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Abnormal growing of the abdomen due to mesenteric lipodystrophy

Abstract: Mesenteric lipodystrophy is a rare and benign disease characterized by non-specific inflammation of mesenteric fat. Nomenclature of the disease changes according to the histological features and clinical presentation. The etiology is still obscure, and the clinical presentation and laboratory tests are non-specific. In the majority of cases, the diagnosis is primarily based on computerized tomography (CT) features. There is no specific treatment available for mesenteric lipodystrophy. Radical surgical treatment is not advised. We report a case of distended abdomen caused by mesenteric lipodystrophy along with ultrasonography (US), CT and magnetic resonance (MR) findings and a review of the literature.

Key words: Abdomen, mesenteric lipodystrophy, sclerosing mesenteritis

Introduction

Mesenteric lipodystrophy is an uncommon and benign disorder characterized by non-specific inflammation and infiltration of the mesentery with lipid-laden macrophages and fibrous tissue. It has been reported in the literature by different names, such as retractile mesenteritis, mesenteric panniculitis, mesenteric Weber-Christian disease, fibrosing mesenteritis, and sclerosing mesenteritis according to the stage of disease (1-6). The varying nomenclature for this disease results from the varying descriptions of histological features that may be observed in this apparently single clinical-pathologic entity (1,6). It is more commonly diagnosed than before due to the increased use of abdominal US and CT. It may present with non-specific abdominal pain or a palpable abdominal mass, or it may be an incidental imaging finding in asymptomatic patients. The exact cause of the disease remains unclear (2-5,7,8).

Signs and symptoms may be related to inflammation or to mass effect on adjacent organs, especially due to entrapment of intestinal loops. Symptoms at presentation include abdominal pain and vomiting, and less frequently constipation, anorexia, weight loss, fatigue, fever of unknown origin, ascites, and pleural and pericardial
effusion. The duration of symptoms can be from days to 10 years (1,4,5,7,9,10). In the majority of cases, the disease is diagnosed predominantly on the basis of CT features (2,4,6,8-10). No therapeutic procedure has been established (2).

Case report

A 50-year-old man was admitted to hospital with distention and growing of his abdomen. His past medical history consisted of subtotal thyroidectomy 11 years ago because of toxic multinodular goiter and cholecystectomy 10 years ago because of cholelithiasis. His abdomen had been growing for the past 9 years and misdiagnosed as obesity. He had hypertension but was not receiving treatment for this. He had neither weight loss nor abdominal pain. Altered bowel habits and fever were not noted. His blood pressure was 160/100 mmHg. His physical examination showed an extreme abdominal distention and an impression of a large diffuse intra-abdominal mass. In laboratory tests, TSH level was 6.8 uIU/mL (normal range: 0.27-4.2 uIU/mL), free T3 and free T4 was normal, total cholesterol was 248 mg/dL (normal range: 120-220 mg/dL), triglyceride was 495 mg/dl (normal range: 65-165 mg/dl). In terms of laboratory findings, sub-clinic hypothyroidism, dyslipidemia, and hypertension were diagnosed. The other laboratory tests were normal. Ultra-sonography (US) did not depict ascites, lymphadenopathy, or a mass. However, diffuse enhancement of mesenteric fat was noted and CT was recommended. The subsequent computerized tomography (CT) scan demonstrated a homogeneous, widely distributed, diffuse growing of the mesenteric fat. Its limits could not be well defined but the entire abdomen was filled with it and it was causing diastasis recti. There were no signs of invasiveness such as lymphadenopathy or displacements of the structures or pseudo capsule (Figures 1-3). After further evaluating with magnetic resonance imaging (MR) also, this giant benign enlargement was diagnosed as mesenteric lipodystrophy (Figure 4). Surgery was not suitable for this patient; thus medical treatment was given.

Discussion

In most patients, the mesenteric lipodystrophy was incidentally identified during a CT examination for an unrelated condition. The prevalence of mesenteric lipodystrophy was reported as 0.6% in a series of 7620 CT scans (8). There were few cases of mesenteric lipodystrophy worldwide or in Turkey (1-10). Although Daskalogiannaki et al. (8) reported a female predominance, Emory et al. (1) noted that this rare and benign disease of mesentery is slightly more common in males (M:F ratio 1.9:1). It usually occurs in middle or late adulthood (1,4,5,7-10).

