

SCLEROSING MESENTERITIS

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Key-word: Mesenteritis

Background: A 55-year-old Caucasian man presented with weight loss, cramping abdominal pains, an increasing abdominal circumference and diarrhea. Physical examination showed no abnormalities besides a puffy abdomen. His past medical history included a recent subcutaneous swelling in the neck, histologically compatible to a benign solitary fibrous tumor. All blood results were within normal limits.

Abdominal ultrasonography showed a tumor with diameter of 6.7 cm, probably originating from the pancreas, with ascites and retroperitoneal lymphadenopathy. This was followed by a CT scan.

CT scan of the abdomen was repeated following therapy.

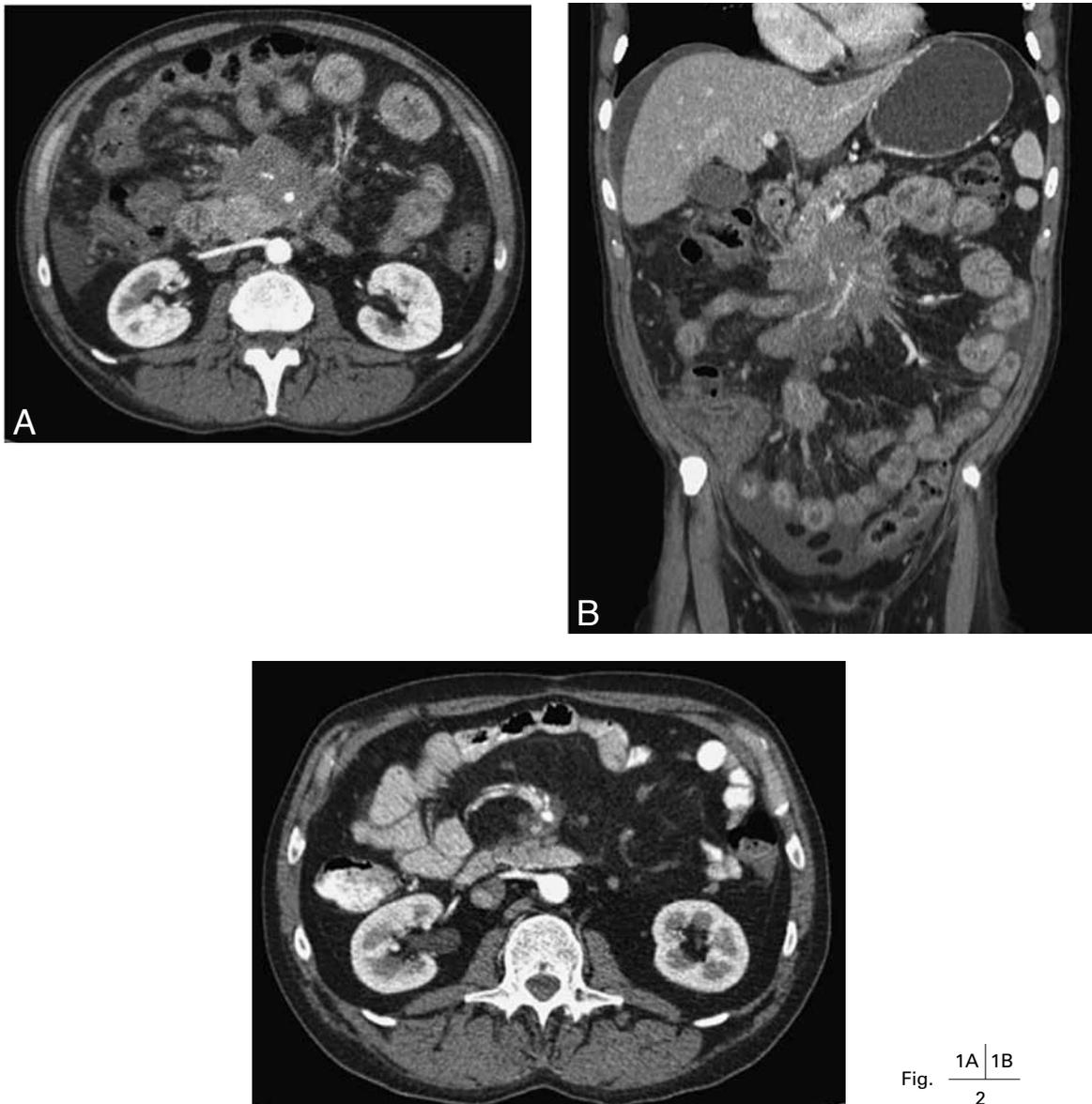


Fig.

