Midaortic syndrome

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DESCRIPTION

A 55-year-old woman was hospitalised for confusion and generalised weakness. Her medical history included refractory hypertension, coronary artery disease and peripheral artery disease. Examination revealed widespread bruits in the carotid, femoral, subclavian and popliteal arteries and a grade 3/6 systolic aortic murmur. Preliminary investigations showed evidence of renal insufficiency. On the third day of hospitalisation, she developed sudden chest pain with elevated levels of troponin. Coronary angiogram was planned after improvement of her renal parameters. During cardiac catheterisation, there was difficulty in palpating femoral pulses. After prolonged attempt, the right femoral artery was cannulated with ultrasound guidance. The guide wire did not pass above the distal thoracic aorta (figure 1). Aortogram revealed complete occlusion of thoracic aorta with suprarenal stenosis (figure 2 and video 1). Prominent collaterals were noted.

'Midaortic syndrome' (MAS) is a term used to describe the localised narrowing of the distal thoracic/abdominal aorta regardless of aetiology. Both congenital and acquired causes have been described. Congenital narrowing has been thought to occur due to incomplete or overfusion of embryonic dorsal aortas during the fourth week of gestation. Acquired causes include giant cell arteritis, Takayasu arteritis, fibromuscular dysplasia, neurofibromatosis, atherosclerosis, retroperitoneal fibrosis, mucopolysaccharidoses, Williams syndrome and Alagille syndrome.[1] Patients with MAS present predominantly with refractory hypertension and if left untreated develop intermittent claudication of legs, congestive heart failure, renal insufficiency and symptoms of hypertension associated end organ damage.[2] Successful control of blood pressure and preservation of end organ function entail a multidisciplinary approach which combines medical management with catheter based and surgical interventions.[3] In untreated patients, mortality rates as high as 90% are observed by the sixth decade of life due to the effects of end organ damage.[1] This patient had no evidence of vasculitis, neurofibromatosis or the above-mentioned genetic disorders associated with MAS. Given the age and presence of coronary and peripheral artery disease, atherosclerosis is the most likely aetiology for MAS in this patient.

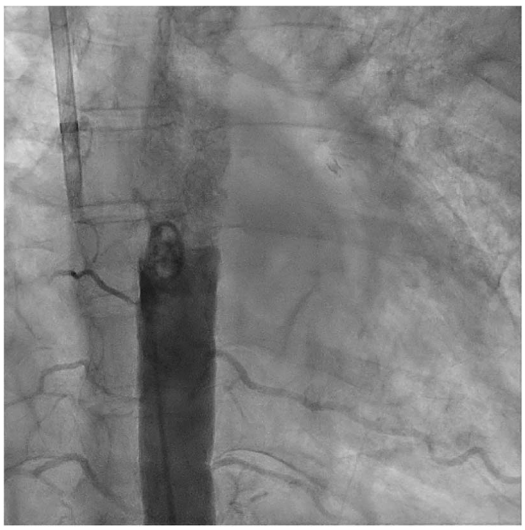


Figure 2 An aortogram revealing complete occlusion of thoracic aorta with extensive collateral vessels.

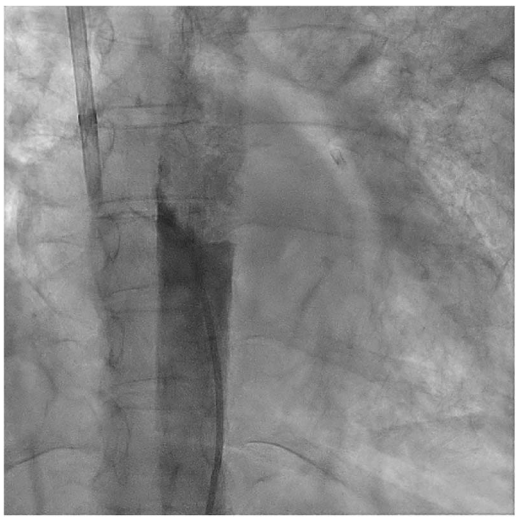
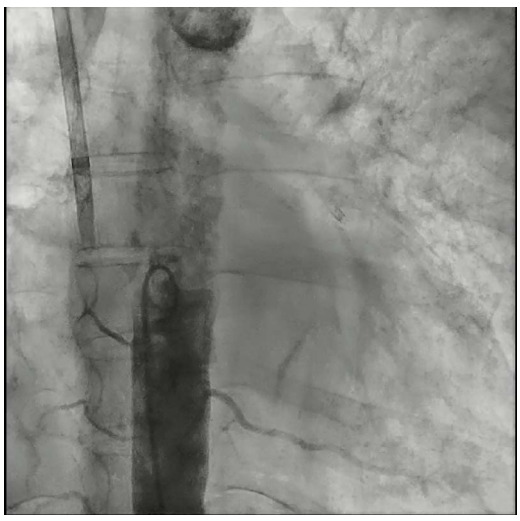


Figure 1 On catheterisation, guiding wire did not pass above the distal thoracic aorta.



Video 1 An aortogram revealing complete occlusion of thoracic aorta with extensive collateral vessels.

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Learning points

- Midaortic syndrome (MAS) refers to the localised narrowing of distal thoracic/abdominal aorta regardless of aetiology.
- MAS can occur due to congenital incomplete or overfusion of embryonic dorsal aortas or secondary to vasculitis, fibromuscular dysplasia, atherosclerosis or retroperitoneal fibrosis. It is also associated with genetic syndromes such as neurofibromatosis, mucopolysaccharidoses, Williams syndrome and Alagille syndrome.
- Untreated MAS has a 90% mortality rate by the sixth decade of life due to end organ damage. This necessitates rigorous blood pressure control and preservation of end organ function through a combination of medical as well as catheter based and surgical interventions.

Contributors KKJ, compiled the case history and investigations, critically reviewed and analysed the data. MK analysed and reviewed the data and prepared the manuscript. DPC and TR helped with editing, formatting and preparing the manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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