Sclerosing mesenteric panniculitis in a young patient : common cause of diagnostic dilemma and treatment refractoriness

Ritu Mehta[1,*], Gaurav P.S. Gahlot[1,*], Prasenjit Das[1], Gaurav Jindal[2], Venigalla Pratap Mouli[2], Kumble Seetharama Madhusudhan[4], Raju Sharma[4], Sujoy Pal[3], Vineet Ahuja[2], Siddhartha DattaGupta[1]

Departments of (1) Pathology, (2) Gastroenterology, (3) Gastrointestinal Surgery and (4) Radiology, All Indian Institute of Medical Sciences, New Delhi, India.
* Both the authors have contributed equally.
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Abstract

Background : **Sclerosing mesenteric panniculitis (SMP) is an idiopathic chronic fibroinflammatory disorder of the intra-abdominal fat.**

Case presentation : **Herein, we report a case of SMP, involving the omentum, mesentery and peri-colic fat in a 18 year old male, who presented with significant and recurrent abdominal distension for 4.5 years. Computed tomogram revealed ascites, with nodular and irregular omental thickening and foci of calcification. Non-specific radiological and histological features made an accurate diagnosis extremely difficult. After a thorough work up and exclusion of other differentials, diagnosis of a nodular SMP (Weber Christian disease) was given. After showing resistance to chemotherapeutic agents, slow response was noted with cyclophosphamide, followed by rapid symptomatic improvement with mesenterectomy.**

Conclusion : **SMP is an uncommon benign mesenteric/ omental inflammation, and is a diagnosis of exclusion. As treatment refractoriness is common, management should be individualized and continued for a long period. Surgical omentectomy may be helpful.** (Acta gastroenterol. belg., **2016**, 79, **254-256**).

Key words : sclerosing mesenteric panniculitis, omental nodular thickening, Weber Christian disease, IgG4 related disease, mesenterectomy, omentectomy.

Introduction

Sclerosing mesenteric panniculitis (SMP) is a rare, benign, idiopathic inflammatory condition (prevalence : 0.6%) of adipose tissue, primarily affecting the root of the mesentery or omentum, mostly in elderly males (M:F- 1.8:1) (1-5). It is characterized by the presence of fat necrosis, inflammatory cell infiltrate and fibrosis. Other known rare sites of involvement are peri-colic fat, retroperitoneum, pelvis, peri-pancreatic region and mesocolon (6). The first known series, was published in 1924, by *Jula et al*. (2), who described the entity as "retractile mesenteritis". Depending upon the predominant finding, three terminologies have been used for similar lesion, as : mesenteric panniculitis- predominance of inflammation and fat necrosis ; mesenteric lipodystrophy- prominent fat necrosis ; sclerosing panniculitis- sclerosing lesion with inflammation. Due to rarity, knowledge is limited, also about the management protocol. Herein, we report a 16 year old boy, who was diagnosed to have SMP, highlighting the diagnostic dilemma and therapeutic challenges.

Case Presentation

This 16 year old male presented 4 years back with expectoration and abdominal distension of 1 month duration, without any history of fever and loss of appetite. Routine biochemical examination revealed raised antithyroid peroxidase 128 IU/ml (0.4-1.1) and antithyroglobulin antibodies 3987 IU/ml (0-5.61 IU/ml), which on fine needle aspiration attributed to Hashimoto's thyroiditis. Bilateral hilar prominence and pleural effusion on chest roentgenogram ; coarse liver echotexture and ascites on ultrasonogram (USG) were noted. Contrast enhanced computed tomogram (CECT) showed omental/ peritoneal bulky homogeneous soft tissue mass, compressing the stomach and small bowel with moderate ascites (Fig. 1A). Radiologically diagnosis of peritoneal carcinomatosis was made. Ascitic fluid glucose was 97 mg/dl and protein level was a 2.6 gram / deal and there was an absence of malignant cells. Anti-tubercular treatment (ATT) was started empirically in a private hospital. After an initial mild symptomatic improvement for 8 weeks, the abdominal distension and ascites reappeared. Hence, an omental biopsy was taken, which showed benign mesenchymal lesion, not otherwise specified. Two subsequent omental biopsies also revealed nonspecific finding. In all, there were polygonal to spindle shaped cells arranged in short fascicles with minimal nuclear pleomorphism, along with sheets of foamy histiocytes, chronic inflammatory cell infiltrate, fat necrosis, touton type of giant cells and intracytoplasmic fat crystals (Fig. 2). He was then referred to our institute, where ascitic fluid tubercular polymerase chain reaction was negative. The serum amylase level and pancreas was normal radiologically. Serum IgG4 level and immunohistochemical (IHC) IgG4/ IgG ratio were also normal, thus

