Sclerosing Mesenteritis: a Diagnostic Challenge

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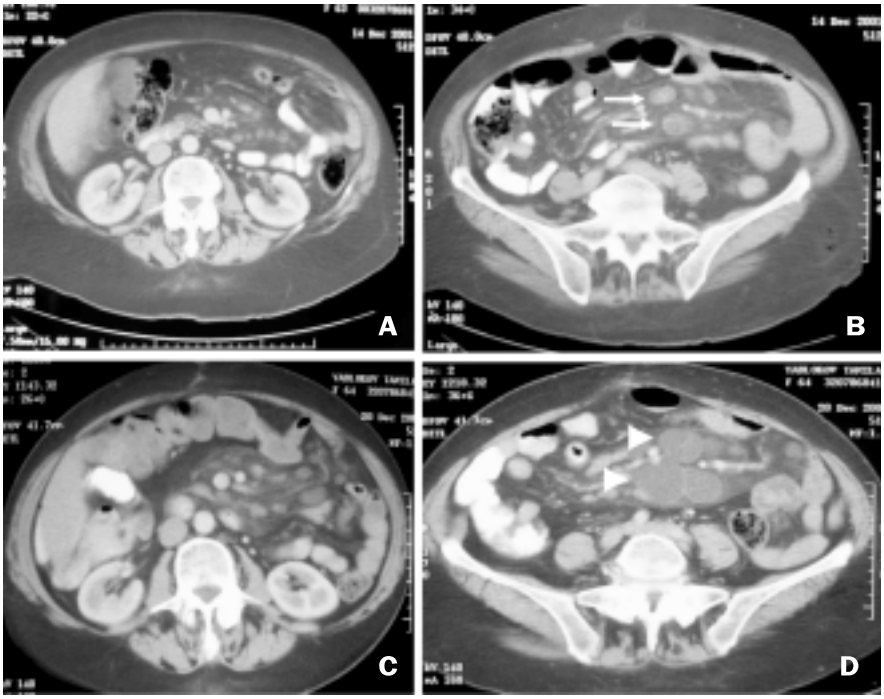
Sclerosing mesenteritis is a rare disorder characterized by a chronic non-specific inflammation involving the adipose tissue of the small bowel mesentery. The cause of the disease is unclear, and autoimmune, infectious, traumatic, and ischemic factors have been reported in the etiopathogenesis [1]. The clinical presentation and laboratory findings are non-specific. A spectrum of findings on abdominal computerized tomography may hint at the diagnosis [1,2] but definite diagnosis requires histologic confirmation [3]. We describe a patient with sclerosing mesenteritis in whom mesenteric lymphadenopathy and large cystic lesions on abdominal CT posed a challenging diagnostic problem.

Patient Description

A 63 year old woman with cholelithiasis was admitted for elective cholecystectomy. On laparatomy, the mesentery was found to be diffusely thickened, with numerous whitish nodes ranging from 0.4 to 2.5 cm in size. The cut section of such nodes was either an elastic-firm yellow tissue or a cyst-like structure containing a milky fluid. Pathologic samples revealed infiltration of the adipose tissue by a predominantly lymphocytic and histiocytic infiltrate surrounding areas of liquefactive necrosis. Fibrotic bands circumscribed lobules of adipose cells. Lymph nodes within the mesenteric fat tissue exhibited features of reactive lymphadenopathy. The patient had no evidence of fat necrosis or cellulitis at other sites, pancreatitis or inflammatory bowel disease. A subsequent CT scan demonstrated soft tissue infiltration of the small bowel mesentery with numerous 1–2 cm diameter low attenuation masses which were thought to be lymph nodes [Figure A]. A few low attenuation masses measured between 8 and 16 Hounsfield units [Figure B]. Tissue cultures for *Mycobacterium tuberculosis* were negative. The diagnosis of lobular sclerosing panniculitis was established.

On follow-up visits the patient still complained of abdominal discomfort. The body temperature was normal. There was no change in bowel habits, nor was there cutaneous eruption, arthralgia or systemic symptoms. Palpation revealed an ill-defined abdominal mass measuring 10 x 18 cm. Results of laboratory examinations, including complete blood count, C-reactive protein, liver and renal function tests, were within the normal range. CT examination demonstrated infiltration and haziness of the mesenteric adipose tissue, containing numerous low attenuation masses [Figure C]. Several round masses, up to 4.5 cm in diameter, with a density corresponding to fluid content (HU 8) were found in the mesentery [Figure D]. A differential diagnosis of infections such as *Mycobacterium tuberculosis* or *M. avium intracellulare,* Whipple's disease and malignancy was considered. In considering such infectious or neoplastic etiology, laparoscopy was performed and biopsies were taken. Histologic examination showed inflammation and areas of fat tissue necrosis. Dilated lymphatic channels were spread amid the inflammatory process. There were no granulomas, vasculitis or malignancy. Excised mesenteric nodes showed non-specific

