Improving Pulmonary Perfusion in a Child with Takayasu’s Arteritis

Bhavin L Ram, Robbie K George, Aruna Bhat
Narayana Institute of Vascular Sciences, Pediatric Rheumatology, Narayana Hrudayalaya Hospital, Bengaluru, Karnataka, India

Abstract

Takayasu’s arteritis (TA) is a form of granulomatous arteritis of unknown etiology, which frequently involves pulmonary artery (PA) also. Here, we describe a report of a 14-year-old boy with TA and disabling pulmonary compromise, who was treated by PA angioplasty and stenting. The fact that he showed such a dramatic response to therapy suggests that it might be appropriate to consider improving pulmonary perfusion not only for pulmonary artery hypertension but also for its respiratory benefits.

Keywords: Pulmonary artery hypertension, pulmonary artery stenting, Takayasu’s arteritis

Introduction

Takayasu’s arteritis (TA) is a form of granulomatous arteritis of unknown etiology. This chronic inflammatory disease involves large- and medium-sized arteries, primarily the aorta and its large branches as well as proximal portions of pulmonary, coronary, and renal arteries, resulting in varying degree of stenosis, occlusion, or dilatation of the involved vessels.[1]

TA is predominantly a disease of young adults in the second and third decades of life, with a female: male ratio varying from 1.3:1 in India[2] to 9:1 in Japan.[3] The onset of illness may be earlier, including in childhood but rarely in infancy. In children, it is a serious illness with a reported mortality of 10%–30% on follow-up.[2,4,5]

Based on the angiographic features, TA is divided into five types, and according to this classification system, involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+) [Table 1].[6]

Here, we describe a report of a young boy with TA and disabling pulmonary compromise treated by pulmonary artery (PA) angioplasty and stenting.

Case Report

A 14-year-old boy was referred to the Pediatric Department of Narayana Hrudayalaya Hospital for the evaluation and management of TA. He was a known case of TA involving aortic arch and its branches [Figure 1] and was on medical management (corticosteroids and other immunosuppressive drugs) for the last 8 months. On admission, he presented with difficulty in breathing on walking a few steps and sometimes at rest, generalized weakness, and occasional fever for the last 1 month.

Clinical findings

His hemodynamic parameters were normal (blood pressure 130/80 bilaterally, heart rate 98 beats/min), except tachypnea (respiratory rate 34/min). Oxygen saturation was 76% on room air and 100% with 5 l of oxygen. Auscultation and percussion were unremarkable, except for the absence of breath sounds in the right lower zone. Left carotid and left brachial pulse were weak compared to the opposite side. His hemoglobin was 10 g/dl, erythrocyte sedimentation rate 9 mm/h, and C-reactive protein 7.68.

The chest X-ray showed mild cardiomegaly with prominent main PA contours and haziness in the right middle and lower lobe. ECHO was suggestive of severe pulmonary arterial hypertension (PAH).

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He was commenced on IV antibiotics for a suspected right lower lobe pneumonia.

There was no improvement seen after a week of antibiotic and supportive treatment. In view of the history and clinical course, a lung (V/Q) perfusion-imaging scan was done that demonstrated nonvisualized left lung with large areas of mismatched perfusion defects in right lung parenchyma [Figure 2]. Computed tomography (CT) pulmonary angiography revealed total occlusion of the left main PA with short segment concentric thickening of the right lower lobar artery with aneurysmal dilatation just beyond its bifurcation [Figure 3].

In view of his worsening and disabling pulmonary compromise despite medical therapy, options to improve his pulmonary reserve and treat the PAH were explored. Our cardiac surgery team has a large experience on pulmonary endarterectomy for pulmonary thromboembolism-associated PAH. However, it was felt that there was no suitable surgical option for the complete occlusion of the left PA. Hence, we offered a right PA branch angioplasty and stenting, primarily with a view to reducing the PAH. The patient and his family were counseled regarding the options of ongoing medical management and the risks of PA stenting that included death and lung loss.

Before the procedure, the child continued to receive steroids and other immunosuppressive agents with a view to decreasing inflammation in the vascular bed before surgery.

