SCLEROSING MESENTERITIS: A CASE OF ACUTE ABDOMEN AND INTESTINAL OBSTRUCTION

[1]Madubogwu Chimezie Innocent, [2]Okani Chukwudi Onyeaghana

[1]Department of Surgery, Chukwuemeka Odimegwu Ojukwu University, Awka Campus, Anambra, Nigeria. (Formally: Anambra State University).
[2]Department of Histopathology, Chukwuemeka Odimegwu Ojukwu University, Awka Campus, Anambra, Nigeria. (Formally: Anambra State University).

ABSTRACT

Sclerosing mesenteritis (SM) is a rare idiopathic disorder characterized by chronic non-specific inflammation involving the adipose tissue of the bowel mesentery. It may be asymptomatic but it commonly presents with abdominal pain. Some individuals may have a palpable abdominal mass and affected individuals may develop small bowel obstruction or acute abdomen.

We report a case of 27-year old man who presented to the Surgery Department at Chukwuemeka Odimegwu Ojukwu University Teaching Hospital (COOUTH), Amaku-Awka, Anambra State with acute abdomen and intestinal obstruction. He subsequently, had a surgical resection, which was histologically confirmed as a sclerosing mesenteritis. Sclerosing mesenteritis is a rare disorder and this is the first case being reported in the literature from South-East, Nigeria. It is therefore, important to alert physicians, more especially the surgeons on the need to have a high index of suspicion in every case of intestinal obstruction.

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INTRODUCTION

Sclerosing mesenteritis (SM) is a rare clinicopathological condition that affects the mesentery of the small bowel and occasionally the mesocolon. It is largely an idiopathic condition characterized by nonspecific inflammation of the mesenteric adipose tissue[1]. It was first described by Jura in 1924 as "retractile mesenteritis" and further labeled as "mesenteric panniculitis" by Odgen later in the 1960s[2-4]. Numerous terms have been used to describe this condition based on the most predominant histological picture. They include: mesenteric lipodystrophy (when fatty degeneration and necrosis predominate), mesenteric panniculitis (when there is marked chronic inflammation involving the adipose tissue), and retractile mesenteritis or mesenteric fibrosis (if fibrosis is predominant)[3,4]. Other names that have been used for this disorder in the literature include mesenteric sclerosis, liposclerotic mesenteritis, mesenteric Pfeifer-Weber-Christian disease, mesenteric lipogranuloma, and xanthogranulomatous mesenteritis[2,5,6]. Today most articles describe the pathologic condition as "sclerosing mesenteritis", and divide it into three major phases of the same pathologic condition, each regarding a progressively worsening state. The pathologic findings begin with fat necrosis and end with regenerative fibrosis. Within this progress of disease state, fat necrosis predominates in the first step, therefore, called mesenteric lipodystrophy. This is then followed by mesenteric panniculitis, characterized by intense inflammatory reaction, and lastly by retractile mesenteritis, when fibrosis becomes the main feature[4].

Sclerosing mesenteritis may be totally assymptomatic, discovered incidentally during routine investigation for other conditions[3,7]. It has been described as a benign, self-limited disorder with a favorable prognosis that may even resolve spontaneously[8]. It may in some cases be potentially severe and debilitating[3]. The most common symptom associated with sclerosing mesenteritis is abdominal pain, which can be severe[3]. Some individuals may have a palpable abdominal mass and affected individuals may develop small bowel obstruction or acute abdomen[3]. A high index of suspicion is needed to suspect this rare entity clinically[5]. Abdominal CT is currently being proposed as an effective method for diagnostic evaluation[9]. The definite diagnosis of sclerosing mesenteritis is established by biopsy[3,6]. Most symptomatic patients might benefit from medical therapy with a variety of drugs including steroids, thalidomide, cyclophosphamide, methotrexate, progesterone, colchicine, azathioprine, tamoxifen, antibiotics and emetine, or radiotherapy, with different degrees of success[2,3,6]. Surgical exploration is however indicated in patients with intractable complications like intestinal obstruction and acute abdomen[3].

