CASE REPORT

SCLEROSING MESENTERITIS - AN ARDUOUS TASK FOR CLINICIANS: RARE CASE REPORT

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ABSTRACT: Sclerosing mesenteritis (SM) is a rare, chronic, benign fibrosing condition of small and large bowel mesentery, found incidentally in half of reported cases. This disease generally has an indolent course, eventually may present with significant morbidities and unusually, death.[1,2] Among the mesentery attached to small and large bowel, SM mainly affects that surrounding small intestine and only rarely the mesentery around the latter. Its etiology is unknown, while the pathogenesis is obscure and pathological characteristics of the disease are nonspecific.[3] Patients may have varied clinical manifestations and present with abdominal pain, fever, chylous ascites, abdominal mass, constipation, gastrointestinal bleeding or diarrhoea and in rare cases, with small bowel obstruction. As clinical presentations and imaging patterns are non-specific, a proper coordination between radiologists, surgeons and pathologists is essential and only histopathological analysis, provides a sound diagnosis of SM masses.[1,2] As very few cases have been reported in literature, we hereby document a rare and interesting case of SM of small bowel in an elderly female.

KEYWORDS: Sclerosing mesenteritis, small bowel, histopathology.

INTRODUCTION: Sclerosing mesenteritis (SM) is a rare, non-specific, benign and chronic fibro-inflammatory disorder of unknown etiology, primarily affecting small bowel mesentery. Root of small bowel mesentery is chiefly involved and infrequently mesocolon, peripancreatic fat, omental and retroperitoneal or pelvic fat.[3]

SM was first known, described and published (1924) under the names of "retractile mesenteritis" and "mesenteric sclerosis". Later, Herrington and associates introduced mesenteric manifestation of Weber- Christian disease", "Isolated lipodystrophy" by Crane and al. and "mesenteric lipogranuloma" by Weeks and co-workers.[3,4]

Extensive elaboration on these terminologies have undergone, on the basis of the histological pattern to better describe the disease, including mesenteric lipodystrophy (predominantly fatty degeneration and necrosis), mesenteric panniculitis (marked chronic inflammation) and retractile mesenteritis or mesenteric fibrosis (predominant fibrosis). This condition has been a grey zone for clinicians due to varied terms and can now be evaluated as a single disease with two pathological subgroups, acute form called mesenteric panniculitis (MP) (predominated histologically by inflammation of the mesenteric fat) and chronic form known as retractile mesenteritis (RM) (mainly fibrosis).

Overall presence of some degree of fibrosis makes the pathological term "sclerosing mesenteritis" an accurate and preferred term in most cases.[2,3,5,6]

SM has 0.6% prevalence in patients undergoing abdominal computed tomography (CT) for varied etiologies, while an autopsy series reported 1%, suggesting many cases go undiagnosed during life.[3,5,6]

Less than 300 cases have been reported in world literature and are often diagnosed incidentally. Mean age range is 50-70 years and M:F ratio being 2:1. Coordination between radiologists, surgeons and pathologists is mandatory for a definitive diagnosis of SM masses.[1,2,3,4]

CASE REPORT: A 65 year old woman, chronic alcoholic, presented with right lumbar pain and mass per abdomen of 4 months duration. Mass was irregular, nodular, firm in consistency freely mobile and measured 7 x 5 cms extending from umbilical region to right lumbar region and 10 cms above inguinal ligament along the midclavicular line.

Routine laboratory investigations and CEA levels were insignificant. Ultrasonography (USG) showed a heterogenous hyperechoeic mass in right lumbar region and mild ascites. These findings were confirmed on contrast enhanced computed tomography (CECT) scan and in addition showed heterogeneous nodular omental thickening. Intraoperatively mass was seen originating from rectus sheath and infiltrating the ileal segment.

Grossly, segment of small bowel measured 11x8x4 cms with a firm nodular mass on serosal aspect involving mesentery, measuring 5x4 cms. Bowel wall adjacent to mesenteric mass was thickened, edematous and markedly congested. Cut section of small bowel showed normal mucosa with marked thickening of serosal layer and mass revealed grey white areas with focal fatty tissue. Mesentery showed 3 small lymph nodes, largest measuring 1.0x0.5 cms.

Microscopy of mass involving mesentery revealed adipose tissue, dense fibrocollagenous tissue, infiltrated by sheets of lymphocytes, numerous plasma cells, foamy macrophages, foci of fat necrosis and polymorphonuclear infiltration. Mucosa of small intestine adjacent to mass was unremarkable and serosa showed congested blood vessels and chronic inflammatory infiltrate. Mesenteric lymph nodes showed reactive changes.

