vagina which on histopathological examination was diagnosed as a case of clear cell adenocarcinoma of the vagina. Case Report: A 27-year-old women with three previous vaginal deliveries presented with complaint of something coming out per vaginum, associated with bleeding and discharge per vaginum for the last one year. She was diagnosed as a case of uterine prolapse with vaginal clear cell adenocarcinoma. Conclusion: Based on the cases mentioned previously and one reported here vaginal clear cell adenocarcinoma is a rare entity and can also exist even without diethylstilbestrol (DES) exposure in utero.

Keywords: Clear cell adenocarcinoma, Vagina, Uterine prolapsed, Excisional biopsy, Radical hysterectomy

INTRODUCTION

Clear cell adenocarcinoma is a rare type of adenocarcinoma. It usually affects the female genital tract. Most commonly it affects the ovary, cervix, and urinary tract including the kidneys [1]. It has been strongly linked to in utero exposure of the female fetus to diethylstilbestrol (DES) [1, 2]. Primary vaginal adenocarcinoma unrelated to DES exposure is very rare [3]. It usually involves anterior or lateral wall of upper vagina or cervix of young females usually in their late teens or early twenties. We hereby report a case of 27-year-old female who presented with clinical features suggestive of uterine prolapse with a polypoidal growth which on histopathology was diagnosed as a case of vaginal adenocarcinoma (clear cell variety).

CASE REPORT

A 27-year-old female patient presented with complaints of something coming out of vaginum for the past one year. It was associated with bleeding and discharges per vaginum for the past one year. She had three previous vaginal deliveries, without any history of abortion. Last delivery was one and half years back. There was no family history suggestive of DES or any toxins exposure. Pallor was present on general examination and all other systems were within normal limits. Inspection of the perineum revealed a polypoidal growth of about 4x4 cm size coming out of the introitus, originating from the anterior vaginal wall (Figure 1). It was firm to soft in consistency and bleed on touch. On per-speculum examination second degree cervical...
descent was present. Growth seemed to arise from the upper one third of anterior vaginal wall in proximity to urethral opening. Cervical lips were free form growth (Figure 2). Vaginal examination revealed a normal sized uterus with no palpable adnexal pathology. On per rectal examination rectal mucosa was free and no parametrial thickening was present. Certain routine and specific investigations were conducted to determine the origin of the growth. Cystoscopic findings were within normal limits. Color Doppler sonography of abdomen did not reveal any evidence of metastasis. Chest X-ray, upper GI endoscopy and colonoscopy were within normal limits. On ultrasonography, excretory system (kidney, urinary bladder and urethra) was normal. On the basis of history and examination the following differential diagnosis were kept in mind: prolapse of uterus with vaginal growth, inversion of uterus, infected cervical polyps, leiomyosarcoma of cervix and vagina. Patient was planned for excisional biopsy of the growth under anaesthesia. Histopathologic examination of the vaginal growth showed stratified squamous epithelium with subepithelial zone showing malignant epithelial tumor composed of papillae and acini disposed in tubulocystic pattern. The cells were round to oval, with high nucleocytoplasmic ratio and vesicular chromatin, prominent nucleoli, hobnailing and areas of cytoplasmic clearing suggestive of clear cell adenocarcinoma of vagina, without surrounding vaginal adenosis (Figures 3, and 4). Based on the above findings staging of this case was T1N0Mo (Stage 1). A radical hysterectomy with vaginectomy with lymph node dissection was done, as patient was willing for removal of the uterus. Histopathology of the specimen showed normal uterine histology. Surrounding vaginal tissue histology showed stratified squamous epithelium with underlying stroma showing fibrocollagenous tissue, blood vessels, smooth muscle bundles and interspersed nerve bundles without any malignant cells. Sections from the cervix showed normal histology and lymph nodes were negative for the malignancy. Patient was followed up at three monthly intervals up to nine months. Clinical examination, pap smear and ultrasonography were unremarkable on follow-up.

**DISCUSSION**

Vaginal clear cell adenocarcinoma is common in late teens and early second decade in females whose mothers took diethylstilbestrol (DES) during pregnancy [4]. Clear cell adenocarcinoma of vagina account for about 5–10% of all vaginal cancers [3]. The absolute risk...
of clear cell adenocarcinoma of vagina in the daughters of DES exposed mothers is 1 in 1000 [5]. In our patient's mother there was no past history of in utero exposure to DES. Clear cell adenocarcinoma of the vagina most frequently arises from vaginal mucosa but may arise from the cervix. It may possibly be derived from vaginal adenosis, but in our case no vaginal adenosis was seen. The tumor size is variable ranging from microscopic to large size. Most of the larger tumors are polypoidal or nodular in appearance and are mostly superficially invasive. Our case is rare because the patient developed primary vaginal clear cell adenocarcinoma, which was not associated with DES exposure in the mother. Furthermore it is the rare reported case in Indian scenario. The diagnosis was made based on the excisional biopsy of vaginal growth which on histopathological examination showed characteristic rounded cells with clear appearance of cytoplasm, high nucleocyttoplasmic ratio with hobnailing and normal histopathologic examination of cervix, uterus and ovaries after radical hysterectomy with vaginectomy. In this cancere, the cells become vacuolated and filled with glycogen, hence the term ‘clear cell’ is given to this cancer. There are a few reported cases of vaginal clear cell adenocarcinoma in which there was no in utero exposure to diethylstilbestrol. Wantanabe et al. also reported a case of advanced primary vaginal clear cell adenocarcinoma not associated with DES exposure in a postmenopausal woman [2]. Nordqvist et al. reviewed twenty one cases of vaginal and cervical clear cell adenocarcinomas, out of which thirteen had no prior exposure to DES [6]. Zeshann-ud-din et al. reported a case of vaginal clear cell adenocarcinoma with associated Mullerian duct anomalies, renal agenesis and situs inversus, with no known in-utero exposure with diethylstilboestrol [7], but in our case no Mullerian and renal anomalies were found.

CONCLUSION

Vaginal clear cell adenocarcinoma is a rare entity and can also exist even without diethylstilbestrol exposure in utero.

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Author Contributions

Rekha Sachan – Substantial contributions to conception and design, acquisition of data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published

Pooya Gupta – Substantial contributions to conception and design, acquisition of data, revising it critically for important intellectual content, Final approval of the version to be published

Patel ML – Substantial contributions to Drafting the article

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES

Strangulated incisional hernia after suprapubic catheter insertion: A case report

Mohanraj Harilingam, Hany Balamoun, Fady Yanni

ABSTRACT

Introduction: Hernia through the suprapubic catheterization site is extremely rare. Attention is required for such hernias as they can get easily complicated by obstruction and strangulation due to its narrow neck. These are life-threatening complications. Case report: We report a case of a 65-year-old man who presented with a strangulated incisional hernia through the SPC site. He was managed successfully by urgent surgical intervention. Conclusion: Incisional hernia through the suprapubic catheterization site should not be forgotten during examination of patients with abdominal pain, to ensure prompt surgical intervention in case of strangulated hernia.

Keywords: Hernia incisional, Suprapubic catheterization (SPC)

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INTRODUCTION

Suprapubic catheter (SPC) insertion is a very common urological procedure. It is practiced widely by a variety of specialties to manage long standing voiding dysfunction or neuropathic bladders. The majority of these procedures are performed without any complications. Common postoperative complications of SPC insertion include hematuria, urinary tract infections and catheter obstruction [1]. We present a rare case of strangulated incisional hernia through the SPC site.

