Hypoplastic Aortoiliac Syndrome Causing Severe Claudication

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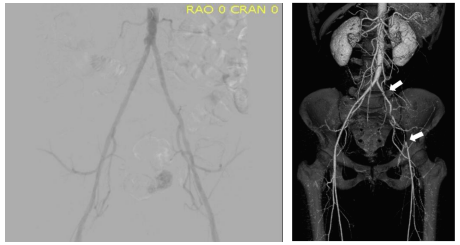
BACKGROUND

Hypoplastic aortoiliac syndrome is a rare condition, occurring almost exclusively in young women of small stature with relatively typical risk factors, including a significant smoking history.

The arteries in HAS may have little if any atherosclerotic changes such as stenosis, kinking or calcification.

Distinct anatomic characteristics of HAS:

- high bifurcation of the abdominal aorta
- straight course of the iliac arteries without the normal characteristic bowing
- acute angle of the aortic bifurcation (20°–30°)
- aortic diameter of 14 mm or less
- iliac artery diameter of 7 mm or less



CTA revealing no atherosclerotic change but showing occlusion of the left external iliac artery and high level of aortic bifurcation with a narrow angle.[4]

SSx: most commonly, intermittent claudication and hypertension.

Dx: Angiography is the gold standard.

Tx: goal - resolve claudication and normalize BP to avoid complications secondary to hypertension.
Treatment of choice remains surgical revascularization; bypass grafts, endarterectomy, and sympathectomy.

Aortofemoral bypass remains the most commonly used grafting technique.

CASE PRESENTATION

48yo F presented to clinic with severe limiting claudication in bilateral lower extremities since 1 year with worsening symptomology with extended periods of walking.

The patient was of small stature with BMI of 21 with significant smoking history of many years. Physical examination revealed 150 cm tall female with systolic BP of 140-170s. Bilateral femoral pulses were poorly palpable.

Laboratory workup were all normal.

Imaging:

Figure 1: CTA: moderate and severe segments of stenosis at infrarenal abdominal aorta with significant involvement at aortic bifurcation. The aorta measured **4mm** at the level of aortic bifurcation and bifurcation was high with a narrow angle.

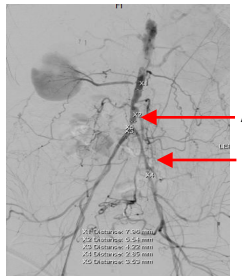


Figure 2: Fluoroscopy showing pathognomonic imaging characteristics of HAS.

Acute angle

Iliacs: Lack of bowing and small caliber

TREATMENT

Operative management:
An intra-abdominal approach was utilized to perform an aorto-iliac bypass grafting with a bifurcating inverted "Y" Gelsoft graft, infrarenal aortic endarterectomy, and right external iliac endarterectomy. An end-to-side anastomosis technique was utilized to overcome size incompatibility between the graft and the native aorta and iliacs.

Postop care:
Postoperatively, the patient was admitted to the ICU for close monitoring and remained hemodynamically stable.

Early postoperative course was complicated with complaints of left leg paresthesias for which CTA was performed and revealed stenosis at the area just distal to the graft-iliac anastomosis on the left. She was taken back the OR and a thrombectomy and balloon angioplasty was performed with relief of the area of stenosis.

She was discharged home on Clopidogrel and Aspirin.

She has since reduced her tobacco habits from 5 cigars/day to 1 cigarette/day.

She has remained free of claudication since her operation.

DISCUSSION

HAS is an uncommon cause of non-atherosclerotic peripheral artery disease. The etiology of HAS remains unclear, however studies suggest that it may be a result of a combination of congenital hypoplasia and atherosclerotic disease.

Most patients are managed surgically with close follow up for long-term patency of the revascularized segments.

Patients should take anti-platelet drugs and be monitored for claudication, ABIs, and CTA.

As the disease process increases the risk for aortoiliac occlusive disease and severe complications of hypertension, it is imperative to identify these patients early and treat them accordingly.

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