INTRODUCTION

Takayasu’s arteritis (TA) manifestations are demonstrable through the usage of various imaging modalities (i.e., echocardiogram, CT, MRI) as well as angiographic studies, which are the unique features of this case.

CASE REPORT

A 12-year-old girl presented to an emergency department in a district hospital with complaints of reduced effort tolerance and on-and-off palpitations for the past one month. She had no recent history of fever and she was admitted with a diagnosis of dilated cardiomyopathy. Cardiomegaly was seen in the chest radiograph but both lungs looked clear. No consolidative changes or pleural effusions were observed.

On examination, she looked slightly pale, but was otherwise active. There were discrepancies in her upper limb and lower limb systolic and diastolic blood pressure (BP) readings. Her right upper limb BP was 124/81 mm Hg, left upper limb was 99/78 mm Hg, right lower limb was 84/67 mm Hg, and left lower limb was 98/65 mm Hg. Her blood analyses showed a raised erythrocyte sedimentation rate of 44 mm/h and CRP 32.8 mg/L. The Mantoux test was negative and venereal disease research laboratory was non-reactive. Antinuclear antibodies and rheumatoid factor were negative. C3/C4 levels were normal. She was treated with anti-heart failure medications.

An echocardiogram was performed and showed global hypokinesia, a dilated left ventricle (LV), reduced systolic function of 28% (by the modified Simpson’s method), moderate MR due to annular dilatation, and a severely stenosed descending aorta with very restricted flow.

A CT angiogram done that later confirmed long segment stenosis of the descending thoracic aorta (Fig. 1A, black arrow) estimated to measure 20.0 mm and the narrowest luminal diameter was measured at 4.0 mm. The proximal left subclavian artery (LSCA) showed significant stenosis as well (Fig. 1B, black arrowhead). The distal LSCA, left axillary, and brachial arteries were faintly visualised suggestive of collateral flow from the left vertebral artery.

Her cardiac MRI revealed a long segment diffuse wall thickening with enhancement involving the descending thoracic aorta and enhancing soft tissues around the branches of pulmonary arteries noticeable in a T1-weighted post-gadolinium fat-suppression sequence (Fig. 1C, white arrows), presumably due to myocarditis.

We report the case of a 12-year-old girl who presented with heart failure symptoms that were treated as dilated cardiomyopathy. The echocardiogram showed a dilated left ventricle, poor ejection fraction, and long segment narrowing of the descending thoracic aorta. A cardiac CT further supported this finding, hence, the diagnosis of Takayasu’s arteritis was proposed. The cardiac MRI revealed a non-ischemic pattern in delayed gadolinium enhancement at the lateral left ventricular wall. She was given steroidal therapy due to vasculitis but was later referred for stenting of the stenosed descending thoracic aorta.

Key words Takayasu’s arteritis (TA) · Cardiac magnetic resonance imaging (cardiac MRI) · Vasculitis · Delayed gadolinium enhancement.
active arteritis. Another important finding was subepicardial delayed enhancement of the inferolateral and anterolateral LV wall, that was concluded to be myocarditis (Fig. 1D, black arrows). The magnetic resonance angiography study also showed significant stenosis of the proximal right renal artery (Fig. 1E, yellow circle). The rest of the coeliac trunk, superior mesent-

Fig. 1. A: CT angiography (CTA) shows long-segment narrowing of descending thoracic aorta (right, black arrow). Image on left is in 3-dimensional volume rendering technique (3D VRT) reconstruction. B: CTA with maximum intensity projection reconstruction (10 mm) shows narrowed left subclavian artery (black arrowhead). Compare to calibre on opposite side (black star). C: Enhancing soft tissues (white arrows) around the branches of pulmonary arteries suggestive of active arteritis in T1 weighted image post gado fat saturation. D: Subepicardial (black arrow) delayed enhancement at mid inferolateral left ventricle wall. E: VRT of abdominal aorta magnetic resonance angiography demonstrated stenosis of proximal third of right renal artery (yellow circle). Compare with normal left side. F: Pre-stent (on the right) depicts a long segment stenosis of descending aorta (36.2 mm) with narrowest calibre of 2.3 mm and post stenting (on the left) angiogram shows good luminal expansion.
teric artery, left renal artery, and both common and external iliac arteries appeared normal. Due to suspected TA with myocarditis, she was given intravenous methylprednisolone therapy for three consecutive days 30 mg/kg (810 mg), followed by oral prednisolone 30 mg once daily. Subsequently, she underwent stenting of the descending aorta. The angiography study revealed the narrowest diameter in the descending aorta was 2.3–2.8 mm and it was 36.2 mm in length. A BeGraft covered stent (Bentley InnoMed, Hechingen, Germany) (12×59 mm) was deployed within this segment with excellent results (Fig. 1F for pre-, post-stenting). She was discharged and is currently under cardiology and rheumatology follow up.

DISCUSSION

TA is an idiopathic, chronic inflammatory disease which predominantly affects the medium and large arteries (e.g., the aorta and its branches) [1]. It is a granulomatous type of vasculitis and most commonly manifests as stenotic lesions, apart from becoming aneurysmal [2].

Typically, patients show decreased or absent pulses with BP discrepancies. This condition is more predominant in females than males, at a ratio of 9:1 [3]. Most adults present with TA at the mean age of 30 years, but children and even infants may also be affected.

The most common clinical features in pediatric patients are arterial hypertension (72.7%), followed by cardiovascular complications (45%), systemic features (36%), neurological deficits (36%), pulmonary complications (27%), and others (skin, etc.) [4].

Gadolinium in cardiovascular magnetic resonance studies is very useful to indicate the presence or absence of vascular and myocardial inflammation in TA. Interestingly, our case demonstrated vasculitis by the presence of enhancing vascular walls and stenosis at various sites including the descending aortic wall, branches of pulmonary arteries, and the proximal right renal artery.

Apart from the vascular enhancement, this case illustrates a non-ischemic pattern of myocardial enhancement i.e. subepicardial enhancement at the lateral LV wall during late gadolini-um; hence, favouring the diagnosis of myocarditis. The incidence of myocarditis in TA has been partially attributed to lymphocytic infiltration, based on a study of Indian cases [5]. This suggests a race or regional distribution as seen in Asia. TA also can cause LV dysfunction in some cases.

The ultimate diagnosis for Takayasu’s disease should be with vascular biopsy which may not be feasible for the adolescent and pediatric patient age groups. Usually, distinct clinical presentations, supported by positive imaging findings like cardiac MRI, are the mainstay of diagnosis. TA is usually treated with glucocorticoids. However, angioplasty or bypass grafts may be essential in dealing with irreversible arterial stenosis.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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