Sclerosing mesenteritis presenting with abdominal mass: Case Report

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Abstract

Sclerosing mesenteritis is a rare disorder characterized by a tumor-like expansion of the mesentery due to variable degrees of fat necrosis, chronic inflammation and fibrosis. It usually involves the root of the small bowel mesentery, but it can also involve the mesocolon, the peri-pancreatic and omental fat, and infrequently the retroperitoneal or pelvic fat. Although sclerosing mesenteritis is non-neoplastic and benign disease it can have fatal outcome. The use of drugs, trauma or ischemia of the mesentery, malignancy, autoimmunity, avitaminosis, pancreatitis, and a history of abdominal surgery has been suggested as possible causative factors. We report a rare case of a 57 year old male patient with sclerosing mesenteritis who presented with acute abdominal pain and intra-abdominal mass. He was misdiagnosed as colonic ischemia by colonoscopy which was partially excluded by the CT (computed tomography) and then underwent exploratory laparotomy and colostomy with mucous fistula. Finally, he was diagnosed correctly by histopathological examination of the paraffin section of the specimens taken during the laparotomy.

Introduction

Sclerosing mesenteritis is defined as a rare, benign process involving mesenteric adipose tissue by three types of pathological changes which are fat necrosis, chronic inflammation, and fibrosis [1-3]. Various names have been used to describe the condition including mesenteric lipodystrophy, retractile or liposclerotic mesenteritis, mesenteric Weber-Christian disease, xantho-granulomatous mesenteritis, mesenteric lipogranuloma, and systemic nodular panniculitis [2, 4-6].

Due to the different and non-specific presentations and findings, the disease poses great diagnostic challenges leading to misdiagnosis in majority of the cases [1, 3, 5, 6]. Although definitive diagnosis requires biopsy and histopathology, various radiological modalities such as multidetector computed tomography (CT) and Magnetic Resonance Imaging (MRI) have been used to make the diagnosis with appropriate clinical analysis [4, 6, 7]. The clinical manifestations are largely non-specific. The patient may be absolutely asymptomatic with diagnosis made incidentally [3-7]. Other findings may include vague abdominal discomfort and a palpable mass, distention, progressive fatigue, weight loss, nausea and vomiting, tender mass in the abdomen, fever, malabsorption syndrome, chylous ascites, and pneumoperitoneum [1, 7-9].

Laboratory workup such as complete blood count, peripheral blood film, erythrocyte sedimentation rate, serum C-reactive protein, amylase, lipase, liver and renal function tests, and autoimmune workup is usually negative in cases of pure sclerosing mesenteritis [2, 8, 9].

Most studies from Western countries have indicated that sclerosing mesenteritis is more common in men than women, on the order of 2:1. Most patients are in the fifth to seventh decades of life, with a median age of 65 years. Pediatric cases are rare because children have lower amounts of mesenteric fat than adults [1, 5, 7]. Added to that, in Western patients, the disease usually involves the small bowel mesentery and extremely rare the sigmoid mesentery [2]. On the other hand, in Far East patients, the disease involves the large bowel mesentery and researchers found that the sigmoid mesentery was most frequently involved among the large bowel mesentery [1, 8]. Therefore, the portion of the mesentery most frequently involved in sclerosing mesenteritis appears to differ between races. Although some genetic factors may potentially be involved, the precise reasons producing these differences have not been clarified [1, 2, 5, 7].

The precise etiology of sclerosing mesenteritis remains unknown. However, the disease appears to be caused by a nonspecific response to a wide variety of stimuli, including abdominal surgery or trauma, autoimmunity, malignancy, ischemic injury and infection. Other factors have also been reported, such as chronic inflammatory conditions, collagen vascular diseases and IgG-4 related disease [2, 9, 10].

Inflammation and fibrosis, as well as a tendency to encase surrounding structures, make surgical management of retroperitoneal inflammatory conditions challenging and dangerous, as it has the potential to damage adjacent organs such as ureters.
and bowel [8-10]. In contrast, medical treatment with immunosuppression is often successful, and can limit the potential morbidity of surgical interventions [1, 7, 9]. In a retrospective and prospective, single-institution (Mayo clinic) study examining treatment outcomes in 92 patients diagnosed with Sclerosing mesenteritis (25 years study), by Akram et al, 45% of patients had surgery. In those patients who were surgically managed, partial to complete resection of the mass was only possible in 30%. The remaining 70% of patients had only segmental small bowel resection, palliative bypass or adhesiolysis secondary to inability to resect the mesenteric mass. Only two percent of patients who were surgically managed responded to surgery alone. However, 41% of patients who were treated medically had clinical improvement [11].

Retroperitoneal inflammatory conditions are often triaged and managed by rheumatologists, general surgeons, gastroenterologists and urologists [1, 3, 5]. While histopathologic confirmation of the diagnosis is required, medical management is a safer and more effective treatment modality than surgery in these patients [2, 8, 12].

