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Segmental arterial mediolysis in splanchnic arteries: an uncommon cause of abdominal pain

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Dear Editor,

We have carefully read the article by Mejías-Manzano MLA et al (1) on IgG4-related sclerosing mesenteritis as a rare cause of abdominal pain.

We herein report the case of a 53-year-old male with an unremarkable history who presented with postprandial diffuse abdominal pain of a one-month duration. Physical examination did not reveal any abnormal findings and the laboratory tests were normal. Abdominopelvic computed tomography (CT)/angio-CT showed an increased caliber of the superior mesenteric artery, resulting from eccentric circumferential wall thickening. The patent lumen had a segment with fusiform aneurysmal dilatation, which was 7 x 26 mm long. These vascular changes extended along a number of distal jejunal branches, which also had complete lumen obliteration (Fig. 1). Abdominal arteries were free from signs of arteriosclerotic disease and the findings were consistent with segmental arterial mediolysis (SAM).

Discussion

SAM is an uncommon non-arteriosclerotic, non-inflammatory, non-hereditary arteriopathy (1/100,000 population/year) of uncertain etiology that primarily involves splanchnic arteries. Clinical manifestations range from no symptoms (8%) to an initial presentation as hemodynamic shock (25%) or death (11%). Abdominal pain is the most common symptom (68%). The definitive diagnosis is histological but is difficult to obtain, characterized by vacuolar degeneration in the media layer of the arterial wall. The clinical diagnosis is made by exclusion, based on imaging findings and the absence of inflammation markers such as ANCA, FANA, C4/C3, anti-La Abs and anti-cardiolipin Abs. The differential diagnosis must include collagenopathies (Ehlers-Danlos, Marfan), vasculitis (Takayasu, Behçet) and fibromuscular dysplasia (2). No therapeutic management guidelines are currently available. Initially, conservative management with stringent blood pressure monitoring and regular radiographic follow-up is preferred. Endovascular surgery is the best option if lesions progress. In patients with shock or intra-abdominal bleeding, emergency surgery or an endovascular procedure is indicated (3).

References

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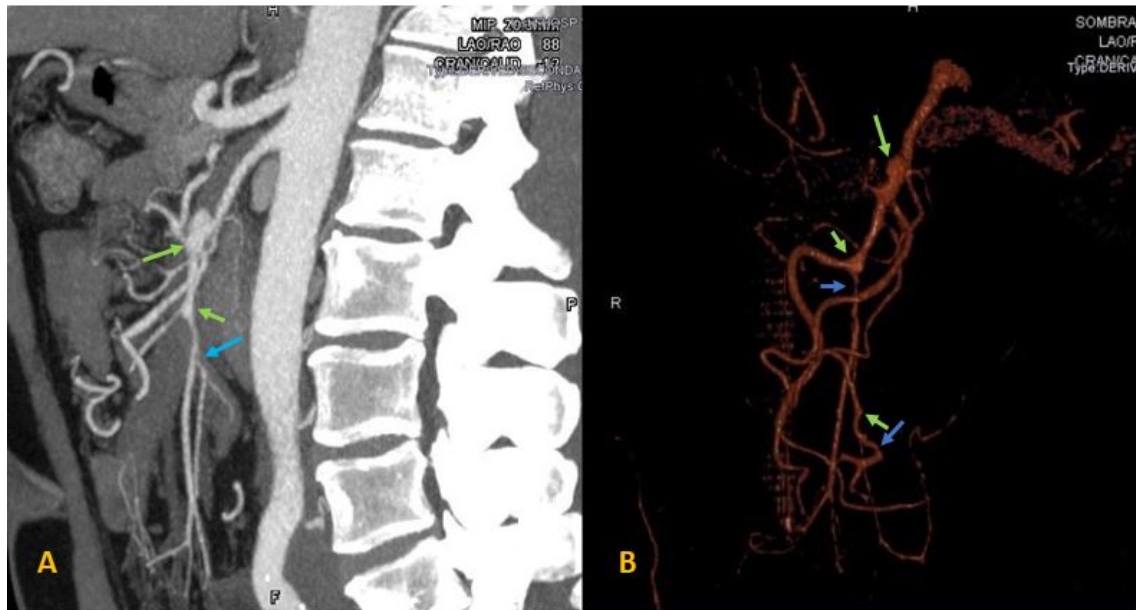


Fig. 1. Abdominopelvic CT/angio-CT reconstructions. Green arrows show the aneurysmal vascular dilatations and blue arrows show the obliterated vascular segments in the superior mesenteric artery and distal branches. The findings were consistent with segmental arterial mediolysis.