Mesenteric Panniculitis – A Rare Diagnosis

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Abstract: Mesenteric panniculitis (MP, also known as sclerosing mesenteritis, mesenterial lipodystrophy, retractile mesenteritis) is a rare immune-mediated inflammatory and fibrosing condition with unknown etiology that affects the mesenteric lipocytes. MP is characterized by degeneration and necrosis of the fat tissue, chronic inflammation and the development of fibrosis within the mesenterium. The most common clinical symptoms are abdominal discomfort and pain, nausea and vomiting, palpable tumor formation in the abdomen, weight loss, fever, and symptoms of bowel obstruction. Some patients are asymptomatic and MP is detected incidentally during imaging studies for other reasons. We present a 62-years-old male patient with tumor formation in the abdomen discovered during ultrasound examination for other reasons and diagnosed on magnetic-resonance imaging as mesenteric panniculitis and discuss the etiopathogenesis, diagnosis and treatment of this rare disease.

Keywords: mesenteric panniculitis, mesenteric lipodystrophy, rare disease, abdominal ultrasound, magnetic-resonance imaging.

Abbreviations: CRP = C-reactive protein, CT = computed tomography, ESR = erythrocyte sedimentation rate, IgG = immunoglobulin G, MP = mesenteric panniculitis, MRI = magnetic resonance imaging.

1. INTRODUCTION

Mesenteric panniculitis (MP) is a rare chronic immune-mediated inflammatory disorder characterized by the development of inflammation, necrosis and subsequent fibrosis in the mesenteric fat tissue surrounding the intestines [1,2,3]. MP was first described in 1924 by V Jura as retractile mesenteritis and in the 1960s WW Odgen named it mesenteric panniculitis. In 1996 RE Mindelzum described the characteristic misty mesenterium image on computed tomography (CT) investigation [4].

MP is also known as sclerosing mesenteritis/ mesenteric sclerosis, mesenterial lipodystrophy, retractile mesenteritis, liposcleroticmesenteritis, mesenteric involvement in Weber-Christian disease, and mesenteric lipogranuloma, mesenteric involvement in Weber-Cristian disease. [1,4,5].It is characterized by three types of pathological changes: chronic non-specific inflammation, fat necrosis and fibrosis. When the histological changes are dominated by inflammatory and necrotic lesions, the term mesenteric panniculitis is applied, and when marked fibrosis is present, the term retractile mesenteritis is more accurate [1]. On the other hand, different severity of fibrosis may be present, and therefore some authors suggest that the term sclerosing mesenteritis is more precise [6]. The development of MP has been associated with different provoking factors, such as abdominal trauma or surgery, ischemia, autoimmune diseases and immune deficiencies, the intake of certain medications, allergies, neoplasms, pancreatitis, etc.[1,6,7,8,9,10].MP is presumed to be immune-mediated disorder developing as progressive chronic inflammation with fibrogenesis, accompanied by systemic (constitutional) symptoms, such as fever and fatigue. Family history of autoimmune disorders is frequently present. Histologically, MP presents with areas of inflammation and necrosis within the mesenteric fat tissue, more frequently around the small intestine and less frequently around the colon. The clinical manifestations are non-specific, mild and often even absent. MP usually has good prognosis and is self-limiting, except for the cases with underlying neoplasm[1,7]. The laboratory findings are non-specific and reveal inflammation – leukocytosis, increased erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) levels. The disease is
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diagnosed with imaging methods and often histological verification is needed. The most important differential diagnosis is the presence of intra abdominal tumor. The treatment consists in surgical removal of the inflammatory infiltrates, anti-inflammatory and anti-fibrotic agents.

We present a case of mesenteric panniculitis detected on ultrasound examination for other reason and verified by magnetic resonance imaging (MRI).

2. CLINICAL CASE PRESENTATION

We present a 62-years-old male patient admitted of the Clinic of Nephrology for diagnostic evaluation of increased serum creatinine (to 162 mcmol/l) and uric acid levels (to 452 mcmol/l). The patient had long-standing history of gout, on treatment with Colchicine 0.5 mg bid. At the admission the patient had acute gout arthritis of the right knee. The clinical-laboratory investigations demonstrated high total cholesterol of 6.15mmol/l, high uric acid of 452 mcmol/l and borderline serum creatinine of 107 mcmol/l. The routine abdominal ultrasound examination revealed cholelithiasis and formation 76/61 mm above and in front of the left kidney (figure 1). Both kidneys had normal ultrasound appearance.

![Figure1. Abdominal ultrasound examination revealing a hypoechogenic formation above and in front of the left kidney.](image1)

The patient was referred for MRI and uncomplicated cholelithiasis with thickening of the mesenteric fat tissue and round areas of decreased signal intensity were demonstrated (figure 2), suggesting the presence of mesenteric panniculitis. Therefore, mesenteric panniculitis, most probably associated with underlying asymptomatic cholelithiasis, was diagnosed and the patient was referred to abdominal surgery for biopsy from the described lesion. Colchicine treatment was continued.

![Figure2. MRI of the abdomen, revealing the fat tissue in the mesenteric root with areas of reticular pattern of decreased signal intensity in T1 and T2 (arrows).](image2)

3. DISCUSSION

MP is a rare fibro-inflammatory disorder with unknown etiology and benign disease evolution, characterized by the development of inflammatory infiltrates within the mesenterium of the small and large intestine, subsequent fat necroses and fibrosis.