DISCUSSION: Sclerosing mesenteritis is a rare disease of unknown pathogenesis represented with various non-specific symptomatologies and has become an area of clinical interest during recent years. There is a lack of established knowledge about etiological factors, natural history, prognosis and therapeutic options of SM, due to rarity of published cases.

Pathophysiology and pathogenic mechanism of SM still remains unknown and seems to be a non-specific response to a wide variety of stimuli.[3,6]

However, possible etiologies include previous abdominal trauma or surgery, powdered surgical gloves induced peritoneal adhesions and fibrosis, autoimmune conditions, infective causes (TB), vascular insufficiency, retained suture material.

Other factors like coronary disease, gallstones, cirrhosis, peptic ulcer, chylous ascites or abdominal aortic aneurysm. A strong relationship between SM and tobacco consumption, its association with other idiopathic inflammatory disorders such as retroperitoneal fibrosis, sclerosing cholangitis, Riedel's thyroiditis and orbital pseudotumours have been documented in recent studies.

Malignant diseases known to be associated with SM are lymphoma, carcinomas of colon kidney, lung, stomach, melanoma, myeloma, Hodgkin's disease, chronic lymphocytic leukemia, thoracic mesothelioma and carcinoid tumor, while the exact pathogenetic link is unclear.[3,6,7]

SM has varied symptoms which include intermittent abdominal pain (34.6%), abdominal mass (30.8%), nausea/vomiting, weight loss, altered bowel habits, with or without fever.[3,6,8,9]

Complete blood count, peripheral blood film, ESR, serum C-reactive protein, amylase, lipase, liver and renal function tests and autoimmune profile is usually insignificant in classical SM.[8]

CT findings of SM are variable and may be seen as increased attenuation ("misty mesentery"), to a solid soft-tissue mass in the small bowel mesentery. Other findings include enveloping of and preservation of fat around the mesenteric vessels, referred to as "fat ring sign". Thus CT plays an important role in suggesting diagnosis in the proper clinical setting and is useful in distinguishing SM from other mesenteric diseases such as lipoma, liposarcoma, lymphoma, carcinoid tumor and mesenteric carcinomatosis.[3,10,11,12,13]

Gunduz et al reported a case of SM in elderly female, while Durst et al. described this condition in middle or late adulthood with slight male preponderance and quoted incidence of SM, in 50% of clinically palpable masses.[14]

Daskalogiannaki et al. stated that most cases of MP are asymptomatic and incidentally detected on abdominal CT for unrelated conditions and Akram et al. documented presence of chylous ascites in 14% cases of MP.[3,6,12,13]

As the clinical presentation is diverse and ranges from being asymptomatic to a debilitating disease, diagnosis of SM is a challenge to surgeons, radiologists, gastroenterologists and pathologists. The diagnosis of SM is usually made by biopsy at laparotomy. Presence of a solitary, multiple or diffuse mass like inflammatory lesion in mesentery together with histological confirmation of fat necrosis, chronic inflammation and fibrosis, strongly suggests diagnosis of SM.[3,14]

This disease has three modes of progression, partial or complete resolution, non-progressive course and an aggressive course that involves fibrosis. Treatment in symptomatic cases should be tailored according to severity and nature of individual symptoms using steroids and immunosuppressive therapy.

Although overall prognosis is good in about 20% of patients, SM is associated with significant morbidity and chronic debilitating course, while recurrence seems to be rare.[15] The disorder has a self- limiting benign course in most of cases and has favourable outcome, if diagnosed accurately on time. Experiences from literatures, implies a dire necessity for more research on this subject.

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FIGURES WITH LEGENDS

Fig. 1: **Gross examination:** Segment of small intestine measuring 11 x 8 x 4cms with nodular mass on the serosal aspect involving mesentery, measuring 5 x 4 cms.

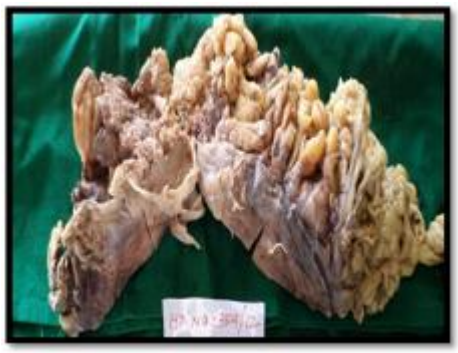

Fig. 1

Fig. 2: Cut section of mesenteric nodular mass: Shows thickened serosa, with grey white areas and focal fatty tissue. Mucosa appears normal.

