Images – Sclerosing mesenteritis presenting with unilateral hydro-ureteronephrosis

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Introduction

Sclerosing mesenteritis (SM) is a rare benign condition that can have many different presentations, including abdominal pain, gastrointestinal symptoms, weight loss, or fever.[1] The pathogenesis of this disease is not well-understood, but it may share a common etiology with other idiopathic primary inflammatory and fibrotic processes, such as mesenteric lipodystrophy (adipocyte necrosis) and mesenteric panniculitis (chronic inflammatory state).[1] The reported prevalence is 0.6–2.5%, with a male to female ratio of 2 :1.[1-4] The non-specific presentation can make the clinical diagnosis difficult. Abdominal exam can identify a mass in less than 50% of cases, while C-reactive protein (CRP) and erythrocytes sedimentation can be a way to follow its response to treatment.[5,6] Abdominal contrasted enhanced computed tomography (CT) scan is the most sensitive imaging modality with two specific signs: "fat ring sign" and "tumor pseudocapsule."[4,7,8] To complete the diagnosis, a pathological examination is often required. This case report highlights how the non-specific presentation of SM can obscure the diagnosis.

Clinical history

A 48-year-old male with no prior medical history presented to the emergency department with back pain complaints and low fever. He was an active smoker with a 30 pack-years history. He did not have any urinary tract symptom and the review of systems was negative. On physical examination, a tender, non-pulsatile abdominal mass was appreciated. Blood work showed a normal creatinine of 94 (estimated glomerular filtration rate [eGFR] 82) and a CRP of 193. Urine analysis was normal and blood cultures negative. Ultrasound revealed severe left hydroureteronephrosis. A contrast-enhanced abdominal CT scan demonstrated a 9.8 x 10.6 cm pelvic mass with a thin membrane and probable liquid content (17 HU) causing severe left hydroureteronephrosis (Fig. 1). A left nephrostomy was placed and intravenous piperacilline/tazobactam administered. The patient was admitted for further investigations.

Cystoscopy and an antegrade pyelogram showed no communication with the urinary system. A magnetic resonance imaging (MRI) scan demonstrated no communication with seminal vesicles or the intestinal tract. This was sampled by needle aspiration; biochemistry results showed serum levels of creatinine and microbiology cultures grew *Propionibacterium acnes*. The patient was treated with a total 14 days of antibiotics. The patient's fever subsided, but due to persistent low back pain, it was decided to proceed with elective surgery to attempt removal.

In the operating room, cystoscopy was normal and a retrograde pyelogram demonstrated a lateralized tortuous left ureter. A left ureteric stent was therefore installed. Through a lower midline abdominal incision, tissues were found to be densely adherent (Fig. 2). At one point, the mass was perforated, draining 200–300 mL of clear liquid. Following irrigation, we removed the inflammatory membrane. Three frozen section analyses identified only inflammatory tissue. The liquid was sent for cytology and microbiology cultures.

The surgical pathology identified the diagnosis of SM. Also, the fluid culture was positive for *Staphylococcus aureus* and the patient was subsequently prescribed three additional weeks of first-generation cephalosporin therapy.

The ureteric stent was removed two months later and a MAG-3 lasix renogram showed no obstruction. The patient recovered well with no other symptoms or sequella.

Discussion

SM is a benign condition, with the literature generally supporting an inflammatory origin of the disease. Emory et al suggests this diagnosis forms a spectrum with other diag-

