Sclerosing mesenteritis presenting as acute abdominal emergency

Sklerozirajući mezenteritis koji se prezentovao kao akutno stanje abdomena

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ABSTRACT

Sclerosing mesenteritis (SM) is a rare and poorly understood chronic benign inflammatory condition of the mesentery that sometimes can be manifested as acute abdominal emergency. In this paper sclerosing mesenteritis was discussed on the occasion of experience in the diagnosis and the treatment of patients in clinical practice, a 62-year-old man who was diagnosed SM on computed tomography (CT) which was confirmed histopathologically. SM manifests grossly as a diffuse, localized, or multinodular thickening of the mesentery. Microscopically, according to the pattern of the disease, SM can be divided into sclerosing panniculitis where necrosis and inflammation predominate and retractile mesenteritis in which fibrosis and retraction are hallmarks of the disease. Clinical presentation of patients suffering from SM is quite diverse – from asymptomatic, which is the most common, to abdominal pain, bloating, nausea, vomiting and febrile episodes. The 'fat ring sign' and peripheral pseudocapsule are virtually pathognomonic CT findings of SM. Management of SM is empirical with no guidelines yet established. SM should be listed in the differential diagnosis in every older patient with diverse abdominal complaints.

Key words: panniculitis, peritoneal; abdomen, acute; tomography, X-ray computed.

SAŽETAK

Sklerozirajući mezenteritis (SM) je retko hronično benigno oboljenje mezenterijuma čija je etiologija nedovoljno razjašnjena i koje se može manifestovati kao akutno stanje abdomena. U ovom radu sklerozirajući mezenteritis je razmatran povodom iskustva u dijagnozi i lečenju bolesnika iz kliničke prakse 62-godišnjeg muškarca kome je dijagnoza SM postavljena na kompjuterizovanoj tomografiji (CT) potvrđena patohistološki. SM se makroskopski manifestuje kao difuzno, lokalizovano ili multinodularno zadebljanje mezenterijuma. Na mikroskopskom nivou, u zavisnosti od oblika oboljenja, SM se može podeliti na sklerozirajući pankulitis kada su nekroza i inflamacija predominantni i retraktilni mezenteritis kada su fibroza i retrakcija vodeće osobine bolesti. Klinička slika pacijenata obolelih od SM je veoma raznolika – od asimptomatskih oblika, što je najčešće, do bola u trbuhu, nadimanja, mučnine, povraćanja i febrilnih epizoda. "Znak prstena od masti" i periferna pseudokapsula su praktično patognomoničan CT nalaz za SM. Terapija obolelih od SM je empirijska bez još uvek utvrđenih vodiča kliničke prakse. SM bi diferencijalno dijagnostički trebalo razmotriti u svakog starijeg pacijenta sa raznolikim abdominalnim smetnjama.

Ključne reči: pankulitis, peritonealni; abdomen, akutni; tomografija, X-zracima kompjuterizovana.

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INTRODUCTION

Sclerosing mesenteritis (SM) is a rare and poorly understood chronic benign inflammatory condition of the mesentery, first described by Jura in 1924¹. However, sometimes it can be manifested as acute abdominal emergency.

A 62-year-old man was admitted to the Emergency ward due to severe acute abdominal pain. On admission, he was pale and prostrated with abdominal tenderness, along with slight leukocytosis. Since, plain radiography and ultrasound of the abdomen were unremarkable, abdominal computed tomography (CT) was performed. A mass-like, hyperatten-

uated small bowel mesentery (Fig. 1A and 1B), with multiple nodes surrounded by a fat halo (Fig. 1A and 1B, arrow) and a peripheral pseudocapsule (Fig. 1A and 1B, arrowhead) were shown. Additionally, slight distension of small bowel loops was depicted (Fig. 2, arrow). Since, the symptoms were pronounced and prolonged, surgery was indicated. Biopsy was performed during the surgical exploration and the diagnosis of sclerosing mesenteritis was made on histopathological examination. However, surgery revealed no obvious abnormalities of the small bowel loops. The patient's recovery was uneventful and he was discharged in good condition with prescribed immunosuppressive treatment. Nonetheless, the patient was lost to follow-up.