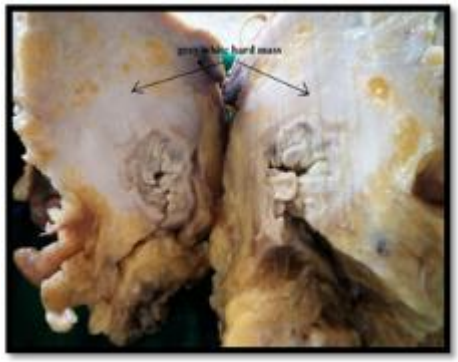

Fig. 2

Fig. 3: Microscopy of nodular mass: Shows adipose tissue and dense fibrocollagenous tissue, infiltrated by sheets of lymphocytes admixed with numerous plasma cells, foamy macrophages and focal fat necrosis (H & E, x 40).

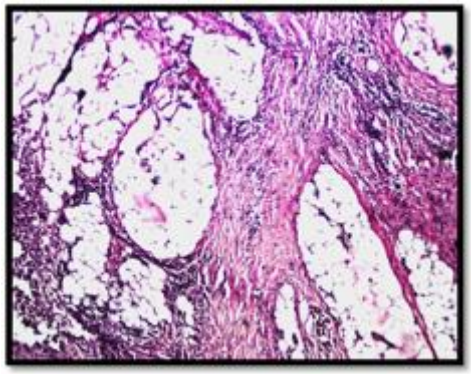

Fig. 3

Fig. 4: Focal aggregates of small lymphocytes, fibrosis and fat necrosis (H & E, x 100).

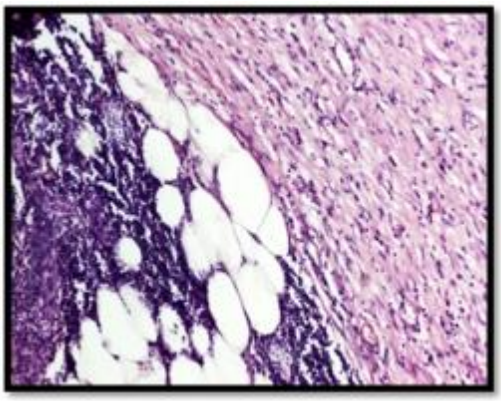

Fig. 4

Fig. 5: Small lymphocytes around capillaries and focal steato-necrosis with lipid laden macrophages and dense fibrosis (H & E, x 200).

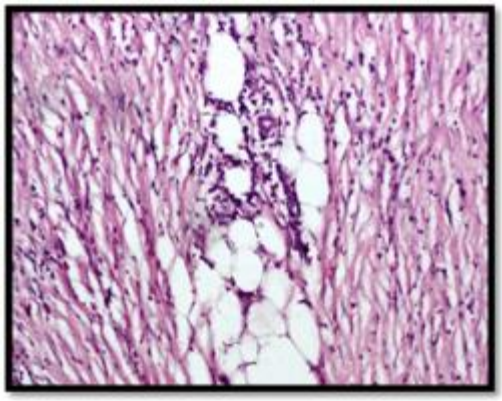

Fig. 5

Fig. 6: Small intestine showing normal mucosa, congested blood vessel and lymphocytic infiltration in the serosa (H & E, x 100).

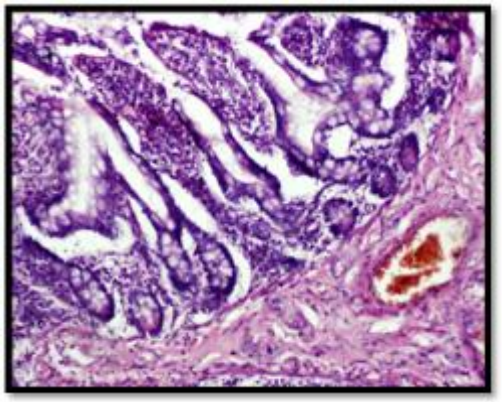

Fig. 6

Fig. 7: Mesenteric lymph node showing reactive changes (H & E, x200).

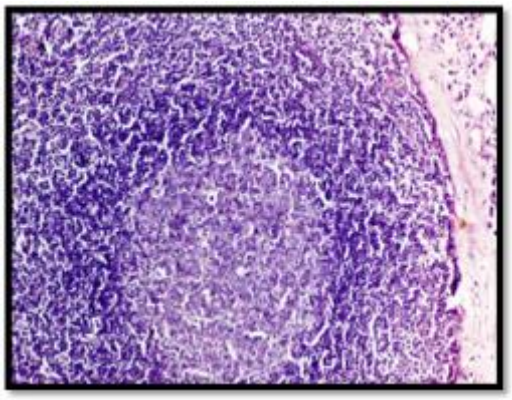

Fig. 7

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