1A	1B
2	

Work-up

Contrast-enhanced CT scan of the abdomen prior to treatment (Fig. 1) included a transversal reconstruction (arterial phase) (A) and a coronal reconstruction (portal-venous phase) (B). An ill-defined homogeneous soft tissue mass around the mesentery root with some radiating strands, attached to the pancreas and duodenum, is visible. The encased arteries and veins are stenosed, but not occluded. This results in prestenotic venous distention and probably in ischemia of small bowel loops and the ascending colon, seen as thickening of the walls. Ascites (and secondary pleural fluid) are probably exsudates due to the ischemia. Furthermore there is a band of soft tissue mass prevertebral on level Th4 to Th10 and soft tissue deposits in mesenterium and omentum are seen.

Contrast-enhanced CT scan of the abdomen (arterial phase) after treatment (Fig. 2) shows tumor regression and normalization of the bowel walls.

Radiological diagnosis

The radiological appearance was first thought to be a lymphoma, with mesenteric and posterior mediastinal localization. Because ultrasonographically guided biopsies were not conclusive, a laparotomy was performed subsequently to obtain a histological diagnosis. Histological diagnosis showed extensive inflammation with fibromatosis, without malignant cells. This is compatible with a diagnosis of *sclerosing mesenteritis* localized in the mesentery root. There were no signs of an underlying disease. The patient was treated with corticosteroids during 15 months. Since a follow-up CT scan showed regression of the tumor, the patient uses azathioprine 150 mg once daily and is doing very well.

Discussion

Sclerosing mesenteritis is a very rare disease characterized by fibrolipomatous thickening of the mesentery and is histologically classified into 3 types: mesenteric panniculitis, mesenteric lipodystrophy and fibrosing/retractile mesenteritis. The disease is also known as systemic nodular panniculitis, liposclerotic mesenteritis and xanthogranulomatous mesenteritis. Sclerosing mesenteritis is the most accepted term, used in pathology.

The tumor is locally aggressive but it is considered benign because it does not metastasize. The clinical presentation is nonspecific and may include weight loss, abdominal pain, diarrhea, nausea and vomiting. There is a male predominance and mostly patients in middle or late adulthood are affected.

The classification refers to the stage of the disease and correlates to the dominant tissue and specific radiological findings.

In the first stage, inflammation and fatty degeneration (fat necrosis) predominate (called panniculitis), seen as a misty mesentery or a well-delineated inhomogeneous mass at the root of the mesentery. There is engulfment of superior mesentery vessels with possibly a fat ring sign (preservation of normal

fat around mesenteric vessels, also called fatty halo), but no vascular involvement. A pseudocapsule (peripheral rim enhancement of connective tissue separating normal mesentery from inflammation) and soft tissue nodules may occur.

In the more advanced phase – as in the presented case – a dense fibrous mass in a thickened mesentery can be seen (fibrosing mesenteritis) with linear radiating strands and traction on the abdominal organs (retractile mesenteritis). Stenosis of the vessels and anatomical changes due to traction can cause ischemia or obstruction of the bowel. Also calcifications (in fat necrosis in the central portion of the mass) and multiple nodular masses (fibromas) in the mesentery can be seen.

To date, it is uncertain whether this disease occurs independently or in association with other disorders, such as infection, trauma, ischemia, malignancies or autoimmune disorders. Some small clinical and histological series suggest a possible role for IgG4-related immunopathological processes.

In the presented case, this could connect the sclerosing mesenteritis with the consecutively found prevertebral soft tissue mass, suggestive for fibrosing mediastinitis/mediastinal fibrosis, and the former fibrous tumor in the neck.

The differential diagnosis of sclerosing mesenteritis contains other tumors such as lymphoma, carcinoid tumor, carcinomatosis, desmoid tumor, inflammatory pseudotumor or liposarcoma as well as tuberculosis and mesothelioma. Histological analysis is required for a definitive diagnosis.

Mesenteric panniculitis can be self-limiting. So, in asymptomatic disease a wait-and-see policy is justifiable. Treatment of the more advanced stadia of this disease may consist of colchicine, tamoxifen, glucocorticoids, other immunosuppressiva like cyclophosphamide or azathioprine, or chemotherapeutics. The results of surgical resection are often disappointing due to vessel compromise, but in case of life-threatening complications like bowel obstruction or perforation, surgical resection is the recommended therapy.

Bibliography

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