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Case Report

Sclerosing mesenteritis - rare cause of abdominal pain and intra-abdominal mass: a case report

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ABSTRACT

Sclerosing mesenteritis is a rare, benign, and chronic fibrosing inflammatory disease with unknown etiology that affects the mesentery of small bowel and colon. The disease has two well-established histological types: the acute or subacute form known as mesenteric panniculitis and the chronic form known as retractile or sclerosing mesenteritis. Because sclerosing mesenteritis lacks special clinical manifestations and typical signs, the patients are very easily misdiagnosed. The correct diagnosis of sclerosing mesenteritis depends on pathological examination after laparotomy. We report a case of sclerosing mesenteritis in a 55-year-old male who presented with chronic abdominal pain and intra-abdominal mass. He was misdiagnosed as lymphoma by Computed Tomography and then underwent exploratory laparotomy. Histopathological examination revealed it to be sclerosing mesenteritis. This patient went well and lives without recrudescence till date.

Keywords: Sclerosing mesenteritis, Abdominal mass, Intestinal obstruction, Exploratory laparotomy

INTRODUCTION

Sclerosing mesenteritis is a rare, benign, and chronic fibrosing inflammatory disease with unknown etiology that affects the mesentery. The condition predominantly occurs in males and is more frequent between the 6th and 7th decades of life.[1] The bowel, adjacent lymph nodes and vessels are usually not affected. On rare occasions, it may involve the mesocolon, peripancreatic region, omentum, retroperitoneum, or pelvis.[2]

In most patients, the condition consists of a mixture of chronic non-specific inflammation, fat necrosis and fibrosis. So the clinical manifestations can vary. Patients may present with abdominal pain, intestinal obstruction, fever, chylous ascites, a mass, constipation or diarrhoea.[3,4] Because its clinical manifestations are non-specific and atypical, the preoperative diagnosis of sclerosing mesenteritis can be very difficult.[5]

We report a case of sclerosing mesenteritis in a 55-year-old male who presented with chronic abdominal pain and intra-abdominal mass. He was misdiagnosed as lymphoma by computed tomography and then underwent exploratory laparotomy. Diagnosis was then confirmed as sclerosing mesenteritis by histopathological examination.

CASE REPORT

A 55-year-old male patient presented with chronic abdominal pain for about 6 months and intra-abdominal mass for about 1 month. The pain was dull aching, intermittent, mild and mainly located in the periumbilical area. He had complaints of nausea and decreased appetite. He gave no history of fever, weight loss, abdominal trauma or any other bowel bladder complaints.

On per abdominal examination, he had 2 firm lumps with restricted mobility involving the left lumbar and right

iliac regions of approximate size 6x5 cm. The laboratory profile of routine blood tests showed total leukocyte count of 15000/cumm and renal and hepatic function tests were normal. He was Australia antigen positive. Ultrasonography showed small edematous bowel loop with ill-defined mass 3.8x3.8 cm in paraumbilical and left lumbar area suggesting retroperitoneal fibroma/ sarcoma. Computed Tomography revealed a soft tissue density mass lesion 50 mmx45 mm in mesenteric tissue near root. Another lesion in mesentery at the level of infrarenal aorta level indicated lymphoma (Figure 1).

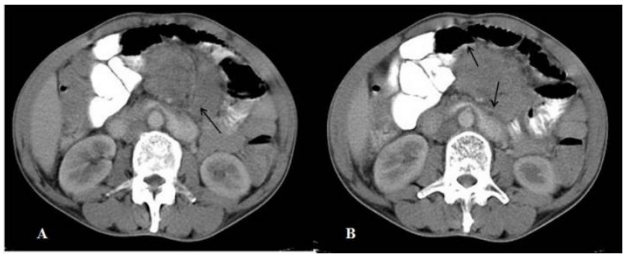

Figure 1: A) Axial CT scan show fatty soft tissue mass (arrow) related to small bowel and mesentery. B) Axial CT scan show peripheral curvilinear band of soft-tissue attenuation (arrow), limiting heterogeneous mass from surrounding normal mesentery known as "tumoral pseudocapsule" sign.