Correspondence to : Dr. Prasenjit Das, M.D., D.N.B., M.N.A.M.S., Assistant Professor, Department of Pathology, All India Institute of Medical Sciences, Ansari Nagar, New Delhi-110029.
E-mail : prasenaiims@gmail.com
dr_prasenpgi@yahoo.co.in

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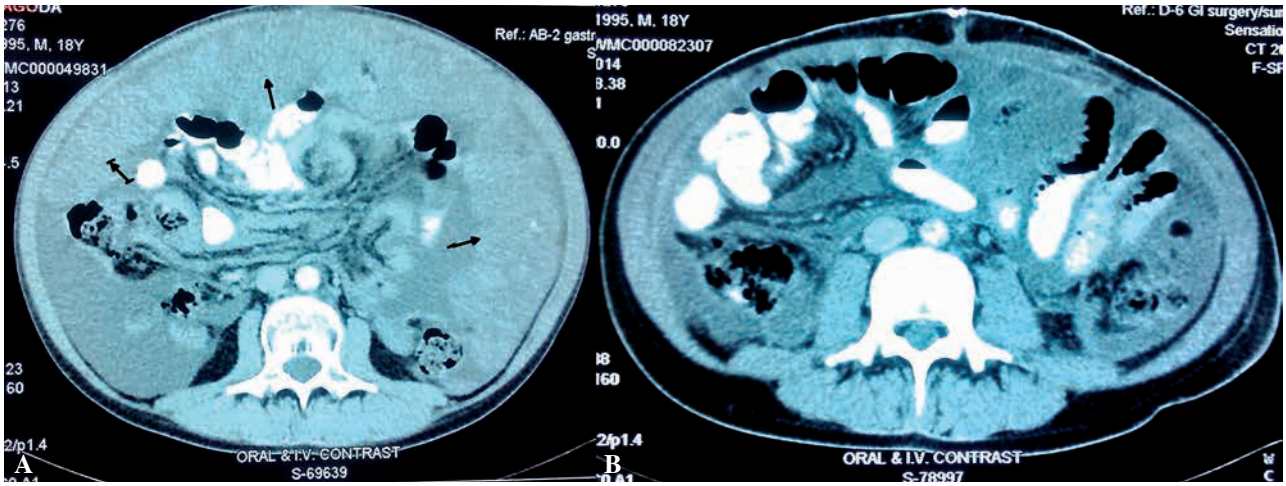

Fig. 1. – A. Axial contrast enhanced CT scan showing nodular omental thickening (arrows) with ascites. B. Axial contrast enhanced CT scan after treatment (surgery and chemotherapy) shows mild ascites with complete resolution of the soft tissue mass.