HU = Hounsfield units



Findings on CT examination performed in **[A and B]** December 2001 and **[C and D]** December 2002. **[A]** Area of increased attenuation within the mesentery comprising numerous hypodense masses. **[B]** Round, fluid-filled cysts, 2.5 cm in diameter (arrows). **[C]** Non-homogeneous fat tissue mass surrounding the mesenteric vessels. The low attenuation lesions are larger than in previous CTs. **[D]** Large, clearly defined, smooth, thin-walled lesions with fluid content (arrow heads).

inflammatory changes. Other nodes were found to have a pseudocapsule with a milky fluid inside. Dilated lymph channels filled with fluid suggested an element of lymphatic obstruction. Tissue Gram stain and Gomori stain were negative. At the time of writing, 3 years after the initial diagnosis of sclerosing mesenteritis, the patient is on symptomatic treatment and is essentially unchanged.

Comment

Sclerosing mesenteritis is part of a spectrum of idiopathic primary inflammatory and fibrotic processes that affect the mesentery. The epidemiology of sclerosing mesenteritis is unknown. An autopsy series reported a prevalence of 1%, suggesting that many cases are undiagnosed during life. A more recent report described a prevalence of 0.6% in over 7,000 abdominal CT examinations [1]. Pathophysiologically, these processes may affect the integrity of the gastrointestinal lumen and mesenteric vessels by a mass effect. These disorders may result in a variety of gastrointestinal and systemic manifestations, including abdominal pain, nausea and vomiting, diarrhea, weight loss and fever. An abdominal mass can be palpated in up to 50% of patients. Masses tend to be deep-seated and poorly defined. Most studies have reported normal laboratory parameters.

The radiologic features are non-specific. Ischemic, neoplastic, infectious and other inflammatory conditions may give rise to similar findings. Ultrasonography may demonstrate well-defined, heterogeneous masses that are predominantly hyperechoic owing to the presence of fat and fibrosis. The most common finding on CT examination is a soft tissue mass in the small bowel mesentery. Mass lesions may be homogeneous or heterogeneous. Two CT findings, "fat ring sign" and "tumor pseudocapsule," are considered to be somewhat specific for sclerosing mesenteritis [2]. The fat ring sign describes the common finding (in 75–90% of patients) of preservation of the densitometric values of fat nearest the mesenteric vessels. A tumoral pseudocapsule (seen in up to 60% of patients) refers to the finding of a hyper-attenuated stripe partly surrounding the mass. These features are not seen in other mesenteric diseases such as lipoma, lymphoma or liposarcoma. The term "misty mesentery" has been used to describe the finding of increased attenuation of mesenteric fat with small lymph nodes but without evidence of a discrete mass. Vascular displacement, encasement or thrombosis may be seen in over one-half of cases. Mesenteric or retroperitoneal lymphadenopathy is present in 20–40% of patients. Cystic components have occasionally been described and may be the consequence of lymphatic or venous obstruction or necrotic liquefaction [4]. In one case, similar to the present patient, sclerosing mesenteritis presented with a multilocular mesenteric mass and cystic spaces corresponding to dilated lymphatic channels [5]. The above features on CT may suggest the diagnosis of sclerosing mesenteritis; however, several other conditions may present a similar appearance. These include lymphoma, carcinoid tumor, carcinomatosis, peritoneal tuberculosis, peritoneal sarcoidosis, primary mesenteric mesothelioma and mesenteric edema. Peritoneal thickening, omental caking, and the presence of ascites with fine mobile septations may suggest the diagnosis of peritoneal tuberculosis. Histologic examination of a sizeable tissue specimen obtained on excisional biopsies is necessary for a definitive diagnosis [2]. CT is the preferred method for periodic assessment on follow-up.

This case demonstrates the diagnostic challenge in finding multiple cystic and nodular mesenteric lesions in a patient with sclerosing mesenteritis. The contribution and limitations of imaging are illustrated, with a definite diagnosis established only after repeated laparotomy and histologic assessment.

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Capsule

Critical period for learning smells

The mammalian sense of smell discriminates an incredible diversity of odors. In the olfactory bulb, each type of odor receptor links up to unique glomeruli that relay the signals up for higher processing in the brain. Zou et al. have now analyzed the process of refinement using mice in which expression of two specific odorant receptors had been marked. Elimination of odoriferous input during specific times of development stalled an intrinsic process by which the inputs from multiple odorant receptors to a given glomerulus are sorted out to produce a mature glomerulus that responds to only one type of odorant signal. Thus, the experience of smell during a critical period is required for maturation of the odorant interpretation system.

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