On exploratory laparotomy a fibrotic mass was detected in the small bowel mesentery from duodeno-jejunal flexure to ileocecal junction (Figure 2). Although a few small intestines were tightly adhered to the mass, the involved intestine had no obstruction (Figure 3). Incisional biopsy of the mass was taken (Figure 4). Biopsy showed fibroadipose tissue with few blood vessels with focal lymphocytic infiltrate. There was no evidence of tuberculosis or malignancy. The inflammation stopped abruptly at the edge of bowel wall (Figure 5). So this case was diagnosed as sclerosing mesenteritis at last by histopathological examination. The patient did not take immuno-suppressants and recovered well after operation.

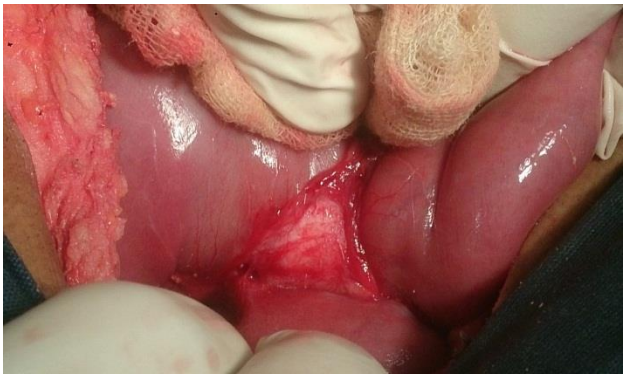

Figure 2: Fibrotic mass in small bowel mesentery from duodeno-jejunal flexure to ileocecal junction.

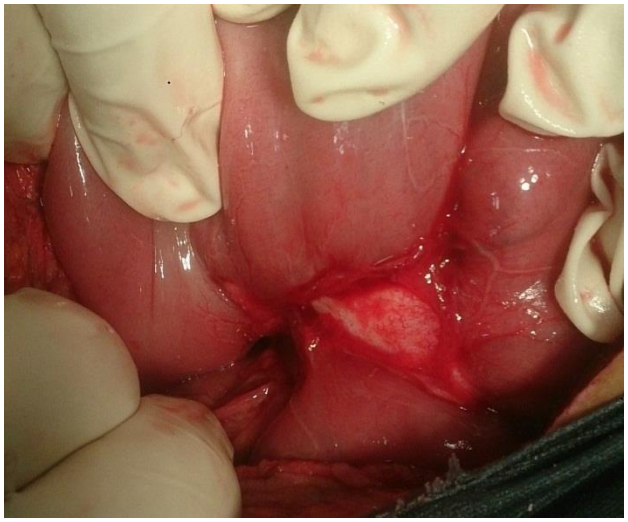

Figure 3: Mass adherent to small intestine but no obstruction.

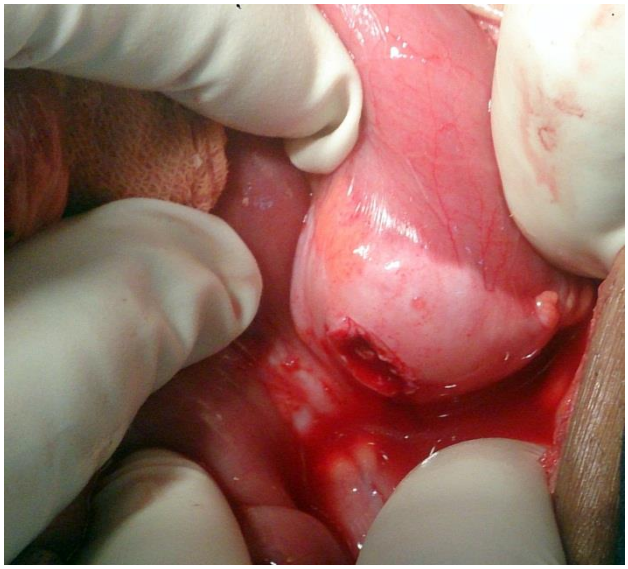

Figure 4: Incisional biopsy from the mass.

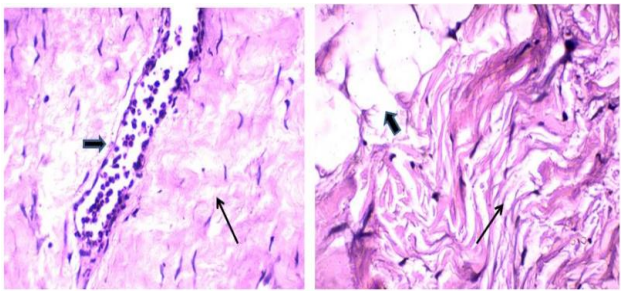

Figure 5: Microphotograph showing fibroadipose tissue with few blood vessels with focal lymphocytic infiltrate. A) H&E (40x) section showing inflammatory cells (thick arrow) in vessel and surrounding fibrocollagenous tissue (thin arrow). B) H&E (40x) section showing fat necrosis (thick arrow) and sclerosing fibrosis (thin arrow).

