A case study of paraneoplastic cauda equina syndrome caused by a gastric adenocarcinoma

Debbie Hunt, Shomari Zack-Williams, Anita Hargreaves, David Monk

ABSTRACT

Introduction: Cauda equina syndrome results from dysfunction of multiple sacral and lumbar nerve roots in the lumbar vertebral canal, leading to impairment of bladder, bowel, or sexual function, and perianal or “saddle” numbness. The most common cause of cauda equina syndrome is disc herniation resulting in compression at L4/5 and L5/S1. However, we will discuss the case of cauda equine syndrome with a paraneoplastic cause. There are only a handful of cases in literature of paraneoplastic cauda equine syndrome, and none specifically as a result of gastric adenocarcinoma. Paraneoplastic neurological syndromes (of which paraneoplastic cauda equine syndrome is one) are described as remote effects of cancer on the neurological system. They are rare, affecting less than 1/10,000 patients with cancer. In this case, the cauda equina was the target for an autoimmune response directed against antigens common to both the cancer and the nervous system.

Case Report: A 71-year-old female was admitted with a two-month history of lumbar back pain, radiating down her thigh, progressive weakness of both legs, numbness of the sacral area, urinary incontinence and 6.4 kilogram unintentional weight loss within 2 months. Abdominal radiograph, breast examination, lumbar puncture, and autoantibodies screens were all negative. Abdominal and pelvic CT, spinal MRI, radioisotope scan and abdominal USS still did not demonstrate any malignant process. One month after admission, the patient deteriorated with sudden abdominal peritonism, tachycardia and hypothermia. An urgent CT was performed, which demonstrated a gastric perforation. A laparotomy was undertaken which demonstrated a 4-cm gastric perforation. Biopsies were taken and the histology subsequently demonstrated a high grade, poorly differentiated adenocarcinoma. From this diagnosis, it was ascertained that she had been suffering from secondary paraneoplastic neuropathy, caused by the gastric adenocarcinoma in the body of the stomach. This specific case has not been reported in literature within the last 10 years.

Conclusion: In conclusion, an unusual presentation of acute and progressive neuropathy without obvious spinal/ cranial aetiology and associated cachexia should prompt thorough investigation to exclude a neoplastic process, as paraneoplastic syndromes may be the first sign of malignancy.

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CASE REPORT

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ABSTRACT

Introduction: Cauda equina syndrome results from dysfunction of multiple sacral and lumbar nerve roots in the lumbar vertebral canal, leading to impairment of bladder, bowel, or sexual function, and perianal or “saddle” numbness. The most common cause of cauda equina syndrome is disc herniation resulting in compression at L4/5 and L5/S1. However, we will discuss the case of cauda equine syndrome with a paraneoplastic cause. There are only a handful of cases in literature of paraneoplastic cauda equine syndrome, and none specifically as a result of gastric adenocarcinoma. Paraneoplastic neurological syndromes (of which paraneoplastic cauda equine syndrome is one) are described as remote effects of cancer on the neurological system. They are rare, affecting less than 1/10,000 patients with cancer. In this case, the cauda equina was the target for an autoimmune response directed against antigens common to both the cancer and the nervous system. **Case Report:** A 71-year-old female was admitted with a two-month history of lumbar back pain, radiating down her thigh, progressive weakness of both legs, numbness of the sacral area, urinary incontinence and 6.4 kilogram unintentional weight loss within 2 months. Abdominal radiograph, breast examination, lumbar puncture, and autoantibodies screens

were all negative. Abdominal and pelvic CT, spinal MRI, radioisotope scan and abdominal USS still did not demonstrate any malignant process. One month after admission, the patient deteriorated with sudden abdominal peritonism, tachycardia and hypothermia. An urgent CT was performed, which demonstrated a gastric perforation. A laparotomy was undertaken which demonstrated a 4-cm gastric perforation. Biopsies were taken and the histology subsequently demonstrated a high grade, poorly differentiated adenocarcinoma. From this diagnosis, it was ascertained that she had been suffering from secondary paraneoplastic neuropathy, caused by the gastric adenocarcinoma in the body of the stomach. This specific case has not been reported in literature within the last 10 years. **Conclusion:** In conclusion, an unusual presentation of acute and progressive neuropathy without obvious spinal/ cranial aetiology and associated cachexia should prompt thorough investigation to exclude a neoplastic process, as paraneoplastic syndromes may be the first sign of malignancy.

Keywords: Gastric adenocarcinoma, Neurological syndromes, Paraneoplastic cauda equina syndrome, Sacral and lumbar nerve

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INTRODUCTION

Cauda equina syndrome is described as dysfunction of multiple sacral and lumbar nerve roots in the lumbar vertebral canal, which can result in a combination of clinical features. However, the term cauda equina syndrome is used only when these include impairment of bladder, bowel, or sexual function, and perianal or “saddle” numbness.