CASE REPORT

A 65-year-old man of Asian origin presented with sudden onset of abdominal pain, abdominal wall lump and vomiting of six hours duration. Four years back, he had transurethral resection of the prostate. Two years after prostate resection, he had retention of urine due to urethral stricture, for which he had SPC. On examination he was hemodynamically stable. Abdominal examination revealed a tender swelling at the SPC scar site which was about five cm above the symphysis pubis. The swelling was tense and tender (Figure 1). Clinical diagnosis of a strangulated incisional hernia through the SPC scar was made. Plain abdominal X-ray confirmed small bowel obstruction. The decision was made to undertake an urgent local exploration of the swelling. The local exploration was done through a vertical incision in the abdominal wall. It confirmed a hernial sac with strangulated small bowel within the sac (Figure 2). The defect was three cm in diameter. It was widened for exposure of the surgical site. Small bowel resection and a hand sewn end to end primary bowel anastomosis were performed. The hernial defect was
carcinoma has also been reported to occur [4, 5]. Incisional hernia through a suprapubic catheter site is a rare but important complication. The possible explanation for the cause of hernia may be the trocar traversing through the peritoneum before entering into the distended bladder. Once the distended bladder decompresses and returns to the pelvis, it provides an opportunity for the peritoneal contents to herniate through the potential weakness of the trocar site [6]. A history of persistent discomfort at the site of a previous suprapubic catheter insertion, coupled with a palpable rectus sheath defect and positive cough impulse form the mainstay of diagnosis [7].

Hernias from the SPC insertion site need to be surgically repaired as the neck is narrow and the hernia is prone to obstruction. Preventive measures to avoid complications include making a low incision rather than a higher incision for the insertion of the trocar to void the distended bladder. Examination of the trocar site is necessary in a patient with a previous history of SPC insertion presenting with abdominal pain and discomfort. Urologists need to be aware of this potential complication of SPC insertion [8].

CONCLUSION

Incisional hernia through the suprapubic catheterization site should not be forgotten during examination of patients with abdominal pain to ensure prompt surgical intervention to prevent bowel strangulation rupture and other life-threatening complications.

*********

Author Contributions
Mohanraj Harilingam – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Hany Balamoun – Substantial contributions to conception and design, analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Fady Yanni – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES

Caspofungin may effective for initial treatment of pulmonary aspergillosis

Alper Sener, Senol Kobak, Timucin Alar,
Ugur Gonlugur, Asli Muratli

ABSTRACT

Introduction: Invasive aspergillosis mostly involves lungs. Patients may present with fever, cough, hemoptysis and many other sign and symptoms. Underlying conditions that compromises pulmonary and immune responses to inhaled Aspergillus species serve as risk factors for pulmonary invasive aspergillosis. We are presenting a case of simple aspergilloma, successfully treated by caspofungin and surgery. Case Report: A 64-year-old male patient has rheumatoid arthritis for ten years. Since three months he had interstitial pulmonary disease secondary to rheumatoid arthritis and hemoptysis. We performed microbiologic and pathologic investigations of the brochoalveolar lavage after computedized tomography for diagnosis of aspergillosis. Caspofungin was used for initial treatment of simple aspergilloma and also for follow-up treatment after apical wedge resection. The patient was discharged from the hospital at eighth week on itraconazol antifungal therapy. Conclusion: Three classes of drugs are available for the treatment of aspergillosis: polyens, azoles and echinocandins. Echinocandins have activity against Aspergillus spp. But have a limited role because they have not been studied for initial treatment of invasive aspergillosis.

Keywords: Antifungal treatment, Aspergillosis, Caspofungin

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INTRODUCTION

Aspergillus spp. may cause a variety of pulmonary diseases, depending on immune status and the presence of underlying lung disease [1]. Aspergillus species are ubiquitous in nature and inhalation of infectious conidia is a frequent event. Tissue invasion is uncommon and occurs most frequently in the setting of immunosuppression associated with therapy for hematologic malignancies, hematopoietic cell transplantation or solid organ transplantation [2]. Most invasive infections are caused by members of A. fumigatus species complex. Caspofungin is an echinocandin class antifungal drug. Echinocandins target fungal cell glucan synthesis by competitively inhibiting the beta-1,3-D-glucan synthase enzyme complex in susceptible fungi. This enzyme complex is
composed of at least two components: catalytic subunits called Fks1p and Fks2p, and a GTP-binding protein, Rh1, which regulate the activity of glucan synthesis [3]. Echinocandins are widely used for the treatment of invasive candidiasis, especially in critically ill and neutropenic patients and less commonly in salvage regimens for invasive aspergillosis [4, 5]. Recent studies conclude that surgical resection should be considered for all patients with pulmonary aspergillosis who have acceptable pulmonary reserve. Echinocandin class of antifungal drugs have not been studied adequately as initial antifungal treatment of pulmonary aspergillosis. In this case we used caspofungin alone for initial treatment of pulmonary aspergillosis and after surgery.

CASE REPORT

A 64-year-old male patient presented to us with hemoptysis for one week. He was a farmer by profession and had rheumatoid arthritis for ten years. He was taking prednisone 20 mg per day for last three months for interstitial pulmonary disease secondary to rheumatoid arthritis and azathioprine 100 mg per day for anti-CCP positivity for last one month. He had fifteen days of cough and productive sputum and was given amoxicillin-clavulonate 2 gm per day by his family practitioner. He was referred to our hospital with hemoptysis of one week duration. The complete blood cell counts and blood biochemistry were within normal range except for high erythrocyte sedimentation rate (110 mm/1st hr) and high C reactive protein (13.7 mg/dL). Chest radiograph showed a suspicious mass in the right apical lobe. Bronchoscopy were done for investigating tuberculosis by acid fast staining of brochoalveolar lavage (BAL). Computed tomography (CT) scan was performed because of suspicious mass on chest radiograph. There was a 5.8x5.1x8.6 cm right apical pleural based cavity lesion with thick irregular wall. It was forming a small air crescent (Figure 1). Asid fast stain of BAL was negative. Routine staining of sputum and BAL showed hyphae with septa. Microbiologic culture showed A. fumigatus. We diagnosed the case as simple aspergillosa and began conventional amphotericin B treatment. Patient has allergic rash after amphotericin B infusion and on third day of hospitalisation he has abdominal pain. Despite his symptoms, hepatic transaminases were within normal range. Acute cholecystitis was diagnosed after general surgery consultation and abdominal ultrasonography showed sludge in the gallbladder. We decided on caspofungin treatment of intravenous loading dose (70 mg/day) followed by 50 mg/day infusion once daily. Patient was evaluated by chest surgery consultation and it was decided to wait for thoracic surgery consultation because of the abdominal complaints. Cholecystitis regressed in one week by medical treatment. Patient was operated at third week of antifungal treatment. Apical wedge resection was performed to remove the cavity lesion and the fungus ball. Pathologic examination of surgical specimen showed hyphae with septa, brochial invasion and interstitial pulmonary disease (Figure 2). Antifungal treatment with caspofungin continued by intravenous route for five weeks after surgery. After eighth week of hospitalisation patient was discharged with itraconazole 200 mg twice daily peroral route. At 12 weeks, antifungal treatment was stopped. Patient’s symptoms such as cough, hemoptysis and productive sputum were not seen after tenth week of treatment.

DISCUSSION

Three classes of antifungal drugs are available for the treatment of aspergillosis: polyens, azoles and echinocandins. Echinocandins have activity against Asperillus spp, but have a limited role because they have not been studied for initial treatment of invasive aspergillosis. Echinocandins are used for treatment of

Figure 1: Air crescent formation with fungus ball in the lungs.

