Fibromuscular dysplasia with multiple visceral artery involvement

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A 19-year-old female presented with episodic abdominal pain not related to food intake. She had occasional vomiting but had no gastrointestinal bleed. Routine ultrasound examination picked up aneurysm of the superior mesenteric artery (SMA). Computed tomography angiogram showed stenosis of the origin of the SMA and multiple aneurysms involving the proximal SMA. A large collateral artery also showed aneurysm at its origin. The celiac artery and hepatic arteries were not visualized. Splenic artery was reconstituted through gastroduodenal artery. Multiple collaterals from the phrenic and mammary arteries were seen to supply the liver. Inferior mesenteric artery was dilated, and a large marginal artery was seen joining the superior mesenteric artery. Right renal artery showed mild irregularity at the proximal part (Cover).

At surgery the stomach was pale, and no pulsations were felt in the splenic and epiploic vessels. Hepatic artery was thin and felt like a cord. The image in (A) shows the aneurysmal segment of SMA. The aneurysmal segment of the SMA was resected and an aorto-SMA interposition graft with polytetrafluoroethylene was performed. The large collateral artery, which had an aneurysm at the origin, was also resected and reimplemented into the graft. A reversed saphenous vein graft was anastomosed to the graft and to the splenic artery (B). The stomach regained color and good pulsations were felt in the epiploic arteries. Biopsy of the aneurysm sac confirmed fibromuscular dysplasia. Patient had complete symptomatic relief. Postoperative angiogram done 2 months later showed patent grafts with good flow distally. The right renal artery lesion had progressed to cause stenosis (C).

Fibromuscular dysplasia or arterial fibroplasia includes a heterogeneous group of nonatherosclerotic occlusive and aneurysmal disease. Four types are described, but medial fibroplasia is the most commonly seen. It is more common in young women, and the exact etiology is not known. Renal and carotid arteries are commonly involved. Rarely, mesenteric and peripheral arteries are involved. Stenotic type is amenable to balloon dilatation but the aneurysmal one requires resection and replacement with a graft.

REFERENCES


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