The most common cause of cauda equina syndrome is compression at L4/5 and L5/S1 from large central lumbar disc herniation. Less common causes include spinal fractures or subluxation, spinal neoplasms of metastatic or primary origin and infective causes.

In this report, we will discuss the case of cauda equina syndrome with a paraneoplastic cause. There are only a few cases in literature of paraneoplastic cauda equine syndrome, and none specifically as a result of gastric adenocarcinoma.

CASE REPORT

A 71-year-old, previously healthy woman, was admitted to the orthopedic ward with a two month history of lumbar back pain, right groin pain radiating down the front of her thigh and progressive weakness of both legs. There was also a seven-day history of numbness of the sacral area and urinary incontinence. In addition, it was also noted that the patient had undergone a 6.4-kg unintentional weight loss within two months, with a current BMI of 16.8.

On examination, the abdomen was soft with generalized tenderness in the lower abdomen and also a localized tenderness in the lumbosacral area. There was no sensory deficit of the legs, normal tone, normal co-ordination and no pain at this stage. However, there was reduced power in the right hip flexors. There was obvious cachexia with marked muscle wasting. Spinal examination was normal. On rectal examination, there was found to be poor anal tone and dubious saddle sensation. These neurological signs fluctuated throughout the course of the patient's stay.

At this stage, an MRI scan was taken of lumbar spine, showed multilevel degeneration, with disc disease and facet joint osteoarthritis. However, it was thought that the degree of narrowing did not appear severe enough to cause the present symptoms. The bladder scan confirmed acute urinary retention, showing a residual volume of 999 ml, and the patient was subsequently catheterized (which stayed in situ for the duration of the patient's stay). An abdominal radiograph also confirmed significant fecal loading. The lumbar back pain worsened over the next week requiring increasing amounts of oral morphine. The patient's weight did not improve either, despite a high protein, high calorie diet. She also experienced worsening abdominal pain and constant nausea.

Over the next three weeks, the patient also suffered

from a number of falls, and progressive loss of sensation in her feet. At this stage the patient was also incontinent of feces, there was a loss of reflexes in both legs and the weight loss had progressed to 13 kilograms, even with enteral feeding. There were also increasingly frequent episodes of confusion. This was thought to be due to a UTI as her urine dipstick was positive for ketones, leucocytes, nitrites, protein and blood and cultured positive for *E.coli*, and she was therefore started on appropriate antibiotics.

Breast examination and mammography did not demonstrate any pathology. Blood tests demonstrated an increased CRP (284 mg/L), increased LDH (717 U/L), increased ferritin (761.4 ng/ml), neutropenia ($1.5 \times 10^9/L$), deranged urea and electrolytes (urea 10.5 mmol/L, Na 124, K 3.2 mmol/L) and normal calcium (2.4 mmol/L), phosphate (1.1mmol/L) and tumor markers (including Ca19-9, Ca15-3 and Ca125). A lumbar puncture showed clear and colorless cerebrospinal fluid with low glucose, high protein and no organisms. Autoantibodies and ANCA were negative. Electrophoresis did not demonstrate any monoclonal bands. Abdominal and pelvic computed tomography, spinal MRI scan (Figure 1), and abdominal USS still did not demonstrate any malignant process. The isotope scan demonstrated increased uptake from the right femoral neck only (there was no known pathology in this region).

One month after admission, the patient deteriorated with sudden severe right upper quadrant and right flank pain, abdominal peritonism, tachycardia and hypothermia. Bowel sounds were no longer present. It was noted that the NG tube feed had been increased that day and the patient had been taking non steroidal anti-inflammatories. There was no previous history of gastric pathology.

An urgent CT scan was performed, which demonstrated a gastric perforation (Figure 2). A laparotomy was undertaken which demonstrated a 4-cm gastric perforation and extensive contamination of the abdomen. Biopsies were taken and the perforation was closed with the Graham Patch. (Radical treatment was initiated as it is known that perforation can occur even in the early stages and seems not to be a negative prognostic factor itself for adenocarcinoma [1]. Drains were inserted in each paracolic gutter. The histology subsequently demonstrated a high grade, poorly differentiated adenocarcinoma.

Postoperatively the patient was transferred to the intensive care unit where she was given both cardiovascular and respiratory support, intravenous tazocin and metronidazole, and total parenteral nutrition feed. She was also maintained on the Hong Kong regime which involves a COX-2 inhibitor and a PPI. While on ITU, the patient suffered from episodes of significant tachycardia and low hemoglobin (7.4 g/L), for which she required two transfusions. The CRP (276 mg/L) and WCC ($17.2 \times 10^9/L$) remained high despite her intravenous antibiotics.

Subsequently, due to the patient's general health, it was decided that the patient was for palliation of symptoms only and she was put on the Liverpool Care Pathway.

The final diagnosis given was that of secondary paraneoplastic neuropathy, caused by a gastric

adenocarcinoma in the body of the stomach, which, in this case, caused a cauda equina syndrome. This specific case has not been reported in literature.

