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Histoplasmosis as a possible cause of retroperitoneal fibrosis and median arcuate ligament syndrome: A case report

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ABSTRACT

INTRODUCTION: Median arcuate ligament syndrome (MALS), a condition of poorly understood etiology, is caused by compression of the celiac artery by fibers of the median arcuate ligament.
PRESENTATION OF CASE: A 46-year-old man with chronic abdominal pain and weight loss was diagnosed with MALS and admitted for surgery. During surgery, extensive retroperitoneal fibrosis around the celiac artery and adjacent aorta was noted. Large necrotizing granulomas and budding yeast, both indicators of histoplasmosis, were found on pathologic evaluation of retroperitoneal tissue removed during surgery.
DISCUSSION: Histoplasma capsulatum may cause pulmonary fibrosis and fibrosing mediastinitis, and the organism may disseminate to reach various internal organs in the immunocompromised individual. Retroperitoneal histoplasmosis has been demonstrated in immunocompromised individuals in the past without retroperitoneal fibrosis. Our patient may have had chronic histoplasma infection of his retroperitoneal lymph nodes, triggering fibrosis in the area surrounding the adjacent celiac artery and abdominal aorta, thus leading to symptomatic MALS.
CONCLUSION: Histoplasma capsulatum, an organism that has demonstrated its capability to cause fibrosis elsewhere in the body, may play a causative role in MALS in our patient, given the finding of retroperitoneal fibrosis in conjunction with retroperitoneal lymph node histoplasma.

1. Introduction

Median arcuate ligament syndrome (MALS),[1] a syndrome of poorly understood etiology, is frequently diagnosed when postprandial abdominal pain, weight loss, and occasionally an abdominal bruit are seen in combination with compression of the celiac artery by fibers of the median arcuate ligament (MAL).[1,2] Variable involvement of the superior mesenteric artery and celiac sympathetic plexus have also been proposed.[3]

2. Presentation of case

A 46-year-old African American man from Texas was admitted for surgery for MALS after an 11-year history of intermittent postprandial abdominal pain and bloating, poor appetite with early satiety, and a minimal weight loss. Before admission, he had computed tomography (CT) and CT angiography of his abdomen, revealing significant narrowing of the origin of the celiac axis that worsened with deep expiration (Fig. 1). Both scans showed no hepatosplenomegaly, lymphadenopathy, or other signs of intraabdominal pathology. In addition, he had a normal gastric emptying study, esophagogastroduodenoscopy, colonoscopy, and chest radiograph. He had a history of hypertension, hyperlipidemia, diabetes mellitus type 1, reflux disease, glaucoma, and minor orthopedic operations. There was no significant work exposure or recent travels. On admission, his blood pressure was 112/65 mmHg, pulse rate 70, respiratory rate 16, pulse oximetry 95% on room air, and temperature 97.0 °F. His physical exam, including an abdominal exam, was normal. Initial lab work was notable for a white cell count 3.4 (10^3 ml^{-1}), normal differential cell count, creatinine 1.23 mg/dl, blood urea nitrogen 18 mg/dl, and glucose 269 mg/dl. His remaining lab results were within normal limits.

He underwent surgery to release the median arcuate ligament and insert an aorto-celiac bypass graft. During surgery, extensive retroperitoneal fibrosis was seen around the celiac artery and the adjacent abdominal aorta, causing MAL compression of the celiac artery. Retroperitoneal tissue was removed for pathological evaluation. Hematoxylin and eosin (H&E) staining of the tissue revealed lymph nodes with numerous necrotizing granulomas (Fig. 2A, B). Within the granulomas, Gomori's methenamine silver (GMS) stain revealed small budding yeast, which is particularly suggestive of histoplasmosis (Fig. 2C). There was no evidence of acid-fast organisms, bacteria, or malignancy. However, DuPont blood cultures