Figure 2: Histopathologic examination shows hyphae with septa, characteristics of aspergillosis (H&E, x 200).
invasive candidiasis especially in critically ill patients and less commonly for invasive aspergillosis especially as salvage treatment. The major advantages of echinocandins relative to other antifungal drugs are their fungicidal activity against Candida spp including fluazole resistant species combined with their relatively low potential for renal or hepatotoxicity [5]. Growth of Aspergillus species is inhibited even by very low concentrations in vitro with the effects predominantly observed at apical sub-apical branching where cell wall remodelling and beta glucan synthase are most active [6, 7]. Azoles are the first agents of choice for pre- and post-operative antifungal therapy. In our patient allergic rash occurred at the beginning of treatment with conventional amphotericin B. As amphotericin B-lipid complex was not available and due to patient’s cholecytic symptoms such as nausea and vomiting antifungal treatment option was switched to caspofungin. Caspofungin was used in standard dose of 70 mg on first day and subsequent dosing was 50 mg/day. In this case we preferred to use caspofungin as initial treatment especially because of the abdominal symptoms. After one week of treatment we made an interval reiewing of treatment for using caspofungin alone or in combination. We made a decision of using caspofungin alone because of improving clinical condition of the patient. Antifungal treatment after surgery of aspergillomas has been strongly recommended by most of the authors [8]. Antifungal treatment options after surgery depend on patient’s clinical situation. Itraconazole and voriconazole are first line agents for oral antifungal treatment of aspergillosis [8]. Duration of antifungal therapy is not well defined in literature. Some authors have recommended lifelong therapy beacuse of the relapses [9]. Others suggest that a four week course of therapy post-operatively is reasonable, if the aspergilloma was fully resected without spillage. If there is spillage, then a minimum of 12 weeks therapy is recommended in an attempt to prevent pleural aspergillosis [8]. In our case, we used itraconazole for seven weeks after caspofungin intravenous therapy.

Risks and benefits of medical and surgical therapy vary with the manifestations of the disease and patients pulmonary status. Surgery is the mainstay of treatment for symptomatic patients with simple aspergilloma to prevent or treat life-threatening hemoptysis [8]. Surgery of pleural based aspergilloma will be risky. Most of the surgens prefer lobectomy or cavernectomy because of the pleural hemorrhagic complications. In our case we preferred pleural based wedge resection of cavitary lesion in order to preserve patient’s vital capacity in the future.

CONCLUSION

Aspergilloma is a fungus ball composed of aspergillus hyphae, fibrin, mucus, and cellular debris found within a pulmonary cavity. In the treatment of aspergilloma, surgery and antifungal therapy must be given together. Choice of antifungal agents is the main cause of successful treatment. Caspofungin shows satisfactory antifungal activity against aspergilloma before and after surgery despite inadequate data about its use.

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Author Contributions

Alper Sener – Substantial contributions to conception and design, Acquisition of data, analysis and interpretation of data, Drafting the article, revising it critically for important intellectual content, and Final approval of the version to be published

Senol Kobak – Substantial contributions to conception and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Timucin Alar – Substantial contributions to conception and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Ugur Gonlugur – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Asli Muratlı – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES

Non-communicating hydrocephalus decades after spinal dermoid resection

Hetal Patel, Michael A Casey, Ajeet Gordhan

ABSTRACT

Introduction: We present a case of acute non-communicating hydrocephalus due to fat droplet obstruction of the cerebral aqueduct, fifty-one years after resection of a spinal dermoid tumor. Case Report: A 71-year-old male presented with sudden onset of neck pain and gait instability. He had prior complete surgical resection of a cervical dermoid cyst in 1959. Follow up magnetic resonance imaging (MRI) in 2007 showed intracranial fat signal droplets within the posterior fossa. An MRI examination obtained in 2012 demonstrated hydrocephalus consequent to fat droplet obstruction at the aqueduct of sylvius which was not present in MRI study done in 2007. Conclusion: To our knowledge there are no published reports of delayed post spinal dermoid resection subarachnoid fat droplet migration leading to acute non-communicating hydrocephalus.

Keywords: Dermoid cyst, Hydrocephalus, Intraspinal dermoid, Cerebral aqueduct

INFORMATION


INTRODUCTION

Intracranial dermoid cyst rupture leading to non-communicating hydrocephalus is well documented in literature [1]. However, there are only two case reports of ruptured spinal dermoid cysts leading to hydrocephalus. In each of these cases fat droplet obstruction within the subarachnoid space occurred prior to resection of the cysts [2]. Cephalad migration of subarachnoid fat droplets from the spine and subsequent intracranial complications has been previously described [3]. Delayed subarachnoid fat droplet migration after spinal dermoid resection leading to acute non-communicating hydrocephalus has not been previously reported in literature.

CASE REPORT

A 71-year-old male presented with sudden onset of neck pain and gait instability. In 1959, he had multilevel laminectomies for gross total resection of a cervical extramedullary, intradural dermoid cyst. No intracranial involvement of the dermoid was noted at the time. The medical and surgical history was otherwise non-contributory.

On physical examination, he was normotensive, alert and orientated with fluent speech. He demonstrated gait ataxia without nystagmus, dysmetria or dysdiadochokinesis.
Cranial nerves were intact with no motor or sensory deficit.

Review of routine follow up magnetic resonance imaging (MRI) scan done in 2007 revealed recurrence of dermoid fat droplets within the cervical spine with cephalad migration into the subarachnoid spaces of the posterior fossa, without hydrocephalus (Figure 1A).

Brain MRI examination obtained during the current admission, revealed obstructive non communicating hydrocephalus consequent to a fat droplet within the cerebral aqueduct that was not present on the MRI study obtained in 2007 (Figure 1 B). Enlargement of the lateral and third ventricles with transependymal cerebrospinal fluid (CSF) flow was identified (Figure 2 A-C). The hydrocephalus and the patient’s symptoms resolved after a ventriculoperitoneal shunt was placed.

DISCUSSION

Dermoid tumors in the central nervous system (CNS) are very rare. Intracranial dermoid cysts account for less than 1% of all intracranial tumors and intraspinal dermoid tumors comprise about 1.1% of all intraspinal tumors [3, 4]. Dermoid cysts form in the third through fifth embryologic weeks when the neural groove is closing to form the neural tube [3]. They result from inclusion of aberrant ectodermal tissue in the spinal canal [4].

Dermoid cysts are comprised of a fibrous external capsule with a layer of stratified squamous epithelium and contain dermal tissue such as sebaceous glands, hair, sweat glands, teeth, and nails [4]. Growth of dermoid cysts over a period of time can result from

Figure 1: Sagittal T1 MRI brain sequence demonstrating (A) fat droplets within the subarachnoid space dorsal to the medulla (striped arrow), (B) a new fat droplet within the cerebral aqueduct (solid arrow) and obstructive hydrocephalus.

Figure 2: Axial MRI brain sequences with (A) solitary T1 hyperintense, (B) axial T2 hypointense, and (C) axial FLAIR hypointense punctate fat droplet signal within the cerebral aqueduct (white arrows).
accumulation of cellular debris [1]. The contents of cysts may vary from patient to patient, which can result in a wide variety of radiographic findings. It is generally accepted, however, that dermoid cysts will appear like fat on computed tomography (CT) and MRI because of their high cholesterol content, yielding hypointensity on CT scan, T1-WI hyperintensity and T2-WI hypointensity on MRI scans [1]. Spinal dermoid cysts can be intramedullary intradural extramedullary or extradural [5]. The majority of intraspinal dermoid tumors occur in the lumbosacral region and rarely in the upper thoracic and cervical regions [5].