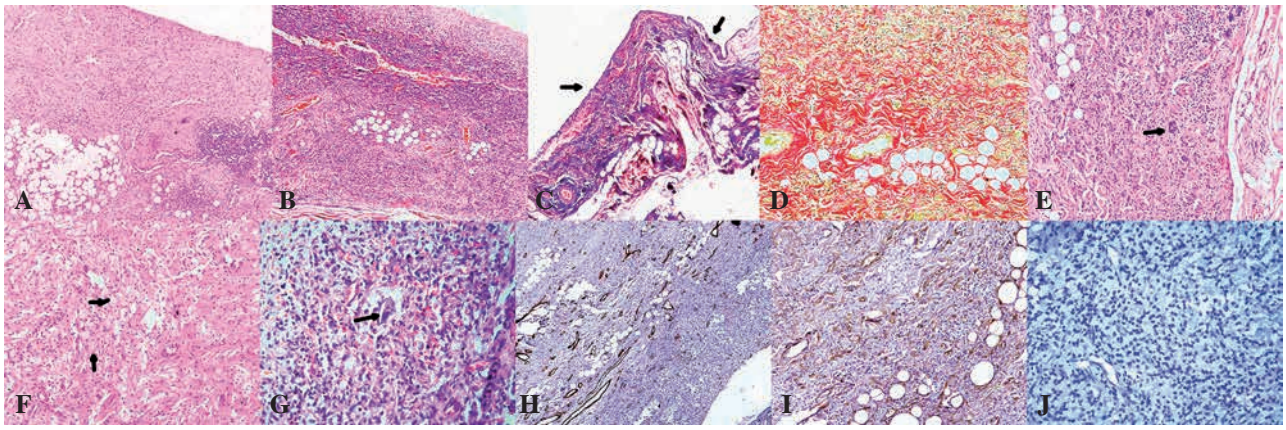

Fig. 2. – Photomicrographs show marked fibrosis and hyalinization of the omental fat, more towards the surface [Figs. 2A & B, H&E ×40]. Masson's trichrome and Sirius stains show deposition of collagen fibers (arrows) [Figs. 2C & D, C (MT) ×40 ; D (SR) ×100]. Many Touton type of giant cells (arrows) [Fig. 2E, H&E ×100], with macrophages containing fat droplets (arrows) [Fig. 2F, H&E ×100] and cholesterol crystals are seen within giant cells (arrows) [Figs. 2G, H&E ×100]. The spindle cells are occasionally positive for smooth muscle actin and CD34 stains [Figs. 2H & I, H (SMA) ×100 ; I (CD34) ×100]. IgG4 stain is negative [Fig. 2J, (IgG4) ×100].

excluding an IgG4 disease. IHC stains for CD20, CD3, CD4, CD8, ER, PR, CD34, and ALK-1 all were negative, while there was diffuse positivity for vimentin and focal positivity for smooth muscle actin and CD34 stains (Fig. 2). Based on overall features, a diagnosis of SMP/ limited Weber Christian disease was suggested. He was started on oral prednisolone, 40 mg OD (once daily) since 2012, tapered to 30 mg OD after a month. However, 15 days after dose adjustment, ascites and abdominal distension recurred, requiring 6 L/week ascitic fluid aspiration. This was followed by tablet Azathioprine, 75 mg OD ; cap thalidomide, 100 mg BD ; tab folic acid, 5 mg OD ; tabs spironolactone, 50 mg+ furosemide, 20 mg OD ; tab calcium carbonate with Vitamin D3 supplementation OD and a short trial of tab colchicine, 100 mg OD, which were continued for another 2-3 months. However, whenever withdrawn, symptomatic abdominal distension returned. The repeat laparoscopic biopsy did not reveal any different pathology. Thereafter, he was started on pulse cyclophosphamide therapy, 600 mg IV, followed by reducing abdominal girth and less requirement for ascetic fluid tap for another 8 months (1 tap/month). Repeat USG though showed minimal ascites, and a persistent omental mass, measuring 6 × 6 cm, which was followed by omental/ mesenteric debulking in 2014. Histological examination of the markedly thickened, multi-nodular and discolored specimen showed similar features as described. Post-surgery, the abdominal girth reduced, with radiological resolution of the omental mass and significant symptomatic improvement (Fig. 1B). He is on regular follow up and didn't require any ascitic fluid tap since last one year. A written informed consent was taken from the patient during management and for preparing the manuscript separately.

Discussion

The term 'SMP' was coined by *Odgen et al*. in 1960 (6). Though the etiology is elusive, trauma, neoplasm, previous surgery, cold, drugs, vitamin deficiency, allergic diseases, abdominal tuberculosis and autoimmune diseases have been attributed to its pathogenesis (7,8). In contrast to conventional belief, the index case was noted in an adolescent and the biochemical findings were suggestive of an autoimmune etiology. The index case presented to us with abdominal distension, due to omental cake formation, though overall, in young patients' symptoms are less evident. Classically, on CT scan, SMP shows thickened mesentery with diffuse, nodular or multinodular enhancement and fat ring sign, reflecting the presence of fat around the mesenteric vessels. However, in the index case, radiology was nonspecific and a diagnosis of peritoneal carcinomatosis was suggested. On HPE, he was diagnosed to have nodular a SMP, after much diagnostic dilemma and excluding possibilities of mesenteric carcinomatosis, pancreatic adenocarcinoma, lymphoma, pancreatitis, inflammatory bowel disease, retroperitoneal fibrosis and IgG4 related diseases (7,9). SMP can be self-resolving, or may respond to immunosuppressive drugs, or may often show chemoresistance, like the index case. In around 20% cases, partial omentectomy or mesenterectomy, may bring down morbidity. Treatment hence should be individualized.

Conclusion

SMP is a rare entity, with variable clinical, radiological and histological findings, which may lead to diagnostic dilemma. It's a diagnosis of exclusion, and histology may serve as the gold standard. Treatment refractoriness is common and should be individualized with emphasis on close long term follow up. Surgical debulking may reduce morbidity in a resistant case.

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