



MID AORTIC DYSPLASTIC SYNDROME : A CASE REPORT OF THIS UNCOMMON VASCULAR ANOMALY

Radiodiagnosis

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ABSTRACT

Mid aortic dysplastic syndrome or Coarctation of abdominal aorta is a rare vascular pathology characterised by localized or extended narrowing of abdominal or distal descending thoracic aorta. With hypertension as its cardinal clinical feature , it mostly remains asymptomatic. It could be congenital or acquired associated with obliterative panarteritis , retroperitoneal fibrosis ,etc. Radiologically , narrowing or segmental stenosis of aorta is seen with formation of multiple collaterals.

KEYWORDS

Mid aortic dysplastic syndrome , Coarctation of abdominal aorta , Segmental stenosis

CLINICAL HISTORY

A 19 years old male presented to medicine OPD at New Civil Hospital , Surat with chief complaint of persistent headache .

General examination findings :Pulse : Normal in upper limbs but weak in lower limbs. Blood pressure : 148/98 mm Hg in upper limbs and 98/58 mm Hg in lower limbs. Systemic examination findings :No cardiac murmurs detected on auscultation . No neurological deficit detected.

Laboratory work-up (including Renal function tests) - Normal

As a part of advanced investigation work-up , Contrast enhanced CT –Angiogram of aorta from its origin to its bifurcation was advised . Findings of aortogram were as under:

1. Approx 5 cm long segment of the descending thoracic aorta extending from **D7-D10** vertebral level shows irregular wall thickening with calcifications causing severe luminal narrowing with maximum narrowing noted at D10 level (3.5mm in diameter)
2. Mild narrowing of celiac trunk noted at its origin (3.6mm in diameter)
3. Abdominal aorta appears hypoplastic with progressive luminal narrowing
4. Multiple arterial collateral channels noted in anterior chest wall (from internal mammary and intercostal arteries) and in anterior abdominal wall (from superior and inferior epigastric arteries).
5. Few arterial collateral channels noted from descending thoracic aorta , left hepatic artery , left gastric artery.

DISCUSSION

Mid aortic dysplastic syndrome or *Middle aortic syndrome* or *Coarctation of abdominal aorta* or *Atypical Coarctation* refers to segmental narrowing of descending thoracic aorta and/or abdominal aorta . First description of MAS was given by Schlessinger and Quain. *Hypertension proximal to the aortic stenosis, and relative hypotension distal to it, are characteristic findings in MAS.*

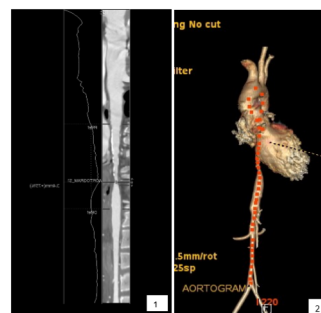
Etiology: **Congenital** - ascribed to a developmental anomaly in the fusion and maturation of the paired embryonic dorsal aortas. **Acquired**- caused by Takayasu's or temporal arteritis (giant cell arteritides), neurofibromatosis, fibromuscular dysplasia, retroperitoneal fibrosis, mucopolysaccharidosis, and the Williams syndrome.

Segmental aortic stenosis may be found at various locations including: Infrarenal, Inter-renal, Diffuse with/without renal artery involvement and with/without post-stenotic aneurysm, Suprarenal. Most frequent location was found to be inter-renal followed by infra-renal.

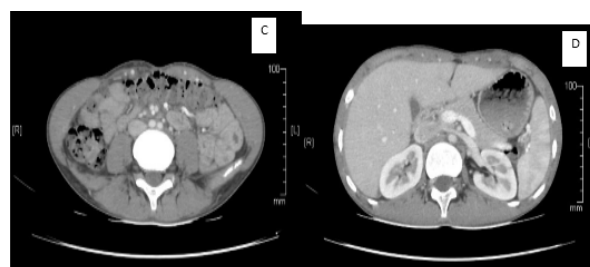
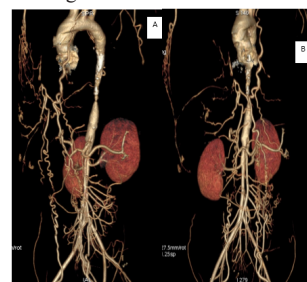
Signs and Symptoms : occur early in childhood and young adulthood. Headache, early fatigue on exertion, and bilateral lower-limb claudication are the common presentations .

It has a high propensity for concomitant stenoses in both the renal (63%) and visceral (33%) arteries.

Diagnosis – done by CT and MR-Angiography
Open surgery is the primary treatment of tubular aortic narrowing (MAS) associated with renovascular hypertension and visceral artery stenosis.



1 & 2 : CT aortogram and volume rendered images respectively showing the stenosed segment of aorta.



A&B : Volume rendered CT-angiography images showing maximally narrowed dorsal aorta at D10 level with narrow celiac trunk at its origin and formation of multiple collateral channels.

C&D :Axial CT images showing multiple arterial collateral channels (arrow) in anterior abdominal wall and from left gastric artery and hepatic artery

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