The clinical presentation of dermoid cysts in the CNS can vary depending upon the location of the lesion. Neurological symptoms can be a result of compression of surrounding anatomical structures [3]. Patients have also been reported to present with obstructive hydrocephalus as a result of fat in the subarachnoid space from a ruptured dermoid cyst. Usually patients presenting with obstructive hydrocephalus have intracranial tumors. There are only two reported cases of hydrocephalus resulting from intraspinal dermoid tumor rupture [2]. In both these cases, hydrocephalus occurred before resection of the dermoid tumor. The patient in our case had a spinal dermoid tumor resection with a subsequent fifty one year delay before developing acute non-communicating hydrocephalus at the cerebral aqueduct.

There are few long term post-operative studies of patients with resected dermoid tumors. One study of a patient with an intracranial dystontogenic cyst removal demonstrated fat in the subarachnoid space after surgical removal of an intracranial tumor. The annual follow-up imaging did not demonstrate migration of the fat. Additionally, there were no complications as a result of the fat [3]. Recurrence of intracranial dermoids after resection has been documented in literature [6, 7].

In our case the patient had resection of a cervical extramedullary, intradural dermoid cyst. Magnetic resonance imaging in 2007 showed fat in the subarachnoid space of the posterior fossa and a patent cerebral aqueduct. The patient then developed acute onset non-communicating hydrocephalus in 2010 as result of a solitary fat droplet obstruction at the cerebral aqueduct. Delayed onset of non-communicating hydrocephalus (after five decades), in the setting of a post-spinal dermoid resection, has not been previously reported in literature.

CONCLUSION

This study illustrates the importance of ongoing imaging and clinical follow-up of surgically treated spinal dermoid cysts. In particular, increased surveillance for non-communicating hydrocephalus should be practised because spinal dermoid tumors can cause intracranial pathology through retrograde flow of fat droplets.

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Author Contributions
Hetal Patel – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Michael A Casey – Analysis and interpretation of data, Final approval of the version to be published
Ajeet Gordhan – Conception and design, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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Peritoneal encapsulation, left paraduodenal hernia with retroperitonealization of inferior mesenteric vein leading to triple obstruction

Imtiaz Wani, Khursheed A Wani, Muneer Wani, Gulzar Bhat, Mubashir Shah

ABSTRACT

Introduction: Encasing of small bowel in an anomalous accessory membrane results in peritoneal encapsulation and is a very rare cause of intestinal obstruction. It is believed to be caused by malrotation of the bowel during the 12th week of gestation and this causes the formation of an accessory sac from the peritoneum, covering the umbilicus. Left paraduodenal hernia is the most common form of congenital internal hernia. Paraduodenal hernia are caused by abnormal rotation of the midgut during embryonic development. This rarely presents as an intestinal obstruction. Congenital adhesions are rare in adults and these may lead to retroperitonealization of inferior mesenteric vein (IMV). Case Report: A case of peritoneal encapsulation and left paraduodenal hernia with congenital adhesions leading to retroperitonealization of inferior mesenteric vein (IMV) in the same patient is reported here. A 28-year-old male presented with recurrent small bowel obstruction and was diagnosed with an intestinal obstruction. Computed tomography confirmed peritoneal encapsulation and left paraduodenal hernia with features of intestinal obstruction. Excision of membrane and reduction of hernia with retroperitonealization of inferior mesenteric vein was done. Conclusion: This is the first case reported in literature where peritoneal encapsulation, paraduodenal hernia and retroperitonealization of inferior mesenteric vein (IMV) occurred together and lead to bowel obstruction. In this case, the origin of three abnormal anatomical structures must have been congenital as there was no history of prior surgery.

Keywords: Encapsulation, Paraduodenal, inferior mesenteric vein (IMV)


INTRODUCTION

The peritoneal encapsulation, first described by Cleland [1] in 1868, is a rare developmental abnormality in which part or the entire small bowel is encased in an accessory sac. It is believed to be caused by malrotation of the bowel during the 12th week of gestation and this causes the formation of an accessory sac from the peritoneum covering the umbilicus [2]. The incidence of this condition is not well known. There are not more than 50 to 60 cases described in literature [3–5]. Most of the cases remain asymptomatic; diagnosed usually as an incidental finding during surgery for unrelated conditions and only rarely present as bowel obstruction [6–8]. Paraduodenal hernia is the most common type of intestinal hernia, accounting for 30–53% of all cases.
and accounts for 0.2–0.9% of all bowel obstructions [9]. This hernia has a variable clinical manifestation and rarely may present as an acute abdomen. In adults, occurrence of congenital bands is an extremely rare condition [10, 11]. The entrapment of the intestine between the bands and the mesentery or by compression of the bowel leads to obstruction. Rarely a vessel with an abnormal position compressing on the gut could lead to bowel obstruction [3].

Clinical diagnosis of bowel obstruction is often difficult to make due to ambiguous presentation in peritoneal encapsulation, paraduodenal hernia, congenital bands and an abnormal vessel compressing the gut [12–14]. X-ray of abdomen, ultrasonography, barium study and computed tomography are contributory in diagnosis of each form of obstruction. The excision of membrane in peritoneal encapsulation is recommended even if this condition is diagnosed incidentally. All paraduodenal hernias must be repaired, including those that are asymptomatic [15]. Release of adhesions in congenital bands and the retroperitonealization of vessel leading to obstruction is recommended.

CASE REPORT

A 28-year-old male presented with generalized abdominal pain, fever and multiple episodes of vomiting. Abdominal pain was episodic and colicky and vomiting was bilious. There was no past history of trauma or surgery. Patient had a past history of three episodes of similar abdominal pain and vomiting three months back. Second episode occurred 12 days after the first episode and third episode occurred 21 days after first episode. Patient was diagnosed as a case of recurrent sub-acute intestinal obstruction by small gut stricture which was confirmed on the barium study at that time. During and after each episode the patient was managed conservatively without any surgical intervention. General physical examination during current episode showed dehydrated appearance, pulse 96/min, blood pressure 110/70 mmHg, and temperature of 99°C. Systemic examination was normal. Abdominal examination revealed distended abdomen, diffuse tenderness and a high pitch bowel sounds. Laboratory parameters were normal. X-ray of abdomen showed multiple air fluid levels. Computed tomography scan of the abdomen revealed conglomeration of gut in a thin sac suggestive of peritoneal encapsulation on the right side. There was displacement of inferior mesenteric vein and the entrapped bowel in left upper abdomen (left paraduodenal hernia) (Figure 1A–B). Gut loops were dilated and had thick walls. Exploratory laparotomy revealed encasement of bowel loops in membrane, typical of peritoneal encapsulation with inter-loop adhesions present in the bowel outside the peritoneal sac (Figure 2). Gut inside peritoneal encapsulation was dilated, which was suggestive of obstruction. Malrotations of small gut loops was seen. After careful dissection of the membrane, adhesiolysis and release of omental adhesions, a vessel lying on ileal loops, outside the retroperitoneum, was seen which was identified as inferior mesenteric vein (IMV) (Figure 3A–B). The bowel proximal to the abnormally placed IMV was dilated with thickened wall which was suggestive of obstruction. This abnormally lying IMV was constrained by adhesions to small bowel. Peritoneal encapsulation and retroperitonealization of IMV were leading to sub-acute intestinal obstruction. Further exploration revealed a loop of intestine impacted in a left paraduodenal fossa. Diagnosis of the left paraduodenal hernia was confirmed (Figure 4A–B). Incarcerated segment of bowel in left paraduodenal fossa had signs of strangulation with thickened walls leading to acute abdomen. Resection of incarcerated segment and the closure of hernial orifice was done. Retroperitonealization of anteriorly placed IMV was achieved. Postoperative period was uneventful and patient was discharged after one week. Patient is well on follow up for last 13 months.