Correspondence: Madubogwu C I.
Department of Surgery, Chukwuemeka Odimegwu Ojukwu University, Awka Campus, Anambra, Nigeria. (Formally: Anambra State University)
E-mail: chymezo@yahoo.com
Phone: +234-803-400-5584.

CASE REPORT:

We report a case of a 27-year old man presenting with 2-week history of severe abdominal pain, 1-week history of abdominal distension and 2-day history of nausea, vomiting and constipation. Abdominal pain was generalized, but more on the right lower abdomen. Prior to the onset of severe abdominal pain, patient had been experiencing intermittent right lower abdominal pain for more than a year. For this reason, he was admitted in a private hospital where a provisional diagnosis of acute appendicitis was made. Laparotomy was performed; however, intra-operatively an impression of appendix mass was made. The surgical wound was closed without tempering with the appendix and the patient placed on antibiotics hoping to be operatated on a later date. Thereafter, patient improved though he still complained of occasional right lower abdominal pain which later became severe 2 weeks prior to presentation to our facility.

On examination, patient was in painful distress, febrile to touch, mildly dehydrated, not pale and anicteric. The pulse rate was 92 beats / minute and the Blood pressure was 130 / 70mmHg. The abdomen was distended with generalized tenderness and guarding more on right iliac fossa and suprapubic regions.

Investigations performed showed leucocytosis with relative neutrophilia, the haemogoblin concentration was 12g / dl, and HIV screening was non-reactive. Abdominal ultrasonography showed a complex mass of bowel loops with surrounding fluid collection in the pelvis and lower abdomen. An impression of ruptured appendix with appendix mass and abscess was made. Patient was worked up for emergency exploratory laparotomy. Intra-operative findings were: retraction and distortion of the loops of ileum, caecum and omentum and the formation of adhesions between them. There was what seemed as walling off an abscess cavity on the pelvis and right lower abdomen; thickening and sclerosis of the terminal 30cm of ileum with patchy degenerative changes of the affected loop of ileum and mesentery. There was also presence of enlarged mesenteric lymph nodes.

Right hemicolectomy with resection of terminal 30cm of the ileum was done. Patient was placed on broad-specrum intravenous antibiotics. The post-operative recovery was uneventful. Histology of the resected specimen showed mesenteric tissue infiltrated by inflammatory cells, myofibroblasts and foamy macrophages with associated infiltration and thrombosis of the vessels traversing the lesion. There was extensive fibrosis with hyalinization, chronic inflammation and fat necrosis surrounded by clusters of foamy macrophages and reactive lymph nodes form islands in the sea of fibrosis and sclerosis. The colon showed focal haemorrhagic infarction. All the lymph nodes isolated showed reactive changes ranging from follicular hyperplasia to intense sinuous histiocytosis. The overall features are in keeping with a sclerosing mesenteritis (figure 1).

