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Emmi Khoo, Dharmenaan Palamuthusingam, Clyson Mutatiri

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Case Report: We report an unusual case of a 74-year-old male patient of Dutch descent presented with intermittent watery diarrhea, recurrent ascites and bilateral lower limb lymphedema.

Conclusion: Symptoms of the patient at initial presentation were non-specific. Bilateral lower limb lymphedema has not been previously identified in literature as a presenting symptom of this disease. Clinicians need to have a higher index of suspicion for the diagnosis of SEP in those that present with bilateral lower limb lymphedema and non-specific abdominal symptoms. The etiology of SEP in this patient remains unclear, which is consistent with most of the cases reported in literature to date.

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CASE REPORT

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Keywords: Ascites, Bowel obstruction, Lymphedema, Sclerosing encapsulating peritonitis

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INTRODUCTION

Sclerosing encapsulating peritonitis (SEP) is a rare condition with varying prevalence around the world. An Australian study identified 54 cases in 14 years, indicating prevalence of 0.7% [1]. This compares with a prevalence of 3.3% described in a UK series [2].

Underlying cause of SEP is believed to be multifactorial [3]. It can occur secondary to several conditions like prior episodes of severe peritonitis, use of beta-blockers, history of peritoneal dialysis, autoimmune disease, intra-abdominal malignancies, exposure to chemicals such as silicosis or asbestosis, endometriotic cyst, uterine leiomyomas, ovarian tumors, sarcoidosis, tuberculosis or it can be idiopathic [4]. There is no significant association between SEP, age and gender [5]. However, notably, idiopathic types are found more commonly among young adolescent female in tropical and sub-tropical areas [4].

CASE REPORT

A 74-year-old male lived his life in Australia and very rarely traveled overseas. He has never smoked tobacco or consumed alcohol. He has a known history of transient ischemic attack four years ago, left knee osteoarthritis,

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depression, and hypercholesterolemia. His medications include escitalopram 20 mg mane, lorazepam 1 mg nocte and rosuvastatin 5 mg mane. Examination revealed normal cardiovascular and respiratory system and his abdomen was soft, with no masses but was positive for shifting dullness. He also had bilateral pitting edema up to his mid-thighs.

Initial biochemistry, full blood count and liver function

tests were unremarkable (Table 1). An echocardiogram showed normal right and left ventricular function and normal valves. An ultrasound (USS) of the abdomen confirmed more than 1 L of ascites but normal liver and spleen dimensions with no ultrasonographic evidence of portal hypertension. The USS of his lower limbs did not demonstrate deep venous thrombosis. Computed tomography (CT) scan of his abdomen showed mild

Table 1: Initial investigations performed

Investigations	Value	Units	Ref. Range
Hemoglobin	131	g/L	120–180
White cell count	4.7	x 10 ⁹ /L	3.5–11.0
Platelet	196	x 10 ⁹ /L	140–400
Creatinine	90	mmol/L	64–108
GFR (estimated)	72	mL/min/1.73m ²	>60
Protein	63	g/L	60–83
Albumin	42	g/L	35–50
Alkaline Phosphatase	217	U/L	56–119
Gamma-GT	29	U/L	<55
Alanine Transaminase	10	U/L	<45
Aspartate Transaminase	14	U/L	<35
Lactate Dehydrogenase	170	U/L	150–280
Calcium (Alb. Corr.)	2.30	mmol/L	2.15–2.60
Phosphate	1.08	mmol/L	0.81–1.45
Magnesium	1.04	mmol/L	0.70–1.10
INR	1.0		0.9–1.2
CRP	2.3	mg/L	<5.0
CA 19.9	<5.0	kU/L	<35
Carcinoembryonic Ag	<1.0	ug/L	<5.0
ePSA	0.14	ug/L	<6.5
Ferritin	172	ug/L	30–300
Transferrin	1.8	g/L	1.6–3.4
Vitamin B12	172	pmol/L	133–680
Serum Folate	29.4	nmol/L	>7.0
25-Hydroxy-Vitamin D	66	nmol/L	50–150
AST (Red cell)	6.2	U/g Hb	3.0–7.0
Ceruloplasmin	1.78	mmol/L	1.33–2.22
Serum Zinc	12	mmol/L	8–18
C3	1.51	g/L	0.90–1.80
C4	0.52	g/L	0.10–0.40
Rheum. Factor (Neph)	<20	IU/ml	<30
Anti-CCP	0	U/ml	<6
Antinuclear Antibody	Negative	Titre	
Extractable Nuclear Ag	Negative		
Anti dsDNA	4	IU/ml	<7
Monoclonal Protein	Not Detected		
IgG	4.8	g/L	7.0–16.0
IgA	0.3	g/L	1.0–4.0
IgM	0.2	g/L	0.4–2.3
Kappa FLC (N Latex)	10	mg/L	7–22
Lambda FLC (N Latex)	16	mg/L	8–27
K/L Ratio (N Latex)	0.62		0.31–1.56
Urine Bence Jones Protein	Not Detected		
Urine Creatinine: Protein	<18	g/mol creat	<15
Urine Cytology	No malignant cells found		
Stool Alpha-1-Antitrypsin	0.5	mg/g dry wt	<1.5
Stool Occult Blood	Negative		