Figure 1(A, B): Computed tomography scan of the abdomen in axial and coronal cuts demonstrating left paraduodenal hernia. Arrows are outlining the hernial sac, with clustered, sharply circumscribed loops of small bowel in the left upper abdomen. In right lower abdomen gut loops conglomerated in peritoneal sac.

Figure 2: Bowel encased in a membrane suggestive of peritoneal encapsulation.
DISCUSSION

Peritoneal encapsulation is a developmental abnormality encasing part or the entire small bowel in an accessory sac derived from the yolk sac [16]. The accessory peritoneal membrane is derived from the peritoneum of the yolk sac which withdraws into the abdominal cavity with the small bowel instead of remaining at the base of the umbilical cord. This is attached to the ascending and descending colon laterally, the transverse mesocolon superiorly and merges with posterior parietal peritoneum inferiorly. The membrane has two openings, one around the duodenojejunal flexure and the other at the ileocecal junction [7]. Peritoneal encapsulation seems to be predominant in males and is not always associated with abnormal intestinal rotation [17, 18]. The condition is largely asymptomatic but some cases may present as bowel obstruction [19, 20]. Peritoneal encapsulation very rarely leads to an acute iliac occlusion or to idiopathic sclerosing peritonitis [21, 22]. This congenital encapsulation can be diagnosed incidentally during laparotomy for other conditions and some cases are diagnosed at autopsy [3, 8]. It can occur with incomplete situs inversus and abnormal artery (both are congenital anomalies) or as an incidental findings at time of gastric surgery or during colon cancer surgery [4, 23].

Paraduodenal hernia is a rare condition in which the small bowel loops are trapped within a congenital paraduodenal fossa [24]. Males are three times more affected than females [25]. Paraduodenal hernia are divided into two subtypes: left or right paraduodenal hernia. About 75% are located on the left side [9]. A case of congenital pyloric stenosis associated with the presence of a paraduodenal hernia in an adult patient has been reported [26]. Paraduodenal hernia lacks a true hernial sac and is termed ‘prolapse’ or ‘procidentia’ rather than a hernia [27]. The left paraduodenal hernia is a result of anomalous rotation of midgut into the developing mesentery of the descending colon with mechanical forces of fluctuating intra-abdominal pressure leading to herniation in places where the peritoneum is yet incompletely fused [27, 28]. They originate at the fossa of Landzert which is just lateral to the fourth segment of the duodenum and behind the inferior mesenteric vein and ascending left colic artery [29]. The sac lies to left of the duodenum. Inferior mesenteric vessels constitute the anterior free margin of the sac [30]. Accurate anatomy is important surgically, as the inferior border of the hernia opening is safest place to widen the neck of the hernia [27].

Symptoms of paraduodenal hernia are non-specific, ranging from recurrent vague abdominal pain, nausea, vomiting and reversible obstruction leading to an acute abdomen [31]. About 50% cases follow an asymptomatic clinical course and are diagnosed incidentally; rest 50% patients with paraduodenal hernia have episodes of intestinal obstruction at some point of their lives [32]. The symptoms depend on patient’s position, with symptoms aggravating while standing and relieved in a

Figure 3(A, B): Gut loops compressed by inferior mesenteric vein lying in the retroperitoneum and site of retroperitonealisation of inferior mesenteric vein.

Figure 4(A, B): Left paraduodenal hernia impacted in paraduodenal fossa and the incarcerated gut (arrow).
supine position [33]. Spontaneous reduction apparently causes the periodic presentation of abdominal pain with normal radiographic studies [34]. The herniated small bowel loops may become trapped within this mesenteric sac. Symptoms may also result from retroperitoneal mass effect.

Adhesions are the cause of about 30–41% of all intestinal obstructions and for the small bowel obstruction, the proportion rises to 65–75% [35]. Only 3% of these are thought to be caused by congenital omental bands formed by abnormal adhesions of the peritoneal folds during embryogenesis [10, 36]. In our case, congenital adhesions might have led to constraining effect on IMV leading to its retroperitonealization which compressed the small bowel leading to obstruction. A congenital constraining of adhesions between the vein and the gut can also lead to bowel obstruction.

In view of the diagnostic difficulty based on clinical findings, imaging studies often play an important role in the diagnosis. Radiographic studies are usually normal or show non-specific features of intestinal obstruction. Plain abdominal X-rays are suggestive of mechanical bowel obstruction which was present in our case. A paraduodenal hernia may be demonstrated by an upper gastrointestinal series performed during a period of acute symptoms. Barium contrast study demonstrates sac-like mass of mildly dilated jejunal loops, left of the ligament of Treitz with mass effect causing displacement of the greater curvature and posterior wall of the stomach [37]. Ultrasonography in internal hernia shows heterogenous echoic abdominal mass with presence of changing cystic or tubular internal components and a surrounding the membrane [38]. On computed tomography (CT) scan, the findings are very non-specific. Small bowel is seen enveloped in a thin membrane and may be suggestive of peritoneal encapsulation [20, 23, 39]. The following findings are seen on CT scan in left paraduodenal hernia [27, 40, 41]:

(a) A focal cluster of small bowel loops positioned abnormally between the pancreatic body or tail and the stomach lying to the left side of ligament of Treitz or behind the descending colon and distal transverse colon.

(b) Bowel loops proximal and within hernial sac show dilatation with air fluid levels.

(c) Narrowing of efferent loops of bowel.

(d) Mass effect causing displacement and indentation of posterior wall of stomach, inferior displacement of duodenojejunal junction and transverse colon and medial displacement of the descending colon.

(e) Inferior mesenteric vein and ascending left colic artery being displaced anterolaterally or upward.

(f) Normal vascular relationship, although there might be mild displacement caused by the herniated loops.

(g) Mesenteric vessels that supply the herniated small bowel segments show crowding at the entrance of hernial sac and the vessels in hernial sac are engorged.

(h) Normal vascular relationship of superior mesenteric artery and vein are usually maintained.

Congenital adhesions involve surgical release of adhesions and bands. Treatment of peritoneal encapsulation involves the total removal of the anomalous membrane and release of both openings. Bowel resection may be necessary if the membrane cannot be stripped atraumatically or if obvious gangrene of the bowel is present. There is no re-operation reported in literature after dividing the encasing membrane in peritoneal encapsulation [4].

Surgical treatment of left paraduodenal hernia follows the basic principles of hernia surgery: reduction of the contents, restoration of normal anatomy and repair of the defect [33, 42]. A retroperitonealization of inferior mesenteric vein involves retroperitonealization of vein after release of congenital adhesions.

CONCLUSION

Occurrence of paraduodenal hernia, peritoneal encapsulation and congenital adhesions leading retroperitonealization of inferior mesenteric vein in the same patient is rare. Left paraduodenal hernia rarely presents as an acute abdomen. Occurring of three abnormal anatomical structures must have had a congenital origin as the patient had no predisposing factors for the development of these congenital anomolies.

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Author Contributions

Imtiaz Wani – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Khursheed A Wani – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Muneer Wani – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Gulzar Bhat – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Mubashir Shah – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES

A case of frontotemporal dementia

Heng Siang Ting, B M Yashodhara, Bashir Ahmad Dar, Uduman Ali Mohamed Yousuf, Adinegara Lutfi Abas

ABSTRACT

Introduction: Frontotemporal dementia is a common cause of dementia. It is easily misdiagnosed as a psychiatric illness due to its presentation with behavioral problems, mutism, language problems and in some cases with aggression and anti-social behavior. Case Report: We report a case of a 72-year-old man who presented with behavioral abnormalities, lack of personal hygiene, personality changes of gradual onset for 2–3 years, suggested by inability to take care of self, mutism, social disinhibition in the form of micturition in the presence of family members. Computed tomography (CT) scan of the brain showed atrophy of frontal and temporal lobes. The family members were counseled for the outcome and prognosis of the case. Conclusion: The diagnosis of frontotemporal dementia is made based on presentation, diagnostic criteria and brain imaging. Definite treatment is not available, but treating according to presenting complaints of the patient is helpful; e.g. if the patient has depression, anti-depressants along with family counselling about the progression of the disease and awareness about requirement for the family support are important.