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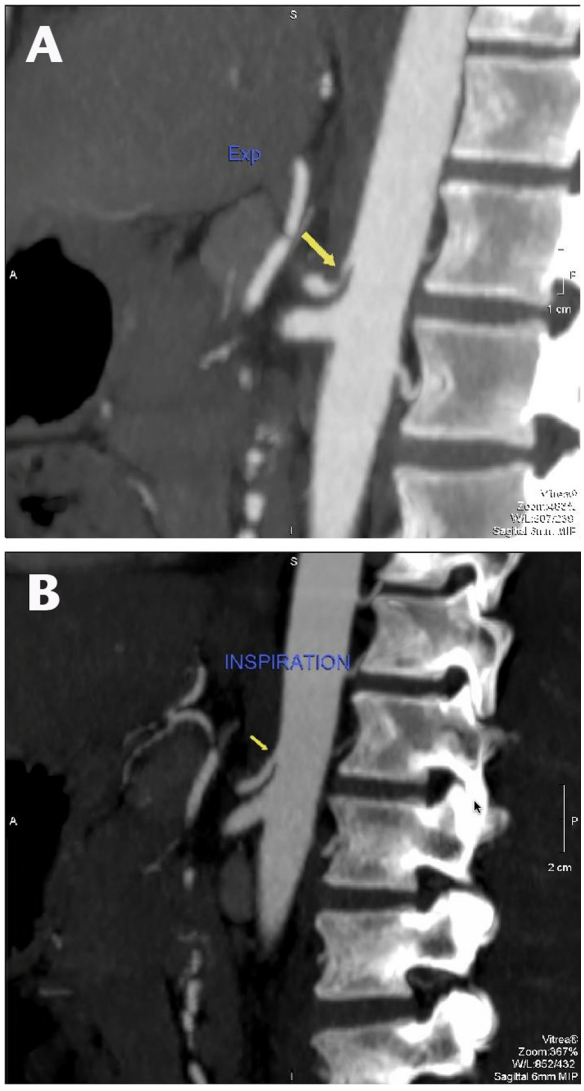

Fig. 1. CT angiography abdomen, sagittal view: compression of the celiac axis. During deep expiration (A), significant narrowing of the origin of the celiac axis is seen, the worst narrowing is located approximately 6.4 mm from its takeoff. During deep inspiration (B), some improvement of the compression is identified.

did not indicate fungemia, and a urine histoplasma antigen test was negative. Voriconazole treatment was started empirically.

3. Discussion

The MAL connects the right and left crura of the diaphragm, forming an arch around the aorta. In some anatomic variants, the MAL may be situated lower than usual and can thus potentially exert external compression on the celiac artery, causing foregut ischemia and possibly a steal syndrome.[4] Good postoperative results after MAL release with celiac sympathectomy suggest that the disease process encompasses periaortic ganglion irritation on the celiac trunk.[3,5]

Histoplasma capsulatum is a dimorphic fungus endemic to the Ohio and Mississippi river valleys. It causes infection in humans when aerosolized spores are inhaled into the lungs. In healthy individuals, the acute infection often has a benign course and may be asymptomatic or commonly mistaken for pneumonia or a simple influenza infection.[6] Histoplasma capsulatum may cause pulmonary fibrosis during chronic infection and fibrosing mediastinitis as an abnormal response to a past histoplasma