Figure 1. Axial (A) and sagittal (B) CT images show a mass-like, hyperattenuated small bowel mesentery. Note the multiple nodes surrounded by a fat halo (arrows) and a peripheral pseudocapsule (arrowhead)

Figure 2. Coronal CT image shows some slightly dilated small bowel loops (arrow)

DISCUSSION

Mesentery, usually refers to the small bowel mesentery, is a broad fan-shaped fold of peritoneum connecting the loops of jejunum and ileum to the posterior abdominal wall, extending obliquely from the duodenojejunal flexure to the ileocaecal valve. It is consisted of blood vessels, nerves and lymphatics that supply the intestine².

SM manifests grossly as a diffuse, localized, or multinodular thickening of the mesentery. Microscopically, according to the pattern of the disease, SM can be divided into sclerosing panniculitis where necrosis and inflammation predominate and retractile mesenteritis in which fibrosis and retraction are hallmarks of the disease. Inflammatory cells found in the infiltrated region are mostly myofibroblasts and foamy macrophages³. Not only small bowel mesentery, but also transverse and sigmoid mesocolon^{4,5} can be affected. Moreover, Scudiere et al. published the 6 cases of SM involving pancreas mimicking pancreatic neoplasm⁶. SM, along with retroperitoneal fibrosis, sclerosing cholangitis, Riedel's thyroiditis and orbital pseudotumor, compose the heterogeneous group of idiopathic fibrosclerotic disorders and pan-inflammatory pseudoneoplasms which unifying feature is mass effect on surrounding structures leading to respective symptoms⁷. Akram et al. found that about 40% cases of SM had history of intra-abdominal surgery⁸. Although benign, SM is commonly associated with malignancies such as lymphoma, lung cancer, gastric cancer, melanoma and carcinoid. The pathogenic mechanism linking these entities is unknown. However, autoimmune reactions or coincidental findings have been postulated⁷.

Clinical presentation of patients suffering from SM is quite diverse - from asymptomatic, which is the most common, to abdominal pain, bloating, nausea, vomiting and febrile episodes. In asymptomatic patients, SM is usually diagnosed by CT as an incidental finding in elderly men in 6th and 7th decade of their lives. Pain is the most common clinical manifestation of SM and usually is due to inflammation in the mesentery or ischemia secondary to compression of the mesenteric blood vessels⁹. Transitory ischemia could be the reason of slightly dilated small bowel loops on CT examination and pronounced abdominal pain in our patient. Rarely, SM can compress bowel leading to intestinal obstruction that could be recurrent¹⁰.

CT findings suggestive of SM are soft tissue density in the root of mesentery, calcifications, preservation of normal fat around the involved vessels and nodes (the so-called 'fat ring sign'), as well as a peripheral pseudocapsule. Mesenteric edema, lymphoma, desmoid, liposarcoma, carcinoid and carcinomatosis should be included in the differential

diagnosis. Although, the 'fat ring sign' and peripheral pseudocapsule are virtually pathognomonic for SM, the definite diagnosis could only be established histopathologically⁷.

Management of SM is empirical with no guidelines yet established. Treatment depends on the stage of the disease: in the early stages, when fat necrosis predominates no treatment is needed since it usually settles spontaneously; in advanced cases when inflammation is the leading manifestation the treatment with corticosteroids, azathioprine and colchicine is advisable. Surgery is reserved for complications such as intestinal obstruction⁹. Unfortunately, we had no opportunity to check the long-term success of the treatment of SM in the present case.

Although benign, sclerosing mesenteritis is the disease that can lead to serious implications if left unrecognized. Therefore, it should be listed in the differential diagnosis in every older patient with diverse abdominal complaints.

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