Keywords: Dementia, Frontotemporal, Abnormal behavior, Poor personal hygiene


INTRODUCTION

Dementia is a common disease in middle age and elderly. It may be due to Alzheimer’s disease, vascular dementia, frontotemporal dementia (FTD) or Lewy body dementia. It may also be due to secondary causes like HIV infection, Huntington’s disease, head injury, Parkinson’s disease, metabolic and endocrine diseases, poisoning, infections like neurosyphilis, vitamin deficiencies, normal pressure hydrocephalus and pseudo-dementia due to depression.

Alzheimer disease patients typically present with problems in cognitive functions; memory, executive functions, language and constructional praxis. On brain scanning there is atrophy of the brain from occipital lobe to parietal lobe and later in the disease of the frontotemporal areas. Patients tend to get lost and find it difficult to get back to their place due to problems in visual-spatial skills. They also lose things easily. Patients of frontotemporal dementia typically have problems in behavior, personality and language when
assessed by neuropsychiatric battery of tests but have intact visual-spatial skills. The presenting features are in the form of behavioral abnormalities like anti-social conduct, poor personal grooming, thefts and vehicle accidents. Generally the diagnosis is not made early by non-specialists, even though it is one of the common causes of dementia.

CASE REPORT

A 72-year-old man presented with history of poor oral intake for two days, chills and rigors, without the presence of fever on daily temperature recording. As he was not able to give accurate history, his daughter was interviewed and it was found that the patient has had personality change for 2–3 years now. It was insidious in onset and progressively worsening. Daughter noticed that patient had problems with memory, particularly short term memory loss, with patient not being able to remember the task he was doing at a time. Patient was also found to have multiple, minor, motor vehicle accidents, but he was not able to recall what happened. Patient also frequently misplaced things. The patient could still recognize family members, and was able to drive out alone without getting lost. For the past one year, patient had a gradual onset of disinhibition. He did not have urinary incontinence, but he would urinate anywhere and anytime at home even in the presence of other family members. He also became more reserved, communicated poorly with family members and at times was found to be non-responsive to questions. Patient’s daughter also noticed that patient has peculiar behaviors on and off, such as sudden thought block and smiling to himself. He did not have visual hallucinations. The patient’s daughter observed that he had vesicular rash on his left thigh which was painful and not itchy. There was no history of previous head trauma, no fever, no movement disorders and no history of frank delusion or hallucination. There was no previous history of psychiatric disorder in the patient or family members. On examination, the patient was alert and conscious with GCS 15/15. However, he was not paying attention to his surroundings. He was afebrile, pulse rate was 83 beats per min, and blood pressure was 128/62 mmHg. A tender, erythematous vesicular lesion was found on left thigh. All cranial nerves were found to be intact. Speech was normal, but at times patient would become mute. His mini mental state examination (MMSE) score was 21/30, as assessed recently on follow up by one of the attending neurologist. He was found to have positive sucking, palmo-mental reflexes. Rest of central nervous system examination and other examinations for respiratory, cardiovascular, gastrointestinal and musculoskeletal systems were essentially normal.

Patient’s full blood count, renal functions, thyroid functions, liver functions and routine urine examinations were normal. He tested negative for HIV, Hepatitis B, Hepatitis C, venereal disease research laboratory test (VDRL), and treponema pallidum hemagglutination test (TPHA). His serum electrolytes were normal. Based on the history and clinical presentation, a diagnosis of dementia with Herpes simplex of left thigh was considered. Computed tomography scan of the brain showed atrophy of frontal and temporal areas of the brain (Figures 1–4).

Figure 1: Bifrontotemporal atrophy and prominent sylvian fissures. Frontal horns of ventricles are just visible.

Figure 2: Bifrontotemporal atrophy and prominent sylvian fissures. Frontal horns of ventricles are clearly visible.
DISCUSSION

Frontotemporal dementia is one of the causes of early-onset dementia (EOD) and has a 2.6% prevalence. Vascular dementia (24.5%), and Alzheimer disease (25.6%) are the most common causes, as noted in a population based study from Japan [1]. Frontotemporal dementia is a focal form of dementia in the presenium, yet remains poorly recognized [2]. It is frequently mistaken for Alzheimer’s disease or psychiatric diseases [3]. The age of onset of FTD is somewhat younger than other degenerative dementias, with a mean age of onset of about 58 years [4, 5]. It is now recognized as one of the three types of frontotemporal lobar degenerations (FTLD): frontotemporal dementia, semantic dementia and progressive nonfluent aphasia (PNFA) [5]. The prevalence of FTD is variable in different age ranges: 3.6/100000 at age 50–59 years, 9.4/100000 at age 60–69 years and 3.8/100000 at age 70–79 years [4]. The reported age of onset ranges from 33 to 80 years [4], with a male predominance in FTD and semantic dementia [5]: Frontotemporal dementia is the second most common cause of dementia in patients less than 65 years of age [5]. Our patient developed frontotemporal dementia of 72 years of age.

Macroscopic examination of the brain of a patient with FTLD typically shows symmetrical focal atrophy of the frontal lobes, temporal lobes. Microscopic examination of the cerebral cortex in most forms of FTLD, shows microvacuolar and neuronal loss. On staining for deposits in neurons, abnormal tau protein aggregates (46%) and ubiquitin-positive inclusions (20%) were found [6]. Our patient had frontal and temporal lobe atrophy on CT scan. The exact etiology and pathophysiology of frontotemporal dementia is incompletely understood but recent studies suggest that genetics plays an important role in disease causation [7]. This is supported by the fact that about 50% of patients with frontotemporal dementia (FTD) report a positive family history [7].

Patients with FTD display a heterogeneous clinical picture, which may include behavioral, cognitive, and motor manifestations [8]. They can be divided into two variants: the behavioral variant and the language variant (also referred as primary progressive aphasia) [8]. The behavioral variant, which was a likely variant in our patient is characterized by progressive personality changes, lack of insight, loss of social awareness, emotional blunting and loss of empathy [8]. The language variant on the other hand can be further divided into a well-defined clinic-pathological entity, semantic dementia (SD), and progressive nonfluent aphasia (PNFA) [8]. Progressive nonfluent aphasia is characterized by a progressive deficit in language, starting from anoma and agrammatism progressing to dysfluent spontaneous speech and eventually mutism [8]. Our patient had shown features of mutism, during his admission in the in-patient department and was also noted by the daughter of the patient. The vesicular rash on the thigh in our patient, was diagnosed to be due to Herpes simplex and was treated appropriately.
Frontotemporal dementia is largely made by clinical assessment, as in our case. Neuroimaging studies are equally important to provide supportive evidence for diagnosis as well as to exclude other structural diseases [8]. Multiple diagnostic criteria to aid in diagnosis (Table 1) have been devised and published [9]. Treatment mode available for frontotemporal dementia is very limited as there are no currently FDA-approved diseases modifying treatments [10]. Hence, off-label medication usage is frequent in the hope to cope with its devastating functional disability. Antidepressant and antipsychotic medications, (US FDA-approved treatments for Alzheimer’s disease) including cholinesterase inhibitors and memantine have been used [10]. Antidepressant and antipsychotic medications [11], have proven to give some benefits while cholinesterase inhibitors have limited efficacy [12]. The role of memantine in frontotemporal dementia is under further evaluation [10].