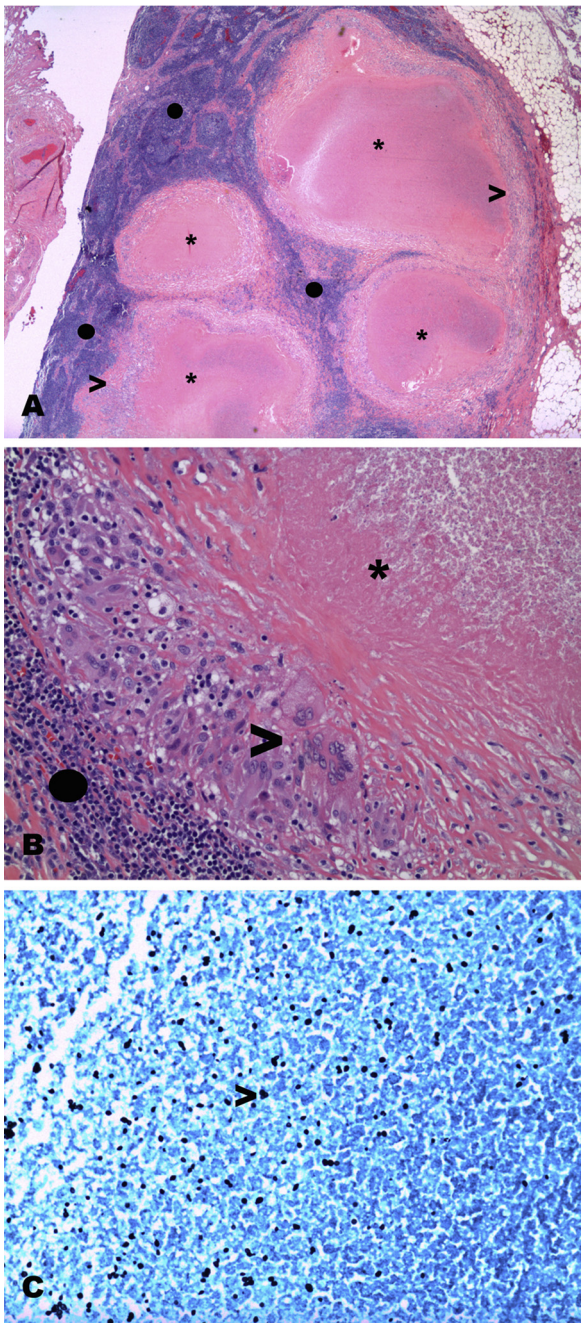

Fig. 2. Lymph Node with Necrotizing Granuloma. The use of H&E staining at 2× (Panel A) and 20× (Panel B) magnification shows macrophages (arrowhead), an area of necrosis (asterisk), and lymphoid tissue (circle). Small budding yeast (arrowhead) was revealed by GMS staining, 40× magnification (Panel C).

infection.[7] Pulmonary histoplasmosis may disseminate via the blood to reach virtually any organ, especially in immunocompromised individuals. Gastrointestinal histoplasmosis has been seen in AIDS patients.[8–10]

Surgery for MALS often reveals a prominent fibrous MAL.[11] A MEDLINE search yielded only three reports of histoplasmosis in retroperitoneal lymph nodes, although all patients had radiographically evident lymphadenopathy, significant comorbidities, and generalized symptoms.[12–14] Our patient never had pneumonia and had no significant travel history or relevant work-related exposures. Histoplasmosis has not previously been described in patients with retroperitoneal fibrosis or MALS. This patient

had large necrotizing granulomas of his retroperitoneal lymph nodes that may have caused extensive fibrosis around the celiac artery and adjacent abdominal aorta, leading to symptomatic MALS.

4. Conclusion

This case underscores the fact that MALS represents not only represents a diagnostic challenge, but also a condition of very poorly understood etiology. We suggest the possibility of histoplasmosis playing a causative role in MALS in our patient, given the finding of retroperitoneal fibrosis in conjunction with retroperitoneal lymph node histoplasma, an organism that has demonstrated it capability to cause fibrosis elsewhere in the body. More case reports are needed to help establish how frequent, if at all, an infectious cause should be seen as a legitimate, and treatable, etiological factor.

Conflicts of interest

None to declare.

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Ethical approval

Written consent has been obtained.

Author contributions

Nicki Niemann contributed to the primary gastroenterology team with Dr. Hochman and drafted case report, chart review, obtained old records, and literature search.

Frank Lyone Hochman contributed to the primary gastroenterology team, critique of draft, and suggestions for improvement.

Richard Sheng Poe Huang analyzed the slides, performed the stains, took pictures, and provided descriptions.

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