**Procedure**

Under local anesthesia (LA), right femoral artery (for monitoring arterial pressure) and vein access were obtained and a 0.035 (Diameter in inch) wire with a vertebral catheter was negotiated into the right main pulmonary trunk. Recorded PA pressures were normal.

Angiogram [Figure 3] demonstrated complete occlusion of the left main PA with a normal right apical and middle artery with 70% stenosis of the inferior main trunk and 90% stenosis of its apical branch.

The inferior main trunk lesion was crossed and glidewire exchanged with stiff wire. After predilatation, a 6 mm × 30 mm balloon-expandable stent was deployed across the main right inferior PA [Figure 4]. Apical branch lesion was crossed with 0.014 (Diameter in inch) glide wire and angioplasty was done with 5 mm × 20 mm balloon. Check angiogram showed good flow across the lesions without any residual flow-limiting stenosis. The procedure was done under therapeutic heparin cover. After the procedure, the patient was kept on therapeutic anticoagulation. Before and after the procedure, there was no difference seen in PA pressure.

**Follow-up**

The patient was kept on therapeutic anticoagulation, and gradually over the next 2 days, he was weaned off the oxygen. Within 1 week, the boy was able to walk without dyspnea on exertion. He was discharged on steroids to maintain suppression of disease activity. He returned to school and normal activities within a month and this is maintained at his 18-month review. We repeated CT pulmonary angiogram at 18-month follow-up, which did not show any flow-limiting stenosis in treated segment [Figure 4]. He continues to remain on regular immunosuppressive treatment on daily low-dose oral steroids and mycophenolate mofetil and shows low disease activity score. We will do next imaging either at 5 years or if the patient becomes symptomatic before that.

**Discussion**

Although TA is predominantly a disease of young adults, it is common in childhood. In children, female preponderance is less obvious. Although the involvement of PA in TA is seen in nearly 70% of the patients on angiographic studies, it is usually mild and most patients with PA involvement remain asymptomatic. The disease involves segmental and subsegmental branches, more in the upper lobes, but larger branches may be involved. A history of hemoptysis, chest pain, disproportionate PAH, or abnormal ventilation-perfusion scan may suggest pulmonary involvement.

Our patient was a known case of TA and was already on treatment for the same, but undiagnosed patients with suspicion
of TA or patients with pyrexia of unknown origin can be benefited with $^{18}$F-fluorodeoxyglucose (F-FDG) positron emission tomography (PET) scan or more precisely $^{18}$F-FDG PET and CT scan to diagnose it very early, particularly in prepulseless stage.\cite{7,8}

The response of PA lesions to therapy has not been well studied. In the presence of symptomatic stenotic or occlusive lesions in TA, endovascular revascularization procedures like angioplasty or stent placement should be considered.\cite{9} Symptomatic PA stenosis is a relatively uncommon manifestation of TA. Clinical observations regarding the safety and efficacy of stent implantation in branch PA and its influence on clinical management have been encouraging.\cite{10} A few studies have shown that percutaneous angioplasty and stent implantation is a safe and effective treatment in patients with pulmonary stenosis caused by TA.\cite{9-11}

Published studies suggest that these endovascular procedures should be undertaken with great care and be reserved for specific indications. Both surgical and endovascular treatments become risky and achieve poorer outcomes, if they are undertaken during a period of acute inflammatory condition. Because of the diffuse, inflammatory, and possibly progressive nature of the disease, surgical treatment is not preferred for TA except for undilatable symptomatic stenotic lesions and large aneurysms.

It is important to optimize immunosuppression in these patients, particularly when the disease is active, before and after surgery to achieve better outcomes.
As is known, lesions in TA are well known to recur and he may well need a further angioplasty in the future. For the present case, we intend to follow him up on clinical grounds with a low threshold for a re-intervention, as long-term follow-up data on children are not available.

**Conclusion**

Our primary intent had been to reduce the PAH to improve his cardiopulmonary reserve and protect the heart. However, we were surprised to find that the PA pressure measurements did not show significant PAH. We proceeded with the stenting in the hope to reduce the ventilation-perfusion mismatch and allow better oxygenation. The fact that he showed such a dramatic response to therapy suggests that it might be appropriate to consider improving pulmonary perfusion not only for PAH but also for its respiratory benefits.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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