Table 1: Clinical and investigational diagnostic criteria for frontotemporal dementia

<table>
<thead>
<tr>
<th>Behavioural</th>
<th>Affective and speech symptoms</th>
<th>Physical signs and Investigational</th>
<th>Exclusions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insidious onset</td>
<td>Depression, anxiety</td>
<td>Pamplomental reflex</td>
<td>Abrupt onset</td>
</tr>
<tr>
<td>Lack of personal hygiene</td>
<td>Emotional unconcern/indifference</td>
<td>Sucking reflex</td>
<td>Head trauma related to onset</td>
</tr>
<tr>
<td>Early signs of disinhibition</td>
<td>Echolalia, progressive reduction in speech and mutism.</td>
<td>Glabellar tap</td>
<td>Cerebellar ataxia, corticobulbar and spinal deficits</td>
</tr>
<tr>
<td>Stereotyped behavior (dapping, dancing, singing etc)</td>
<td></td>
<td>Normal EEG</td>
<td>Brain imaging (post-central structural/functional deficits or multifocal infarcts on CT or MRI)</td>
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<tr>
<td>Loss of insight</td>
<td></td>
<td>Brain imaging findings</td>
<td></td>
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<tr>
<td>Utilization behavior — unrestrained exploration of objects in the environment</td>
<td></td>
<td>(frontal and temporal lobe atrophy)</td>
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</table>

CONCLUSION

Frontotemporal dementia is easy to diagnose and should be suspected in middle age and elderly persons presenting with gradual onset of personality changes, aggressive behavior, apathy and mutism. It is easily diagnosed by standard criteria and brain scan findings. Family history may be positive in some cases.

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES

An ectopic pheochromocytoma on cardiac metaiodobenzylguanidine scintigraphy in a patient with Takotsubo cardiomyopathy

Kenji Sadamatsu, Yuya Yoshidomi

ABSTRACT

Introduction: Pheochromocytoma is one of the diseases that is known to cause Takotsubo cardiomyopathy, however, because of its rare occurrence a diagnosis of pheochromocytoma may be overlooked in patients with Takotsubo cardiomyopathy. Case Report: We report a case of a patient with Takotsubo cardiomyopathy which was discovered when the patient visited her doctor for consultation for a breast mass. During the visit she developed transient left ventricular dysfunction. Computed tomography scan detected a paraortic mass. After recovery, she underwent simultaneous operations for the breast cancer and the paraaortic mass and developed an acute hypertensive crisis. The paraaortic mass was diagnosed as ectopic pheochromocytoma. On retrospective examination, metaiodobenzylguanidine scintigraphy, which was undertaken previously to assess suspected Takotsubo cardiomyopathy, showed an abnormal uptake in the paraaortic mass which was suspicious for pheochromocytoma. Conclusion: Pheochromocytoma can be detected as an incidental finding on metaiodobenzylguanidine cardiac scintigraphy, which is used to evaluate patients with Takotsubo cardiomyopathy or heart failure.

Keywords: Radioisotope, Computed tomography, Paraganglioma, Metaiodobenzylguanidine

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INTRODUCTION

Takotsubo cardiomyopathy has been widely recognized as a differential diagnosis for patients with chest pain. Several mechanisms, including microvascular dysfunction, have been proposed as a possible cause of this disorder. The abnormal uptake on iodine-123 labelled metaiodobenzylguanidine cardiac scintigraphy suggests that an excessive release of catecholamines has an important role in the pathophysiology of the disease [1, 2]. Pheochromocytoma is one of the diseases that is known to cause reversible left ventricular dysfunction, however, because of its rare occurrence its diagnosis may be missed in patients with Takotsubo cardiomyopathy.

CASE REPORT

A 61-year-old female who was visiting a doctor for consultation for the presence of a breast mass, suddenly developed headache and chest pain and therefore was transferred to a hospital. Although her electrocardiogram revealed no significant ST-T change, the troponin I level was 3.57 ng/mL. Coronary angiography was normal and left ventriculography demonstrated anterior hypokinesis.
postoperative chemotherapy for breast cancer. One year after the operation, CT scan did not demonstrate any evidence of recurrence of the pheochromocytoma or the breast cancer.

DISCUSSION

Cardiac scintigraphy with Iodine-123 labelled metaiodobenzylguanidine, a radiopharmaceutical agent that is taken up by adrenergic nerve endings, has been reported to be useful in patients with heart failure or left ventricular dysfunction. A reduced uptake ratio of the heart to mediastinum on the planar image has an independent prognostic value in patients with symptomatic heart failure [3]. In patients with Takotsubo cardiomyopathy, myocardial metaiodobenzylguanidine scintigraphy can reveal transient defects and a high washout ratio [1, 4], which suggests that there is an impaired activity of the sympathetic nervous system in such patients. On the other hand, metaiodobenzylguanidine scintigraphy has also been used for detecting pheochromocytoma inspite of the difference of labelling with Iodine-123 or iodine-131 [5], and has a high sensitivity of 77–100% and a specificity of 95–100% [6]. Diagnostic imaging is useful for both the diseases and the incidental finding of pheochromocytoma by metaiodobenzylguanidine cardiac scintigraphy is possible. However, the diagnosis of pheochromocytoma might be difficult without a suspicion [7], because the pheochromocytoma is considered a rare disease, and thus the entire findings of Iodine-123 labelled metaiodobenzylguanidine cardiac scintigraphy must be carefully reviewed in patients with heart failure and/or Takotsubo cardiomyopathy. In addition, the pheochromocytoma can be an etiology of heart failure and/or Takotsubo cardiomyopathy [8]. Metaiodobenzylguanidine scintigraphy might also serve as a single step assessment to predict the prognosis and to rule out the etiology for patients with heart failure and/or Takotsubo cardiomyopathy.

The recurrence of pheochromocytoma and Takotsubo cardiomyopathy is not rare and the surgical removal of a pheochromocytoma does not necessarily lead to a long-term cure of pheochromocytoma or hypertension, even in the case of a benign tumor. The recurrence rate of pheochromocytoma is reported to be 16%, and recurrence is more likely in patients with familial pheochromocytoma or familial paraganglioma, right adrenal tumors and extra-adrenal tumors [9]. The recurrence rate of Takotsubo cardiomyopathy has been reported to be 1.6–11.4% [10], and this type of cardiac dysfunction sometimes results in hemodynamic collapse, although the transient left ventricular dysfunction in our case was dependent on the attacks of the pheochromocytoma. Therefore, long-term monitoring is indicated in such patient for the recurrence of pheochromocytoma with Takotsubo cardiomyopathy.

Figure 1: Left ventriculography at the first admission at (A) end-systole and (B) end-diastole showed anterior hypokinesis (arrows) at the mid-ventricular level, (C) Abdominal computed tomography scan showed a 4.2x4.0x7.7 cm heterogeneously enhanced mass (arrow) in the left abdomen, (D) Iodine-123 metaiodobenzylguanidine planar imaging demonstrated an abnormal uptake in the left abdomen (arrow), which was accordant to the site of the paraaortic mass.
CONCLUSION

Pheochromocytoma can be detected as an incidental finding on metaiodobenzylguanidine cardiac scintigraphy done for evaluating patients with Takotsubo cardiomyopathy or heart failure.

Acknowledgements
The authors wish to thank Dr. Daisuke Mori for his evaluation of the pheochromocytoma specimen.