The most common clinical findings are abdominal pain, abdominal mass, and symptoms of obstruction (1). Additional presenting complaints included abdominal distention, diarrhea, constipation, anorexia, weight loss, fatigue, fever of unknown origin, ascites, and pleural and pericardial effusion (1,2). Strikingly, our patient, with such a remarkable growth in the mesentery fat, had neither obstruction symptoms nor the other complaints except abdominal distention.

The lesions were described as a single mass (mostly), multiple masses, and diffuse thickening of the mesentery (1). When mesenteric lipodystrophy manifests as a speculated mesenteric mass with or without calcification, the most important alternative in the differential diagnosis is carcinoid metastasis to the mesentery (6). In this patient, mesenteric fat had spread to the entire abdomen and was diffusely thickened.

Figure 1. Pilot image of CT showing apparent abdominal distention.
The duration of symptoms was reported as 3 weeks to 2 years or days to 10 years (1,8). Herein, progressive growth of the abdomen, which began after the cholecystectomy, means a 10-year duration.

The maximum transverse diameter of mesenteric fatty masses ranged from 7.1 to 15.2 cm (mean 9.5 ± 1.4 cm) in the series reported by Daskalogiannaki et al. (8). Similarly, Emory et al. (1) described single or multiple masses ranging from 1 to 40 cm in size (average 10 cm). As may be seen in Figures 1 to 4, the enlargement was diffuse in this case and therefore its limits could not be measured.

The etiology of mesenteric lipodystrophy remains unknown. The vast majority of cases are thus considered idiopathic. In a small subset of cases, however, the disease may be associated etiologically with trauma, previous surgery, infection, vasculitis, or autoimmune mechanism (1,2). Although generally mesenteric lipodystrophy was reported as a benign condition of unknown origin, some studies marked malignancy, mostly urogenital or gastrointestinal adenocarcinomas or lymphomas or immune suppression as in AIDS or idiopathic inflammatory disorders, as the etiologic factor (2,8). Sub-total thyroidectomy because of toxic multinodular goiter and cholecystectomy 1 year before his complaint may be partly responsible for the etiology of this case but autoimmune conditions were obscure.

A definitive diagnosis of mesenteric lipodystrophy requires a surgical excision biopsy and pathological analysis, but the incidental benign and often asymptomatic nature of mesenteric lipodystrophy does not justify biopsy in all cases (2,3). In the
majority of cases, the disease is diagnosed predominantly on the basis of CT features. In the study by Daskalogiannaki et al. (8) only 4 of the 49 patients had a biopsy proven diagnosis. On CT, the hallmark of mesenteric lipodystrophy is increased density of the mesenteric fatty tissue (approximately -40 to -60 HU) as compared to the attenuation values of normal retroperitoneal or subcutaneous fat (-100 to -160 HU). This hyper-attenuating fat surrounds the mesenteric vessels, but does not displace them. The mesenteric lesion, however, may show some regional mass effect by displacing locally small bowel loops (3,6,8). Because of the apparent diagnosis after US, CT, and MR, surgical excision and histopathological analyses were not carried out in this patient as mentioned in the literature.

No therapeutic procedure has shown unequivocal efficacy, and treatment of the symptomatic form is not well established. Most patients continue to be managed conservatively with follow up CT to monitor for any disease progression or any potential complications (2). The prognosis of mesenteric lipodystrophy is good. In a study by Emory et al. (1), no evidence was found to suggest an association with any syndrome and there were no reports of aggressive growth, recurrence, or regression subsequent to the initial diagnose after following 42 patients for an average of 9.5 years. Once the diagnosis is established, asymptomatic mesenteric lipodystrophy may be left untreated and observed. Some authors reported medical treatment with anti-inflammatory or immunosuppressive agents in highly symptomatic diseases (1,2,5). Surgical treatment should be exclusively attempted when intestinal obstruction or ischemia occur. Most of the time, complete excision of the lesions is not possible (1,9,10). We prescribed drugs for his hypertension and hypothyroidism. Dyslipidemia might be due to hypothyroidism and nothing was prescribed since TSH limits will return to normal levels. Furthermore, consistent with the literature we prescribed colchicines as an anti-inflammatory drug because of disturbing distention, and planned future visits (5). At the first visit, after 1 month, the distention was not bothersome, and he was checked for hypertension and hypothyroidism. A further visit was planned 5 months later to monitor his CT findings.

We presented a very rare case of mesenteric lipodystrophy as a cause of severe abdominal distention. To the best of our knowledge this is the first study to describe such a huge enlargement of mesenteric fat with CT and MR findings.

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References