The diagnosis of paraneoplastic cauda equina was a presumptive diagnosis based on the extensive negative findings and final histology. There was not enough time preoperatively (between the finding of the adenocarcinoma on computed tomography and the emergency laparotomy) to perform onconeural antibodies. Furthermore, postoperatively, the patient deteriorated very quickly, giving little time for either, assessment of the effects of treatment of the gastric adenocarcinoma on the cauda equina syndrome (a condition to be satisfied for a conclusive diagnosis of paraneoplastic syndrome to be made), or for testing of onconeural antibodies (another condition).

Furthermore, computed tomography, magnetic resonance imaging and isotope bone scanning showed neither evidence of an abnormality of the spine (for example any evidence of spinal metastasis/sufficient disc protrusion to produce the symptoms) or abnormality of the cauda equina itself. This led us to the conclusion that the symptoms were caused by a paraneoplastic phenomenon as all other plausible causes had been excluded.

DISCUSSION

Paraneoplastic neurological syndromes, of which cauda equina syndrome in one, are defined as the remote effects of cancer that are not caused by the tumor and its metastasis, or by infection, ischemia or metabolic disruptions. These cases are rare, affecting less than 1/10,000 patients with cancer [2]. In 2007, criteria were developed for diagnosing PNS. In short, the criteria include

- (i) a classical neurological syndrome is observed (one of which is sensory neuropathy);
- (ii) the condition improves after cancer treatment;
- (iii) or if non-classical neurological syndrome, that onconeural antibodies are present [3].

There have been cases of PNS reported from metastatic breast cancer [4–5] prostate cancer, small cell lung cancer [6] and lymphoma and it is thought that these tumors secrete a protein, which is taken up by the nerves, causing them to hypertrophy due to osmotic retention of water, causing compression leading to the neurological symptoms described [7]. There are also cases described in literature where the cauda equina is a target for an autoimmune response directed against antigens common to both the cancer and the nervous system, designated as onconeural antigens [2]. Therefore, the best way to diagnose these cases is to identify one of the anti-onconeural protein antibodies. These antibodies can guide the search for the primary malignancy (when often these are not clinically overt). This is important as the best way to stabilize the paraneoplastic syndrome

Figure 1: Magnetic resonance imaging showing no intrinsic cervical/thoracic cord abnormality. There are, however, spondylotic changes in the mid-cervical spine resulting in spinal canal narrowing – although not deemed to be extensive enough to cause the symptoms discussed. There is also a small central disc protrusion at T5/6.

Figure 2: Computed tomography image showing marked ascites, free intraperitoneal gas as a result of a large perforated gastric ulcer along the anterior wall of the stomach body.

is to treat the primary cancer. However, other papers refute this, claiming that less than 50% of patients with paraneoplastic syndromes have paraneoplastic antibodies [3].

Similar cases of paraneoplastic neuropathy affecting the cauda equina describe progressive distal limb paresthesia, weakness, heaviness and gait difficulty. There was also loss of vibration and proprioception. Nerve conduction studies showed evidence of axonal peripheral neuropathy and somatosensory evoked potential studies showed impaired conduction [7, 8].

Similar to our case, these patients described symptoms which progressed over many months to years. In a case report of a 45-year-old woman with a metastatic cauda equina tumour from breast cancer [5], the patient presented four years after she had undergone resection, without spinal column or brain metastasis. This particular case however, in contrast to our case with generalized hypertrophy of the cauda equina, demonstrated a well enhanced intradural extramedullary mass. In contrast to our case however, these patients showed diffuse abnormal thickening and enhancement of the cauda equine nerve roots on MRI scan [8], whereas in our case, there was no abnormality of the cauda equina on any imaging modality.

Additionally, while there have been no reports of gastric cancers causing a cauda equine syndrome specifically, there have been cases described in literature of other paraneoplastic neurological manifestations of gastric carcinoma, including one case of Stage 1b gastric cancer associated with spontaneous muscle atrophy of both hands. The cause of the atrophy was not apparent with neurologic diagnostic modalities, and therefore the case was assumed to be due to a paraneoplastic manifestation [9]. There is also a case report of systemic polyarteritis nodosa leading to the discovery of an asymptomatic, surgically curable gastric adenocarcinoma, and a diagnosis of paraneoplastic systemic angiitis was given [10].

CONCLUSION

In conclusion, an unusual presentation of acute and progressive neuropathy without obvious spinal/cranial aetiology and associated cachexia should prompt thorough investigation to exclude a neoplastic process, as paraneoplastic syndromes may be the first sign of malignancy.

Author Contributions

Debbie Hunt – 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
Shomari Zack-Williams – Analysis and interpretation

of data, Revising it critically for important intellectual content, Final approval of the version to be published
Anita Hargreaves – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
David Monk – 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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