Author Contributions
Kenji Sadamatsu – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Yuya Yoshidomi – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES

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Hirayama disease: Role of diffusion tensor imaging

Dinesh Sharma, Shruti Thakur, Chidanand Chavan, Anupam Jhobta

CASE REPORT

A 28-year-old male patient presented with progressive weakness of both forearms and hands along with clawing of hands for past eight years. Right side was affected more than the left. There was no history of similar illness in the family. There was no past history of any prolonged illness in childhood. There was no history of sensory, pyramidal or bladder dysfunction. There was atrophy of muscles of both forearms and hands. Muscle power was normal in both upper limbs. Nerve conduction studies showed reduced amplitude in bilateral median and ulnar nerves. Tendon reflexes were normal. Magnetic resonance imaging (MRI) examination was performed in non-flexion and flexed position. Subsequently, diffusion tensor imaging (DTI) was performed in non-flexion, which very clearly showed the cord atrophy at C7–T1 level (Figure 1). This cord atrophy was not appreciated on MRI flexion and non-flexion studies. The diagnosis of Hirayama disease (HD) was thus established on DTI.

Dinesh Sharma¹, Shruti Thakur², Chidanand Chavan³, Anupam Jhobta⁴

Affiliations: ¹MD, (Radiodiagnosis), Assistant Professor, Department of Radiodiagnosis and Imaging, Indira Gandhi Medical College Shimla, India; ²MD, (Radiodiagnosis), Senior Resident, Department of Radiodiagnosis and Imaging, Indira Gandhi Medical College Shimla, India; ³MD, Student (Radiodiagnosis), Junior Resident, Department of Radiodiagnosis and Imaging, Indira Gandhi Medical College Shimla, India; ⁴MD, (Radiodiagnosis), Associate Professor, Department of Radiodiagnosis and Imaging, Indira Gandhi Medical College Shimla, India.

Corresponding Author: Dinesh Sharma, Assistant Professor, Department of Radiodiagnosis and Imaging, Indira Gandhi Medical College Shimla, India 171001; Ph: +919418499909; Fax: +911772658339; Email: dineshss108@gmail.com

Received: 30 October 2012
Accepted: 17 November 2012
Published: 01 March 2013

Figure 1: Diffusion tensor magnetic resonance image acquired in non-flexion shows atrophy of spinal cord at C7–T1 level (arrows).

DISCUSSION

Hirayama disease (HD) is a non-progressive, juvenile, spinal muscular atrophy of distal upper limbs and is a kind of cervical myelopathy related to flexion movements of the neck. Imbalance in growth of
vertebrae and dura mater causes a loss of normal dural slack in extension. As a result of this, the tight dural canal causes compression of the spinal cord [1]. It is suggested that HD might be due to microvascular changes following chronic trauma to spinal cord during flexion and extension of the neck [2]. On non-flexion MRI studies, asymmetric cord atrophy especially at the lower cervical level is highly suggestive of HD [3]. Furthermore, the magnetization and diffusion MRI histograms of the cervical cord suggest that the cord damage in HD extends beyond that seen on routine MRI scans [4]. In our opinion, this atrophy of the spinal cord is best demonstrated on diffusion tensor imaging. This makes diffusion tensor imaging a valuable tool in diagnosing Hirayama disease without subjecting the patient to a flexion study of the cervical spine.

CONCLUSION

Diffusion tensor imaging is simple, fast and accurate method for diagnosing Hirayama disease without subjecting the patient to flexion magnetic resonance imaging study.

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Acknowledgements
Dr Ashwani Tomar, Dr Vijay Thakur, Dr R.G. Sood, Department of Radio diagnosis and Imaging, Indira Gandhi Medical College Shimla, India –171001.

Author Contributions
Dinesh Sharma – Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Shruti Thakur – Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Chidanand Chavan – Acquisition of data, Drafting the article, Final approval of the version to be published
Anupam Jhobta – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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REFERENCES
Acute renal failure caused by tumor-induced hypercalcemia successfully treated by denosumab

Kouichi Kuninaka, Hiromu Tengan, Tadashi Nishimaki

To the Editors,

Denosumab, a human monoclonal antibody that binds to and neutralizes the receptor activator of nuclear factor-κ ligand (RANKL), has recently become available for the prevention of skeletal-related events (SRE) of bone metastases from breast cancer. In a recent study, denosumab was shown to be more effective than zoledronic acid in delaying SRE [1], and it has been shown that hypocalcemia occurred more frequently and renal failure less frequently, in patients treated with denosumab as compared to those treated with zoledronic acid [1-3]. In light of these studies, we used denosumab to successfully treat a patient presenting with acute renal failure caused by a tumor-related hypercalcemia arising from breast cancer bone metastases.

A 53-year-old woman was admitted to the orthopedics ward due to walking difficulties. Investigations revealed compression of the spine by metastatic carcinoma of the thoracic vertebrae. An emergency decompression surgery was performed. Pathological examination of the resected specimen revealed metastatic carcinoma of probable breast cancer origin that was immunohistochemically positive for estrogen receptor, negative for progesterone receptor, and 2+ for HER2 receptor (additional fluorescence in situ hybridization revealed significant amplification of HER2/neu). The patient had undergone surgery for left breast cancer 13 years ago in our hospital and had shown no evidence of recurrence at her last visit to the outpatient clinic three years ago. A positron emission tomography (PET) scan after the decompression surgery revealed multiple bone metastases (in the thoracic and lumbar vertebrae, sternum, humeri, scapula, ribs, pelvis, and femur). Lung metastasis and serum calcium level started to increase 29 days after admission. Tumor-related hypercalcemia was strongly suspected and the patient was treated with elcatonin (80 U/day), normal saline (1,000 ml/day) and furosemide (40 mg/day) for six days from day 29, but the serum calcium level continued to increase and renal dysfunction developed (Figure 1). The patient developed severe appetite loss and renal dysfunction with creatinin clearance of 26 ml/min. With these findings a diagnosis of acute renal failure due to tumor related hypercalcemia was made. Usually, we use zoledronic acid to treat tumor-related hypercalcemia, but it could not be used in this case because of renal dysfunction. As an alternative, on day 49, we administered single dose of denosumab (120 mg) to treat the acute renal failure caused by tumor-related hypercalcemia. The serum calcium level decreased to within normal limits after only five days, and renal functions improved and completely recovered after a month (Figure 1). The patient is now under treatment with exemestane and trastuzumab. Denosumab is still being administered every four weeks and the serum calcium level and renal function are stable owing to a daily intake of 600 mg calcium and 0.25 μg vitamin D, respectively.

Enhanced bone resorption is the primary cause of tumor-induced hypercalcemia. The release of tumor-derived mediators induces this increase in osteoclast-mediated resorption. The interaction between osteoclasts and cancer cells is mainly mediated by parathyroid hormone-related protein (PTHrP), that activates osteoblasts to produce RANKL which stimulates osteoclast differentiation [4]. Denosumab, human RANKL antibody inhibits this vicious cycle. Moreover, denosumab binding to RANKL is metabolized by peptides and cleared by the reticuloendothelial system and probably does not have a nephrotoxic effects.
Figure 1: Changes in serum calcium (albumin adjusted, black line) and creatinine (grey line) levels in response to denosumab treatment. The dotted lines indicate the normal range for serum calcium and the solid line indicates the upper limit for serum creatinine.


**Guarantor**
The corresponding author is the guarantor of submission.

**Conflict of Interest**
Authors declare no conflict of interest.

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**REFERENCES**


**Author Contributions**
Koichi Kuninaka – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Hiromu Tengan – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Tadashi Nishimaki – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published