CASE REPORT

Mid-aortic syndrome secondary to Takayasu's disease

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Accepted 25 October 2017

SUMMARY

A 24-year-old young woman with good past health presented with hypertension during body check. Blood erythrocyte sedimentary rate level was elevated. CT and MR angiography revealed moderate stenosis of abdominal aorta and bilaterally renal arteries. She was diagnosed with mid-aortic syndrome caused by Takayasu arteritis based on clinical, radiological and pathological findings. She was treated with abdominal aorta resection, graft replacement and aorta-renal bypass. Pathology was compatible with Takayasu arteritis. Postoperatively, her blood pressure was normalised. She was prescribed with long-term prednisolone and aspirin for her arteritis.

BACKGROUND

Mid-aortic syndrome (MAS) is an important clinical entity to recognise and treat accurately. It is a rare cause of uncontrolled young hypertension with poor outcome if left untreated.

A thorough physical examination could assist in early recognition and prevent secondary end organ damage.

CASE PRESENTATION

A 24-year-old young woman with good past health presented with hypertension during body check. Clinically she was asymptomatic. She did not complain of claudication. On physical examination, her brachial blood pressure was elevated, measuring 158/90 mmHg. Bilateral renal bruit and scapula bruit were positive. Radial femoral delay was positive. She is a non-smoker non-drinker.

Her mother has medical history of hypertension.

INVESTIGATIONS

Baseline investigations include chest radiograph and ECG were normal. Serum renin was elevated to 6.85 ng/mL/hour, aldosterone was elevated to 784 pmol/L and erythrocyte sedimentary rate was elevated to 36 mm/hour.

CT and MR angiography showed 50%–75% stenosis of the abdominal aorta at L1–L2 levels (figures 1 and 2). Celiac trunk and origins of bilateral renal arteries were stenotic. There was a saccular aneurysm arising from celiac trunk (figure 3). Origin of superior mesenteric artery was occluded, and distal part was reconstituted by extensive collaterals. Extensive vessels and collaterals are present in the abdominal wall connecting the subclavian artery to the internal mammary, superior and inferior epigastric arteries and ending in the external iliac arteries (pathway of Winslow) (figure 4).

DIFFERENTIAL DIAGNOSIS

- ▶ Congenital MAS
- ▶ Acquired MAS, which can be associated with
  - Neurofibromatosis
  - William syndrome
  - Fibromuscular dysplasia
  - Mucopolysaccharidosis
  - Takayasu arteritis

TREATMENT

The patient was admitted for surgery in view of the celiac artery aneurysm with risk of rupture, stenotic aortic segments potentially leading to critical organ ischaemia and uncontrolled hypertension resulting from renal artery stenosis.

She was treated with resection of abdominal aorta with graft replacement and bilateral aorta-renal bypass. Medically, she was prescribed with long term prednisolone, aspirin and antihypertensive.

OUTCOME AND FOLLOW-UP

Pathology showed intimal and medial fibrosis and granulation tissue with chronic inflammatory cell infiltrates in the aortic wall, consistent with Takayasu arteritis (figure 5).

On follow-up, the patient was in a good state of health, with arterial blood pressure normalised at 128/79 mmHg. Antihypertensives treatment was weaned off gradually. Follow-up CT examination showed patent anastomoses and bypass graft. Clinically she was well and asymptomatic.

DISCUSSION

MAS is an unusual cause of arterial hypertension in the upper extremities. This syndrome may be caused by Takayasu arteritis, which causes stenosis of the large vessels.¹ The initial description of Takayasu arteritis is credited to Mikito Takayasu, a Japanese ophthalmologist, who reported a 21-year-old woman who had sudden visual loss and an anastomotic wreath of vessels around the optic disc.²

Takayasu arteritis is a chronic, idiopathic, inflammatory disease that primarily affects large vessels such as aorta, its major branches and the pulmonary and coronary arteries. Histopathological specimens are seldom available as a result of large vessels commonly affected. The histopathological appearance of Takayasu arteritis can mimic other arteritis, such as giant cell arteritis, and renders the diagnosis difficult. Hence, the diagnosis of Takayasu arteritis is based on the combination of clinical information, laboratory evaluation and diagnostic imaging.³

Our patient presented with a young onset of hypertension at the age of 24 (younger than 40

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To cite: Cheng KCA, Li Y-L.
BMJ Case Rep Published
Online First: [please
include Day Month Year].
doi:10.1136/bcr-2017-
221729

Figure 1 Sagittal maximum-intensity projection reformatted image shows marked narrowing of the abdominal aorta at L1 and L2 levels.

