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American Journal of Interventional Radiology

Case Report *Vascular Interventions*

Endovascular treatment and long-term safety for pulmonary artery stenosis due to Takayasu's arteritis – A case report

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Received: 07 May 2024
Accepted: 28 July 2024
Published: 20 September 2024

DOI
10.25259/AJIR_23_2024

Quick Response Code:

ABSTRACT

Takayasu's arteritis is a rare large vessel vasculitis which can involve the pulmonary arteries and progress to pulmonary artery (PA) stenosis with pulmonary hypertension. We present a case of Takayasu arteritis complicated by bilateral pulmonary stenosis and pulmonary hypertension that resolved after PA stenting and angioplasty. This case demonstrates the efficacy of endovascular intervention and sustained safety during 10 years of follow-up.

Keywords: Pulmonary artery stenosis, Pulmonary hypertension, Safety, Takayasu arteritis

INTRODUCTION

Takayasu's arteritis (TA) is an idiopathic inflammatory vascular disorder that may involve the thoracoabdominal aorta, its branches, and the pulmonary arteries. Our patient presented with rapidly progressive dyspnea and pulmonary hypertension due to bilateral pulmonary artery (PA) stenosis secondary to TA. She was treated by endovascular therapy and has remained stable over 10 years of follow-up.

CASE REPORT

A 68-year-old woman with no significant past medical history presented in June 2012 with several months of progressive dyspnea on exertion, exertional dizziness, fatigue, and extremity weakness associated with intermittent low-grade fever and a 10-kg weight loss. Her work-up demonstrated anemia and elevated (C-reactive protein; 7.9 mg/dL). There was moderate cardiomegaly on chest radiography. A transthoracic echocardiogram (TTE) showed pulmonary hypertension with an estimated right ventricular systolic pressure of 70 mmHg and a large pericardial effusion. Her cardiac stress test showed no evidence of ischemia or scar and a normal left ventricular (LV) ejection fraction. Computed tomography (CT) angiogram and subsequent MR angiogram revealed high-grade bilateral PA stenoses and marked aortic wall thickening [Figure 1]. The constellation of these findings was consistent with large vessel vasculitis, most likely TA.

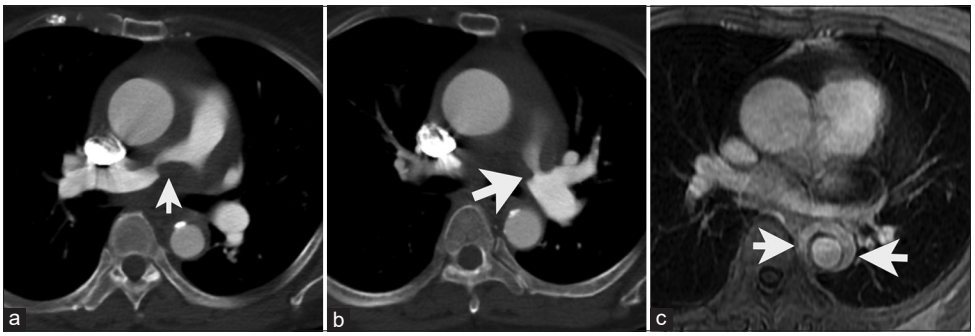

Figure 1: A 68-year-old female presented with progressive dyspnea on exertion, exertional dizziness, fatigue, and extremity weakness. Computed tomography angiogram demonstrating (a) high-grade stenosis at the origin of the right pulmonary artery (white arrow) and (b) circumferential stenosis of the proximal left pulmonary artery (white arrow). MR angiogram showing (c) gadolinium-enhancing adventitial thickening of the descending thoracic aorta (white arrows).