A small number of MP patients have been described in the literature (probably, less than 500)[1]. MP develops more frequently in Caucasians, showing male predominance (male: female ratio = 2:3: 1) and its incidence increases with age[1,7,8,9]. It is extremely rare in children, probably because of the lower volume of the mesenteric fat tissue in this age group.
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MP probably develops as inflammatory changes with necrosis and subsequent fibrogenesis within the mesenteric fat tissue in response to different stimuli, such as trauma/surgery, systemic or localized intra abdominal infection, mesenteric ischemia or abdominal tumor. Abdominal trauma or surgery precede MP in the majority of cases [1]. Intake of medications (beta-blockers, anti-arrhythmics, etc.), peritonitis, pancreatitis, cholelithiasis/ cholecystitis, urine leakage, autoimmune and fibrosing conditions, and tobacco smoking have been described in association with MP [1,10,11]. Some of the patients have underlying neoplastic disease: solid (lung, small or large intestinal tumor, renal and gastric cancer, melanoma) or hematological neoplasm (lymphoma or leukemia) [1,8,9,10,11]. In the majority of cases the pathological changes (different degree of inflammation, necrosis and fibrosis) involve the mesenterium of the small intestine (in >90% of the cases) and less frequently – the large-intestinal mesenterium. Peripancreatic, mesocolic and retroperitoneal, omental and pelvic involvement have also been described [1]. Therefore, systemic fibroinflammatory condition, resembling retroperitoneal fibrosis, cannot be excluded [1].

MP is presumed to be an IgG4-related disease, developing as a pathological immune response against the structurally changed mesenteric fat cells or interstitial structures (probably, like in idiopathic retroperitoneal fibrosis in which the autoimmune response is suspected to be directed against ceroid, and in other systemic sclerosing conditions), or as a late immune-mediated consequence of chronic infections, ischemia or trauma to the mesenteric structures, tumors in the mesenteric area, etc., leading to autoimmune aggression with inflammatory, necrotic and fibrotic changes [12].

Three histological stages have been described [1]:

- Inflammatory – foam macrophages with minimal inflammatory infiltrates within the mesenteric fat tissue. At this stage clinical symptoms are usually absent.

- Appearance of mesenteric infiltrates consisting of plasma cells, polymorphonuclear leukocytes, giant cells (“foreign body” type) and persistence of foam cells within the mesenteric fat tissue. Lipocytic necroses may be detected. At this stage usually non-specific clinical signs are present – abdominal pain, fever, fatigue, obstructive symptoms.

- Development of fibrous infiltrates and symptoms of bowel obstruction (pain, palpable abdominal formation).

This clinico-histological staging is not definitive, because usually all three processes (inflammation, fat necrosis and fat necrosis) may be observed in one biopsy.

The clinical-laboratory investigations usually reveal non-specific inflammatory changes – increased leukocyte count, ESR and CRP levels, and increased acute-phase markers.

The clinical manifestations of the disease are non-specific: fatigue, malaise, fever, decreased appetite, weight loss, abdominal pain, palpable abdominal formation. Rarely, in underlying intra abdominal neoplasms, bleeding, jaundice, bowel obstruction/perforation can be observed [1,2,5].

The diagnosis is based on imaging studies – abdominal CT and MRI, revealing the characteristic mesenteric involvement, and on histopathological investigations [1,4]. Both imaging and histological studies allow differential diagnosis with intra abdominal neoplasms. Abdominal ultrasound may reveal intra abdominal formation that requires further investigations. The CT and MRI show different degree of “maturity” of the inflammatory and fibrous infiltrates and allow detection of other inflammatory or neoplastic processes.

The differential diagnosis should be made with infiltrative abdominal processes, affecting themesenteric structures – carcinoma, lymphoma, lymphosarcoma, carcinoid and desmoids tumors, mesothelioma, mesenteric metastases, infections (histoplasmosis, tuberculosis, Whipple’s disease), amyloidosis, foreign bodiesm inflammatory reaction against tumors and intra abdominal inflammatory collections – abscessus and phlegmona.

The natural history of MP, when unassociated with underlying neoplasms, is slow and usually benign. The disease can undergo spontaneous resolution or lead to advanced fibrosis with bowel obstruction. The diagnosis is usually made 2-11 years after the first disease symptoms [1] due to the non-specific character of the clinical and laboratory manifestations.

The treatment is aimed at detection and elimination of the underlying condition (inflammation or tumor within the abdomen), anti-inflammatory (corticosteroids, thalidomide, cyclophosphamide, azathioprine) and anti-fibrotic medications (colchicines, tamoxifen)
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[1]. Different antifibrotic strategies have been tried, including gestagens, surgical excision, radiation therapy, have been tried, but have been abandoned due to further increase of fibrosis.

In our patient, the disease was detected during routine ultrasound investigation for other reason, without abdominal pain or palpable tumor formation, and was verified by MRI. Underlying non-complicated cholelithiasis was also detected. The cholelithiasis probably has preceded MP by many years and could have lead to its development due to asymptomatic persistent inflammation caused by the gall bladder stones. No intra abdominal neoplasm was detected. The concomitant colchicin treatment for gout is beneficial for MP and was therefore continued.

In conclusion, mesenteric panniculitis is a rare fibro-inflammatory disorder involving mesenteric fat tissue, developing in response to other inflammatory, ischemic or neoplastic intra abdominal disease, or as a part of a systemic fibro-inflammatory disease. MP is frequently detected during routine imaging studies for other reason, or due to constitutional symptoms, bowel obstruction or palpable abdominal formation. The evolution of the disease is usually beneficial and spontaneous remissions have been described. Currently, there is no standard therapy and the treatment is aimed at elimination of the underlying conditions (ischemia, cholelithiasis, medications, infections, etc.) and suppression of inflammation and fibrogenesis. The presented patient demonstrates a rare case of mesenteric panniculitis associated most probably with asymptomatic cholelithiasis, and underlines the importance of team approach in the diagnosis and treatment of this rare condition.

REFERENCES