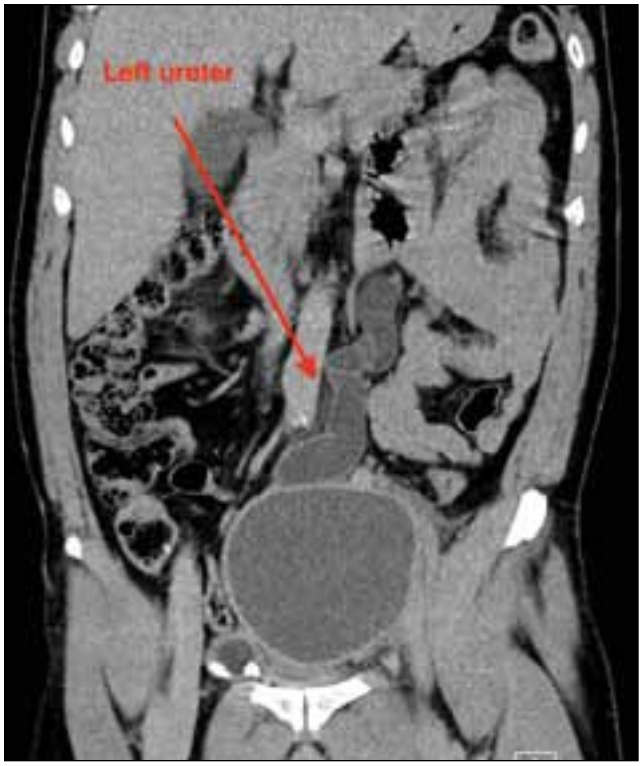

Fig. 1. Pelvic mass and hydro-ureteronephrosis on computed tomography scan.

noses, such as mesenteric lipodystrophy (fat necrosis) and mesenteric panniculitis (chronic inflammation and necrosis).[1] Defined by the macroscopic pathology, SM can be classified as three different types: 1) diffuse thickening of the mesentery; 2) single knotty thickening at the root of the mesentery; and 3) multiple knotty thickenings of the mesentery.[9,10] Two series of 53 and 84 patients show a variable division of 17–42% type 1, 32–70 % type 2, and 13≤26% type 3.[1,10] An association between prior abdominal surgery and SM has been variously reported at 5–41%.[1,6] An immunopathological etiology has also been suggested, with a possible link with IgG4 levels.[6] Paraneoplasic syndrome may be a cause of this inflammatory disease, with some studies suggesting a link in 1–70% of cases.[1,2,10] Both renal cell carcinoma and prostate adenocarcinoma are included as cancers potentially associated with SM.[2,10]

According to CT scan and autopsy series, the prevalence of SM is 0.6–2.5%. Further, SM appears to occur most commonly between the age of 50 and 70, with a male to female ratio of 2:1.[1-4]

Clinical manifestations are often vague, including abdominal pain, gastrointestinal symptoms, weight loss, or any local mass effect. In our case, the patient experienced back pain due to severe obstructive uropathy. Physical examination is uncommonly contributory, with less than 50% of patient presenting a palpable mass, and laboratory findings

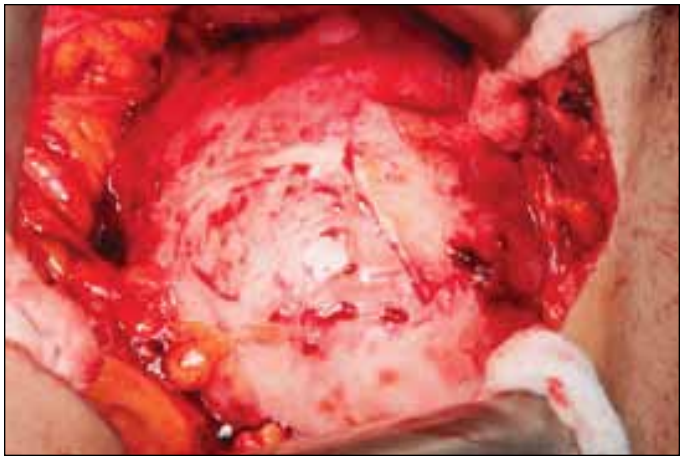

Fig. 2. Pelvic mass during surgery.