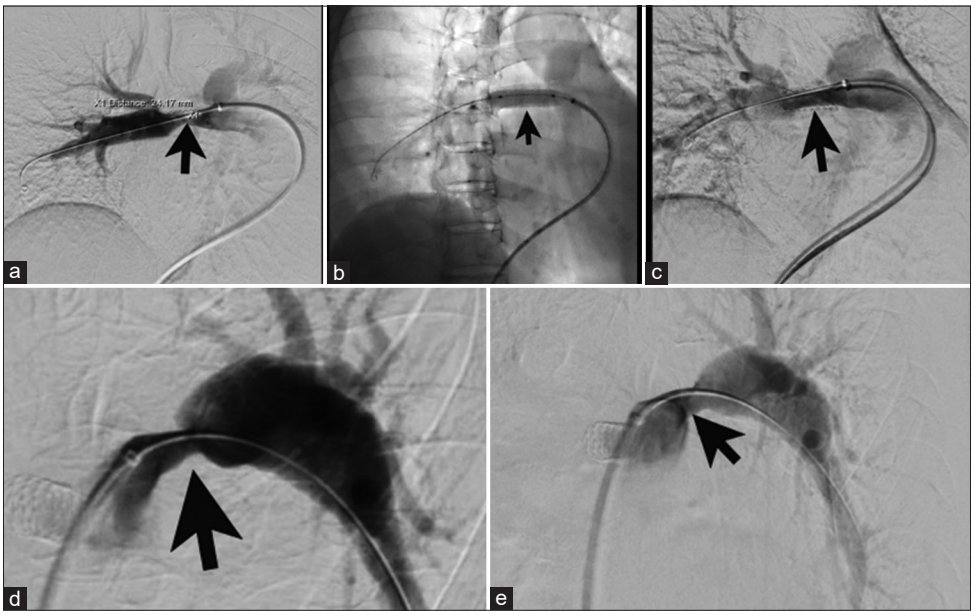

Figure 2: A 68-year-old female with pulmonary artery stenosis undergoes endovascular therapy. Digitally subtracted right pulmonary angiogram showing (a) 24-mm long stenosis of proximal right pulmonary artery (black arrow); (b) placement of balloon-expandable stent across the right pulmonary artery stenosis (black arrow); (c) digitally subtracted angiogram following placement of stent (black arrow); (d) left pulmonary stenosis before angioplasty (black arrow); (e) elastic recoil of left pulmonary stenosis (black arrow) following angioplasty.

Based on the high-grade PA stenoses and the patient's progressive symptoms, bilateral selective PA angiography was performed in June 2012. The digital subtraction angiography demonstrated a 24 mm-long high-grade stenosis in the proximal right PA with a pressure gradient of 65 mmHg, as well as a focal (<10 mm) stenosis in the proximal left PA consistent with TA [Figure 2a]. The high-grade stenosis of the right PA was stented with a 10 × 29 mm Genesis stent (Cordis, Miami Lakes, FL) followed by 12 mm balloon dilation of the stent. A post-stent angiogram showed marked improvement in the pressure gradient between the right (27/2 mmHg) and main PA (37/9 mmHg) which had decreased from 65 mmHg to 10 mmHg [Figure 2b and c].

The focal stenosis in the left PA was not considered amenable to stenting due to the location of the stenosis and was treated with angioplasty using a 12-mm balloon. Post-angioplasty imaging showed modest improvement in the pressure gradient (46/12 mmHg proximal and 28/11 mmHg distal to the angioplasty) with the final pressure gradient of 18 mmHg remaining somewhat elevated due to elastic recoil of the stenosis [Figure 2d and e].

The patient tolerated the procedures well. Her dyspnea, fatigue, and supplemental oxygen requirements improved significantly over 48 h. She was started on prednisone 60 mg/day and aspirin 81 mg/day. Two months later, she was no longer complaining of fatigue or dyspnea. She then underwent a slow steroid taper to 5 mg daily. After 1 year, she began re-developing shortness of breath on exertion.

A chest CT scan in November 2013 demonstrated stenosis of the left main PA which had progressed since prior imaging and was nearly occlusive. Based upon this finding, she underwent repeat bilateral selective PA angiography in December 2013, 18 months after her initial endovascular therapy. The left PA had nearly occlusive high-grade stenosis with a 50 mmHg gradient [Figure 3a]. This was stented with a 10 × 19 mm Genesis stent followed by 12-mm balloon dilation; the post-procedure systolic gradient was 0 mmHg [Figure 3b]. A focal restenosis of the previously placed R PA stent was then dilated with a 12-mm balloon [Figure 3c].

Repeat chest CT imaging in January 2020 demonstrated widely patent PA stents [Figure 4]. A repeat TTE (August 2022) was significant only for mild concentric LV hypertrophy and trace tricuspid regurgitation with an estimated RVSP of 23 mmHg. She has been followed for over 10 years from her second stenting procedure without any reoccurrence of dyspnea.

DISCUSSION

TA is a chronic non-specific inflammatory disease, which predominantly affects the aorta and its major branches with a higher incidence in women than in men.[1] The pathologic changes in TA are characterized by granulomatous pan arteritis with adventitial thickening and cellular infiltration of the tunica media, myofibroblast proliferation resulting in intimal hyperplasia and fibrosis of the tunica media and intima that eventually leads to stenosis.[2] Because histopathologic specimens of affected large vessels are seldom available, and the fact that the histopathologic appearance of TA can mimic another arteritis, the diagnosis of TA is typically based on the combination of clinical presentation, laboratory evaluation, and diagnostic imaging.[1]

PA involvement in TA is usually accompanying aortic involvement and has been reported on the autopsy of 20–56% of TA patients.[2,3] The rate of PA hypertension in TA

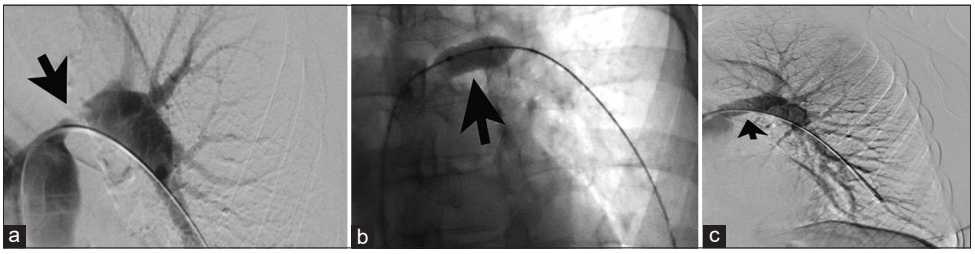

Figure 3: A 70-year-old female with reoccurrence of dyspnea due to pulmonary artery stenosis undergoes repeat endovascular intervention. Selective left digital subtraction arteriogram showing (a) nearly complete occlusion of the left pulmonary artery (black arrow); (b) balloon expandable stent placement across the left pulmonary artery stenosis (black arrow); (c) after stent placement showing resolution of stenosis (black arrow) and increased perfusion of left pulmonary artery branches.