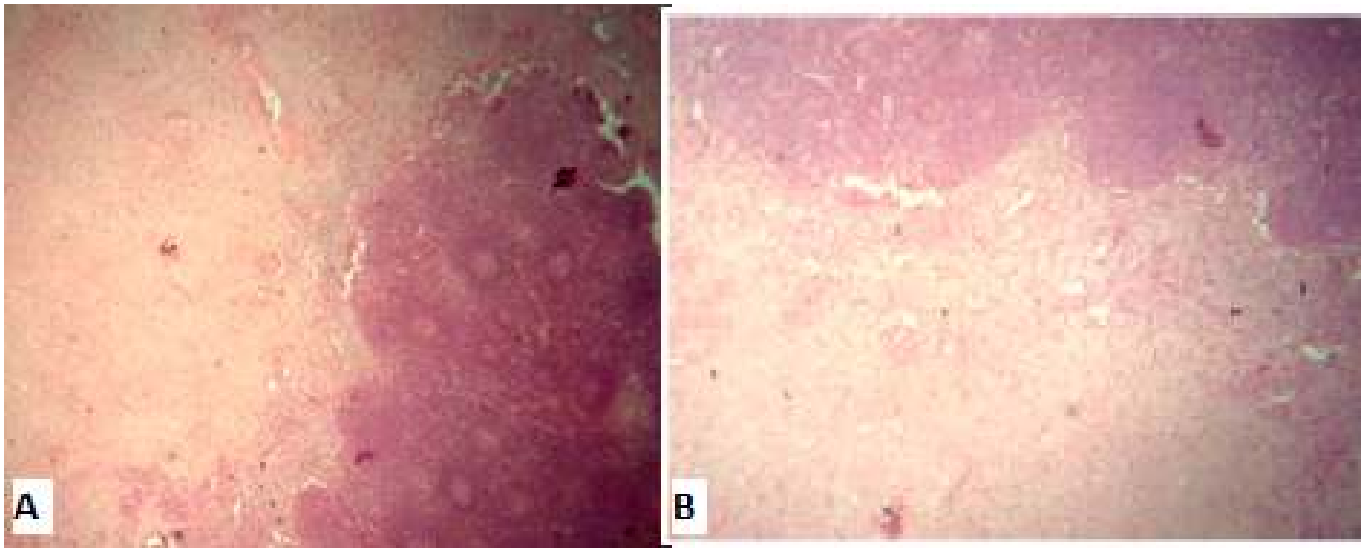

Figure 1 Sclerosing Mesenteritis. A, Low power view showing reactive lymphoid follicles with prominent germinal centres forming an island on a background of sclerosing fibrosis. B, There is replacement of fat lobules by extensive sclerosis, fibrosis, and inflammatory infiltrates of foam cells and lymphocytes, sharply demarcating the reactive lymphoid follicles.

Patient has been on follow up visits for 6 months and has remained symptomless. Post-operative abdominal CT scan was requested for but yet to be done due to financial constraint.

DISCUSSION:

Sclerosing mesenteritis (SM) is a rare idiopathic disorder characterized by chronic non-specific inflammation involving the adipose tissue of the bowel mesentery[1]. The condition has been described as rare but may actually be under diagnosed due to the relative ignorance of the condition amongst both surgeons and radiologists[5]. Less than 300 cases have been reported in the world-wide literature under several names: retractile mesenteritis, sclerosing mesenteritis, mesenteric panniculitis, liposclerotic mesenteritis, isolated lipodystrophy of the mesentery, mesenteric lipomatosis, and lipogranuloma of the mesentery, and mesenteric manifestations of Weber-Christian disease[2,4,5,9]. Only a few large series have been published in world literature[3,10-12] most other published reports are of small case series[2,5,6] or single patients like the current case. Due to the paucity of published cases there is limited information on the exact cause, the etiological factors, the natural history, the prognosis, and the treatment options of patients with SM[3,6]. Sclerosing mesenteritis is a disease of middle-aged or older adults, diagnosed primarily during the 6th–7th decade of life, the incidence increases with age[3,6,8]. Paediatrics cases are very uncommon, probably because children have less mesenteric fat than adults.[2,6]. The disease appears to be twice as common in men as in women[2,3,6,8]. Our index patient is a male and relatively younger than the usually finding in most cases.

The disease is often asymptomatic and indeed most patients in any given series were incidentally identified during a CT examination[6,9]. Most symptoms associated with SM are caused by the direct mechanical effect of the mesenteric mass encasing the bowel, blood vessels and lymphatics[6]. The most common symptom associated with sclerosing mesenteritis is abdominal pain, which can be severe[3]. Other symptoms include: nausea, vomiting, constipation, diarrhea, fatigue, malaise, fever, night sweats, abdominal distention, loss of appetite, and weight loss[3]. Akram et al[3] found the most frequent presenting symptoms to be abdominal pain in 70%, bloating and distention in 26%, diarrhea in 25%, weight loss in 23%, nausea and vomiting in 21%, anorexia in 16%, constipation in 15%, fever in 6% and night sweats in 3%. Rajendran and Duerksen[1] presented a case of SM manifesting as protein loosing enteropathy with features of diarrhea and peripheral oedema. In some cases, affected individuals may develop small bowel obstruction or acute abdomen with abdominal tenderness, palpable abdominal mass and chylous ascites.[3] Our index patient presented with features suggestive of acute abdomen and intestinal obstruction: severe abdominal pains and distension, nausea, vomiting, constipation and fever.