heterogeneity of the greater omentum in the upper abdomen, some circumferential thickening of the sigmoid, and few sclerotic foci in axial skeleton.

A abdominal paracentesis performed was cloudy in appearance but cytology and biochemistry were unremarkable. The surgeons performed a sigmoidoscopy and subsequent biopsies revealed normal large intestinal architecture.

A diagnosis remained elusive and the patient was treated symptomatically with bumetanide 1 mg mane but there was no clinical improvement.

Ultimately, a radionuclide lymphoscintigraphy confirmed bilateral distal ‘dermal backflow’, consistent with bilateral lymphedema. A provisional diagnosis of lymphangitis was made based on these findings he was commenced on prednisolone 50 mg daily.

After four weeks on steroid treatment, his edema had reduced significantly and his bowel symptoms improved.

Despite improved symptomatology, he was revisited by the surgeon in clinic, who continued to be suspicious for intra-abdominal malignancy and insisted on diagnostic laparoscopy being performed. At the time of the operation, the small bowel looked normal with white plaque-like patches in some areas. The surgeon converted to an exploratory laparotomy via a midline incision and noted the cecum feeling and appearing fibrotic and having some whitish areas resembling scarring in pelvic peritoneum. The sigmoid colon was adherent to pelvic sidewall, and the omentum was shrunken with obliteration of lesser sac. Several peritoneal biopsies were taken.

The histology of peritoneal biopsy revealed tissues containing bland spindle cells separated by collagen, some of which is thick and eosinophilic; with rare mitotic figure noted but no atypical figures were identified. Omental tissue biopsies showed thin submesothelial zone of collagenous connective tissue containing cytologically bland spindle cells, similar to those seen in the peritoneal biopsy. These histological features were consistent with sclerosing peritonitis. However, at this point in time the diagnosis SEP was overlooked for multiple reasons. Firstly, the experienced surgeon did not feel that at the time of the operation, the macroscopic appearance of the abdomen was not consistent with the typically described ‘cocoon-like’ appearance. Secondly, the patients’ presentation was not a characteristic manifestation of SEP that had been previously documented in literature.

Unfortunately, the patient developed acute bowel obstruction and died from sepsis eight months later.

DISCUSSION

Underlying cause of SEP is believed to be multifactorial [3]. It can occur secondary to several conditions like prior episodes of severe peritonitis, use of beta-blockers, history of peritoneal dialysis, autoimmune disease, intra-abdominal malignancies, exposure to chemicals such as silicosis or asbestosis, endometriotic

cyst, uterine leiomyomas, ovarian tumors, sarcoidosis, or tuberculosis [4]. Or it can be idiopathic. There is no significant association between SEP, age and gender [5]. However, notably, idiopathic types are found more commonly among young adolescent female in tropical and subtropical areas [4].