are non-specific.[6] Anemia and hypoalbuminemia may be found, while CRP and erythrocyte sedimentation can be useful as markers of treatment response.[5] Complications of SM include obstructive uropathy, bowel obstruction, chylous ascites, and chronic mesenteric ischemia. Contrast enhanced CT scan is the best imaging modality.[2,6] Typical findings are a solid fatty mass in the mesentery with lymph nodes surrounded by a pseudocapsule ("tumor pseudocapsule sign," Fig. 3). The preservation of the densitometric values of fat around the vessels is called the "fat ring sign" (Fig. 4).[4,7,8] This sign may help distinguish SM from lymphoma, carcinoid tumor, or carcinomatosis. MRI has not been widely studied in SM. Typically, pathology is required to confirm the diagnosis. Histopathology can show fat necrosis, fibrosis, and chronic inflammation with lymphocyte infiltration.[1]

There is no proven treatment, but empiric treatment is recommended based on symptoms. Corticosteroids or tamoxifen are reasonable first-line treatment options. Surgical intervention is warranted for obstructive complications (either urinary, bowel, or vascular), but does not cure the disease.[6]

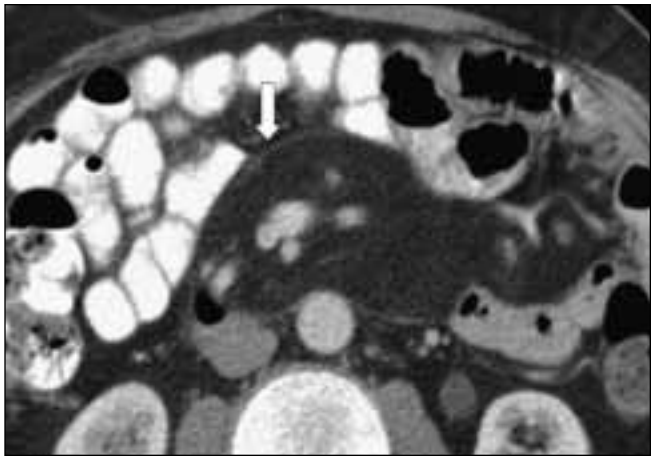

Fig. 3. Tumor pseudocapsule sign, van Putte-Katier et al.

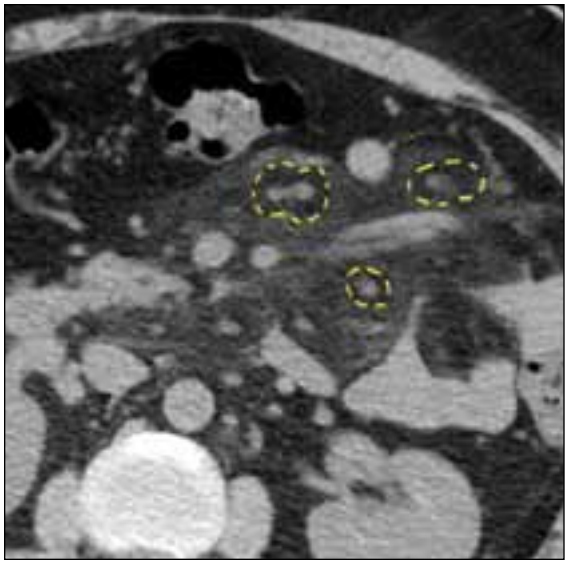

Fig. 4. Fat ring sign, case courtesy of Dr. Hani Salam, Radiopaedia.org, rID: 10092.

A 2014 study followed patients with mesenteric panniculitis (MP) (same spectrum of disease) for five years and found a significantly higher incidence of cancer within the MP group compared to the control group (14% vs. 6%).[4] This suggests that further followup may be warranted for these patients.

Conclusions

SM is an uncommon diagnosis with a variable and nonspecific presentation. Further research is needed to understand the potential link with urological cancers and the appropriate followup.

Competing interests: Dr. Toren has been an advisory board member for Abbvie, Ferring, and Pfizer; has received honoraria from Innocrin Pharma and Janssen; and has participated in clinical trials supported by Roche. Dr. Cloutier has received speaker honoraria from Boston Scientific and Storz; and participated in a clinical trial supported by Medpace. Dr. Turcotte reports no competing personal or financial interests related to this work.

This paper has been peer-reviewed.

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