The exact cause of sclerosing mesenteritis is idiopathic[1,6]. The pathogenic mechanism seems to be a non-specific response to a wide variety of stimuli.[6] Some individuals with sclerosing mesenteritis have had prior abdominal surgery or trauma[3,12]. Emory et al[12] reported 84% of their patients had a history of previous abdominal trauma or surgery. Akram et al[3] reported a history of abdominal surgery in about 35% of their patients. The patient in this index case had a year history of an attempted appendicectomy which was abandoned because of finding of what appeared as an appendix mass. Some researchers have proposed that sclerosing mesenteritis is associated with autoimmune disorder[9] but no study has been able to substantiate the claim. Some individuals with sclerosing mesenteritis have had an underlying form of cancer (malignancy) such as lymphoma, Hodgkin disease, leukemia, gastric carcinoma or colon cancer. Daskalogiannaki et al[9] found an association of SM and malignancy in 69.4% of their patient but more than half of the cases were extra-abdominal in location. Wikes et al[13] discovered malignancy in 38% of their 108 patients mostly colorectal, lymphoma and urogenital tract in that order. Vlachos et al[6] found colon cancer in 60% of the 5 cases they studied. In other reports, no increase in the occurrence of cancer is seen[14]. Some other researchers have speculated that, in some cases, sclerosing mesenteritis may be a paraneoplastic syndrome[13].

Individuals with sclerosing mesenteritis have a soft-tissue mass that is detectable by radiographic study such as abdominal ultrasound, computerized tomography (CT) scanning or magnetic resonance imaging (MRI). Ultrasound typically demonstrates distortion and thickening of the root of the mesentery with slight decrease in echogenicity. Mass effect may be evident. A halo of sparing around vessels may be also seen on ultrasound as a region of hyperechoic fat. Abdominal CT has been proposed as the standard diagnostic radiological investigation[9]. CT criteria for the diagnosis of SM were the following: a solitary well-defined mass composed of inhomogeneous fatty tissue with attenuation values higher than those of the retroperitoneal fat at the root of the small-bowel mesentery, engulfment of superior mesenteric vessels without vascular involvement, and no evidence of invasion of the adjacent small-bowel loops even if displaced[9]. Our patient was only able to do abdominal ultrasound which showed a complex mass of bowel loops with surrounding fluid collection in the pelvis and lower abdomen, suggesting probably a ruptured appendix with a mass and abscess. The definite diagnosis of SM is established by biopsy. Variable combinations of foci of fat degeneration and necrosis, a nonspecific and predominantly lymphocytic inflammatory infiltrate and fibrosis have been reported[3,8].

There is still no general consensus on the treatment of SM[3,6]. Treatment is reserved for the symptomatic cases. Asymptomatic cases are usually observed over time. A lot of authors have reported good response of symptomatic cases to a variety of anti-inflammatory, immunomodulatory, and antifibrotic agents like steroids, thalidomide, cyclophosphamide, methotrexate, progesterone, colchicine, azathioprine, tamoxifen[3,6]. Surgical exploration and resections has been reserved for those with intractable complications like acute abdomen or intestinal obstruction or when other indications for abdominal exploration cannot be ruled out[3]. However attempted surgical resection or debulking usually does not result in resolution of the symptoms or prevent disease progression[3,6]. The patient in this case study only received bowel resection and has remained asymptomatic for more than six months after.

CONCLUSION

Sclerosing mesenteritis can manifest as acute abdomen or intestinal obstruction. A high index of suspicion is needed in making the diagnosis and confirmation of diagnosis is by biopsy. Surgical exploration and resection is still useful in complicated cases.

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