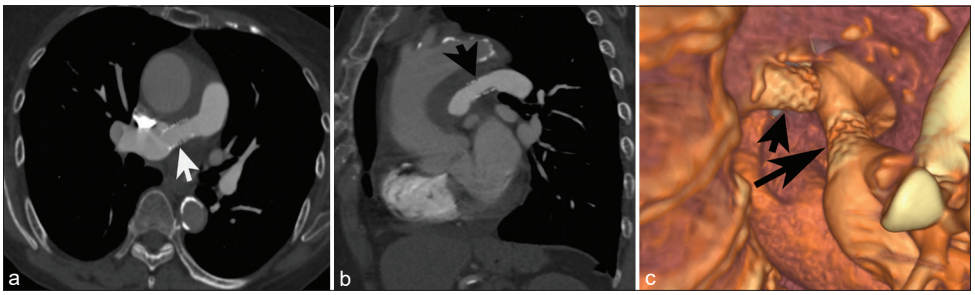

Figure 4: A 76-year-old female with a history of takayasu arteritis of the pulmonary arteries has follow-up imaging with axial computed tomography (CT) angiogram 8 years following right pulmonary artery stent placement. (a) widely patent right pulmonary artery (white arrow); (b) sagittal oblique CT angiogram with widely patent left pulmonary artery (black arrow); (c) 3D CT angiogram image from posterior oblique perspective showing widely patent left (short black arrow) and right (long black arrow) pulmonary arteries.

patients is approximately 12 to 13% in various reports and is associated with a less favorable prognosis.[2]

High-dose glucocorticoids remain the standard medical therapy for TA. Achieved remission is rarely stable with steroid wean and often requires prolonged high-dose treatment.[2,4] In addition, no remarkable change has been shown in PA stenoses following glucocorticoid therapy.[5] Surgical treatment has been successfully performed for the PA stenosis. However, surgical limitations including graft re-occlusion, anastomotic site aneurysm, and morbidity of thoracotomy have precluded the widespread use of this therapy.[3]

Kreutzer *et al*. performed balloon dilation of multiple vessels in 11 patients. The authors suggested that balloon dilation offered successful short-term reduction in right ventricular hypertension and improved respiratory symptoms.[6] Rothman *et al*. reported that 4 adult patients with multiple intralobar pulmonary arterial stenoses all responded acutely to balloon angioplasty and stent placement.[7] Qin *et al*. reported 4 patients with bilateral high-grade PA stenosis treated with angioplasty and stenting (3 patients) and angioplasty alone (1 patient) showing sustained improvement of symptoms in long-term follow-up (mean 34 months) in three patients treated with stent placement versus restenosis at 18 months in the single patient treated with angioplasty alone.[3] Complications have been reported in recent literature to include in-stent stenosis, reperfusion pulmonary edema, vascular dissection, hematoma at puncture site, and hemoptysis.[8-10]

Two observational studies of patients undergoing pulmonary angioplasty have demonstrated improvement in functional class, 6-minute walk test (6MWD), NTproBNP, echocardiographic parameters, cardiac catheterization, and cardiac cause of death.[9,10]

Although endovascular therapy of PA stenosis has been discouraged in the active phase of TA,[3] the nature and severity of the symptoms such as our patient may warrant prompt intervention. Angioplasty of the stenotic PA segment followed by stent placement may have a better outcome than angioplasty alone, with no apparent advantage in using drug-eluting stents.[3,4] However, anatomic considerations can preclude the use of stent in certain cases (as initially in our patient's left PA). Finally, as TA is a progressive inflammatory disease, corticosteroids and regular follow-up are needed to monitor patients for restenosis and new lesions.

CONCLUSION

In conclusion, angioplasty and stent implantation may be a safe and effective treatment for patients with severe pulmonary hypertension due to TA. This is the first report demonstrating 10 years of follow-up after endovascular therapy for TA with resolution of pulmonary hypertension and without any evidence of disease reoccurrence. More studies are needed to clarify the safety profile and longevity of disease-free status post-angioplasty and stent implantation of stenotic PAs for pulmonary hypertension in TA.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Vahdatpour C, Clark T, Palevksy H. Endovascular treatment and long-term safety for pulmonary artery stenosis due to Takayasu's arteritis– A case report. Am J Interv Radiol. 2024;8:14. doi: 10.25259/AJIR_23_2024