years old), bruit over abdominal aorta and arteriogram abnormality. Therefore, she met the diagnostic criteria of Takayasu arteritis based on the 1990 American College of Rheumatology (ACR) classifications. She was diagnosed of Takayasu arteritis based on the clinical and radiological findings. The ACR diagnostic criteria of Takayasu arteritis which would require at least three of the six criteria to be met. The other three ACR diagnostic criteria would include claudication of extremities, decreased brachial artery pulse and blood pressure difference greater than 10 mmHg.⁴

For assessment of Takayasu arteritis, angiography, particular digital angiography traditionally has been the procedure of choice. However, increasingly CT and MRI angiography used as the diagnostic techniques of choices.² The typical manifestation for Takayasu arteritis is the concentric mural thickening of the involved arteries. Other findings included stenosis, occlusion, ectasis and aneurysm. Abdominal aorta aneurysms would lead to fatal consequence, such as aortic rupture.⁵

Different radiological techniques are also available for better anatomical delineation of the arterial lumen, such as curved planar reformation, multiplanar reconstruction and volume rendered imaging. Moreover, Doppler ultrasound is also a useful procedure for the assessment of vessel wall inflammation, extracranial vessels and vessel wall thickness.³

Takayasu arteritis is a chronic, relapsing disease that is associated with significant morbidity and disability. A cohort study of 88 patients from India shows a 5-year survival rate of 91% and

Figure 2 Three-dimensional volume-rendered image again shows stenosis at the mid abdominal aorta, obliteration of the superior mesenteric artery (SMA), and numerous collaterals.

Figure 3 Axial CT image in arterial phase shows significant collaterals at the Winslow pathway (arrows).

Figure 4 Axial CT image in arterial phase shows significant narrowing of the abdominal aorta (arrow) with coeliac artery aneurysm (asterisk).

10-year survival rate of 84%. Congestive heart failure was the most common reported cause of death.²

Treatment aim of Takayasu arteritis is to control disease activity and preserve vascular competence. High-dose corticosteroids are the mainstay of treatment for Takayasu arteritis. Approximately half of the patients treated with steroids will respond.³ In patients with symptomatic stenotic or occlusive lesions, interventional or surgical therapy are required. Percutaneous transluminal angioplasty and bypass surgery would be the most common treatment.⁵ Bypass surgery and reconstruction demonstrated better sustained patency than percutaneous transluminal angioplasty. Complications of bypass procedures included restenosis, thrombosis, infection and haemorrhage. Cleveland clinic reported 36% of 44 bypass procedures had restenosis or occlusion which requires additional procedural intervention. Hence, vigilant clinical, laboratory and radiological follow-up are essential postoperatively to detect early complications.

The vascular interventional procedures carry a high rate of

Figure 5 Section of the vessel wall shows patchy chronic inflammatory cell infiltrates within the adjacent fibroadipose tissue.

restenosis, especially the times of active disease. Therefore, surgery is usually postponed and recommended until Takayasu arteritis is in clinical remission. Cleveland clinic defined clinical remission as follows: (1) absence of clinical or laboratory markers of active disease and absence of radiographic evidence of new vascular lesions for more than 6 months and (2) treatment with less than 10 mg/day of prednisone.²

In our case, the patient only presented with incidental hypertension during body check-up. Otherwise she was clinically asymptomatic. The paucity of clinical symptoms and signs limits the early detection of Takayasu arteritis. Correlation between acute phase reactants (erythrocyte sedimentation rate and C-reactive protein) and radiological findings is also poor. Takayasu arteritis disease activity maybe subclinical and not recognised by laboratory surrogate markers in up to half of all patients. These are the reasons rendering the early diagnosis and subsequent follow-up of Takayasu arteritis difficult.²

In summary, MAS is an important clinical entity to recognise and treat accurately.

A thorough physical examination including blood pressure measurement would assist in the early recognition of MAS. A vigilant approach and follow-up at the postoperative period would be essential for early detection of disease relapse.

Learning points

- ▶ Mid-aortic syndrome (MAS) caused by Takayasu arteritis is a rare cause of uncontrolled hypertension in the young.
- ▶ Most complications are due to long-lasting malignant hypertension. They included hypertensive encephalopathy, cerebrovascular accident, congestive heart failure and renal failure.
- ▶ Medical treatment with surgical correction is effective in MAS.
- ▶ Blood pressure measurement and accurate physical examination are essential at every paediatric consultation.

Contributors KCAC is the primary and corresponding author. Y-LL assisted in preparing the manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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