DISCUSSION

Sclerosing mesenteritis is a rare disease of unknown etiology that is characterized by tumor-like mass composed of chronic nonspecific inflammation, fat necrosis and fibrosis. Chronic bellyache and intraabdominal mass are its main clinical manifestations.[6] Although various causes have been suggested, including trauma or ischaemia of the mesentery, self-immune, the exact etiology of the disease still cannot be affirmed.[7] Etiological associations of mesenteric panniculitis include malignancy, infectious disease, autoimmune disease, vasculitis, cirrhosis, peptic ulcer disease, Gardner's syndrome, orbital pseudotumor, pancreatitis, abdominal aortic aneurysm, previous abdominal surgery and elevated IgG4 related disease.[8]

It has been linked with malignancy, in particular lymphoma, gastrointestinal and genitourinary. 30% - 50% patients remain asymptomatic. In terms of autoimmune disease, its associations have been with autoimmune haemolytic anaemia, coeliac disease, thyroiditis, primary sclerosing cholangitis, rheumatoid arthritis, lupus and polychondritis.[9]

Approximately 90% of cases involve the small-bowel mesentery and changes are more commonly centered to the left of the midline corresponding with the jejunal mesentery.

It can be categorized into three pathological changes: chronic non-specific inflammation, fat necrosis and fibrosis.

Based on the different course of disease, many terms have been used to describe sclerosing mesenteritis, including mesenteric lipodystrophia, retractile or liposclerotic mesenteritis, mesenteric Weber-Christian disease, xantogranulomatous mesenteritis, mesenteric lipogranuloma, and systemic nodular panniculitis.[2] This varied terminology has caused considerable confusion, but the condition can now be evaluated as a single disease with two pathological subgroups. If inflammation and fat necrosis predominate over fibrosis, the process is known as mesenteric panniculitis; when fibrosis and retraction predominate, the result is retractile mesenteritis. The overall presence of some degree of fibrosis makes the pathologic term sclerosing mesenteritis more accurate in most cases. In most cases, sclerosing mesenteritis involves the alvine mesentery, but it can also affect the mesocolon, peripancreatic region, omentum, retroperitoneum, or pelvis.[4]

Till now, just about 300 cases have been reported in world literature.[10]

Few scholars have suggested that abdominal CT scan and Magnetic Resonance Imaging (MRI) take important roles in suggesting the accurate diagnosis and can be used for distinguishing sclerosing mesenteritis from other mesenteric diseases with similar imaging features such as carcinomatosis, carcinoid tumor, lymphoma, desmoid tumor, and mesenteric edema. Two CT findings are considered somewhat specific for this disorder: a "fat ring sign" that reflects the preservation of fat around the mesenteric vessels, and the presence of a "tumoral pseudocapsule," which is detected in 50% of patients.[11]

The imaging appearances of sclerosing mesenteritis vary depending on the predominant tissue component (fat necrosis, inflammation, or fibrosis). So the accurate diagnosis of sclerosing mesenteritis can be established only by evaluating a biopsy specimen. It is very easy to be misdiagnosed not only by the radiologists and surgeons, but by the pathologists. Some authors recommend that asymptomatic or those with mild symptoms may be left untreated and observed, surgical resection is advocated for patients with life-threatening complications such as bowel obstruction or perforation. Potential treatment options include surgical resection, immunomodulatory (steroids, azathioprine, cyclophosphamide), antifibrotic agents (thalidomide, tamoxifen, pentoxyfylline NSAIDS (colchicine) and conservative management.[12-14]

A review of the literature suggests that currently there is no standard treatment and management should be guided by patients symptoms. Mesenteric panniculitis is rare; as a result evidence for treatment is limited to individual case reports.

It has a favourable prognosis and may be self-limiting.

CONCLUSION

Our case illustrates that the diagnosis of sclerosing mesenteritis can be difficult preoperatively. Tissue diagnosis is absolutely essential to avoid misdiagnosing a malignancy as sclerosing mesenteritis on radiological appearance. Knowledge of such rare conditions motivates us into further work on its etiopathogenesis and management. The rarity of this case makes it interesting.

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