Common clinical features at presentation of SEP are abdominal pain, nausea, vomiting, loss of appetite, constipation and weight loss [5]. Other than symptoms of bowel obstruction, SEP may present with features of fluid overload like ascites and edema. In those undergoing PD this is attributable to the progressive loss of ultrafiltration. This is a result of peritoneal scarring and adhesions by fibrocollagenous membrane which leads to the loss of surface area for ultrafiltration [6]. Repeated episodes of peritonitis are believed to accelerate the process [7].

Preoperative diagnosis is non-specific. Imaging techniques are helpful but not diagnostic. Plain abdominal X-ray may show signs of bowel obstruction [8, 9]. Small bowel intestine loops congregated in a single area in the peritoneal cavity, encased by a soft tissue-density mantle may be seen on abdominal CT scans [10]. Other features like signs of obstruction, agglutination and fixation of intestinal loops, mural thickening, peritoneal thickening or calcification and reactive adenopathy may also be present [10]. Ultrasonography may illustrate ineffective peristaltic contractions, dilated fixed intestinal segments matted together and tethered posteriorly, intraperitoneal echogenic strands, trilaminar appearance of the bowel wall, and loculated free fluid [9, 11, 12]. The “cauliflower sign” is the conglomeration of bowel loops that appear to adhere to each other surrounded by a sac-like structure. This along with delayed transit of contrast medium are characteristics of SEP and can be found on contrast study including barium X-ray and contrast CT scan [8].

Ultimately, the diagnosis of abdominal cocoon is based on intraoperative and histopathology findings. During surgery, encapsulation of entire or partial intestine in thick white fibrous “cocoon-like” membrane is seen; and histology of membrane reveals proliferation of fibrocytes and enrichment of collagen fiber, with non-specific inflammatory reaction and vascular proliferation [8].

Surgical adhesiolysis is the gold standard in treatment of patients with recurrent bowel obstruction secondary to SEP [8, 11]. Other alternate treatment options for milder cases include TPN and nasogastric decompression [11].

Many reports in literature describe the beneficial effects of immunosuppressive agents on the progression of SEP [12–14]. Patients with SEP have been successfully treated with corticosteroids alone or in combination with azathioprine [11, 14, 15]. Examples of other immunosuppressants like colchicine, tamoxifen, renin- angiotensin inhibitors, phosphatidylcholine and antifibrotic agents have also been trialed in management of mild disease and outcomes were inconclusive [11, 12]. A study in Japan showed that two-year survival among patients treated with steroids was 73%; while among patients not receiving such treatment, the two-

year survival was only 48% [11]. It was proposed that immunosuppressive therapy could slow the progression of SEP by reducing the production of inflammatory mediators, which promote fibrinogenesis; hence, formation of fibrous capsule [12]. Due to the sporadic nature and low incidence of the disease, there is a lack of literature to conclude on the significance of long-term immunosuppressive therapy and treatment durations have varied between 3–4 months to life-long treatment [11, 14, 15].

This patient's symptoms at initial presentation were non-specific. His symptoms centered mainly on bilateral lower limb non-pitting lymphedema with ascites and some weight loss, with no indication of bowel obstruction. In a case series by Ping et al., all five of their patients experienced symptoms between 2 weeks to 10 years, before a diagnosis was confirmed. Those symptoms included recurrent episodes of intestinal obstruction, colicky abdominal pain, non-bilious vomiting, abdominal distension and constipation [16]. In our case, underlying malignancy had been considered high on the list of differentials. In retrospect, there were some initial features which were suggestive of SEP but this diagnosis had not been entertained preoperatively. His first episode of acute bowel obstruction occurred 10 months after this.

The aetiology of SEP in this gentleman remains unclear, which is consistent with most of the cases reported in literature to date.

CONCLUSION

This case report highlights that clinicians need to have a higher index of suspicion for the diagnosis of sclerosing encapsulating peritonitis (SEP) in those that present with bilateral lower limb lymphedema and non-specific abdominal symptoms.

Author Contributions

Emmi Khoo – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published

Dharmenaan Palamuthusingam – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published

Clyson Mutatiri – Substantial contributions to conception and design, Acquisition 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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