Case Report

57 years old male patient known case of type II diabetes on oral hypoglycaemics. The patient was admitted by the gastroenterology team as a case of abdominal pain with mucus discharge per anus and tenesmus but no blood per rectum. During this time, examination showed left iliac fossa mass about 5 x 5 cm. Colonoscopy was done and showed diffuse congestion in the rectum and sigmoid with cystic-like mucosa, suggestive of pneumatosis cystoid intestinalis (Figure 1). Multiple biopsies were taken from the sigmoid and the rectum which showed no pathologic findings. Chronic phase of ischemic colitis has been considered. His laboratory investigations showed WBC 9.3 x 10^9/L, Hb 17.3 g/dL, CRP 286 mg/L and lactic acid 2 mmol/L. Consultation with the Colorectal surgeon was done and he kept the patient NPO (Nothing Per Oral) and CT (Computed Tomography) of the abdomen and pelvis with oral and rectal contrast done. These showed circumferential diffuse wall thickness slightly irregular affecting the left colon, sigmoid and rectum with marked peri-colonic fat stranding, no collection detected and no enlarged peri-colonic lymph node. The ascending colon and the transverse colon were not significantly dilated. The radiological impression was suggestive of infectious or inflammatory colitis rather than neoplastic or ulcerative colitis (Figure 2).

The patient was taken for laparotomy exploration based on picture of severe abdominal pain and impending intestinal obstruction. The laparotomy revealed fibroed, markedly oedematous mesentery of the entire left colon extending down to the upper. The surgeon could not proceed for left procto-colectomy because there was diffuse bleeding, significant in amount from the divided inflamed edematous wide mesenteric base. Also the patient became hypotensive. Inotropes and blood were given by the anesthetist. Eventually the surgeon did diversion colostomy with mucus fistula and multiple biopsies were taken from the mesentery and the colonic wall. Hemostasis was achieved, two drains were placed. The patient was then admitted in ICU post-operative, and discharged the next day to the surgical ward. The patient was improving regarding the abdominal pain with functioning colostomy. He was discharged in good health with daily dressing and follow up in 3 weeks.

Histopathology of the descending colon showed acute serositis, no mucosal necrosis and no dysplasia or neoplasm seen. The excisional biopsies of the appendices epiploicae showed fat necrosis, acute inflammation, mesothelial hyperplasia and vascular congestion consistent with epiploic appendagitis and no evidence of neoplasm. The excisional biopsy form the omentum showed acute serositis and vascular congestion with no evidence of neoplasm. Based on the radiological, surgical and histopathological findings, the patient diagnosed as mesentritis.

On the follow up visits 3 weeks post-operative abdominal wound completely healed, prednisolone and tamoxifen were started based on literature review of similar cases. The patient condition improved with disappearance of the abdominal mass in the following visits.

Discussion

In this report, we present a patient with histologically proven acute sclerosing mesentritis which was diagnosed during a difficult surgery with subsequent good response to oral prednisolone and tamoxifen with complete disappearance of the inflammatory mass and resolution of clinical symptoms.

Part of the controversy over the etiology and classification of this disorder is due to its rarity. About 300 cases have been reported in the literature, all heterogeneous in terms of natural history, clinical presentation and effective treatment [13-15]. The first series about the disease was published in 1924.
compromising 34 cases [16]. In a series of 92 patient at Mayo clinic over 25 years by Akram et al, the most frequent presenting symptoms were abdominal pain in 70%, bloating and distention in 26%, diarrhea in 25%, and weight loss in 23% [11]. CT with oral and rectal contrast showed Sclerosing Mesentritis as an incidental finding in 10% of cases when an abdominal surgery (3%), computed tomography (CT) scan (5%), or autopsy (1%) was performed for another indication, and there were no symptoms attributable to mesenteric disease. The diagnosis was established at laparotomy with biopsy in 65%, laparoscopy with biopsy in 25%, and CT-guided biopsy in 10%. Laboratory parameters were unremarkable. In 61% of cases, an abdominal CT showed a single soft-tissue mass in the root of the mesentery, often containing calcification. In 34% of cases, there was subtle increased density of the mesenteric fat, suggesting mild mesenteric fibrosis or inflammation. Histologically, a variable combination of fibrosis, chronic inflammation, and fat necrosis was noted. The most frequent histologic finding, noted in 53%, was prominent fibrosis with scant inflammation and some fat necrosis [11].

The diagnosis of sclerosing mesenteritis is usually made by biopsy at laparotomy. The presence of a single, multiple or diffuse mass-like inflammatory lesion in the mesentery, together with a histological confirmation of fat necrosis and inflammatory reaction or fibrotic infiltration in the mesenteric lesions, strongly suggests the diagnosis of sclerosing mesenteritis [16].

The best treatment for sclerosing mesenteritis remains unclear. Asymptomatic or mild clinical forms may sometimes be left untreated with spontaneous recovery. Surgical resection is required for patients with intestinal obstruction and perforation, and immunosuppressive therapy with corticosteroids, thiadomide, and other drugs has been recommended by some authors [17] (Figure 3).

Conclusions

Sclerosing mesenteritis is a rare idiopathic disorder that involves predominantly the small bowel mesentery with varying degrees of fibrosis, inflammation and fat necrosis. Diagnosis of this nonspecific benign inflammatory disease is a challenge to surgeons, radiologists, gastroenterologists and pathologists. Its clinical presentation is quite diverse and ranges from being asymptomatic to a debilitating disease. CT features of the disease, usually highly suggestive, have recently been delineated clearly. Approximately half of the patients may not require any treatment. However, in symptomatic cases treatment should be tailored according to the severity and type of individual symptoms. Patients with bowel obstruction should undergo surgery, while those with non-obstructive symptoms might benefit from steroid therapy alone or in combination with other drugs. Overall prognosis is usually good and recurrence seems to be rare. However, we recommend long-term follow up to document treatment results, because of the spars number of cases and presence of many treatment modalities.

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