

Endovascular biopsy in Takayasu arteritis

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Abstract

Takayasu arteritis is a chronic inflammatory arteriopathy of the large vessels, mainly the aortic arch and its branches. The disease progression varies, ranging from a rapid progression to quiescence reached within 2 years. The activity of the disease is evaluated by biochemical markers, but at times, there is a discrepancy between the clinical picture and biochemical markers. Histopathology helps in assessing the disease activity, but it is not included in the antemortem diagnosis because of the invasiveness of the procedure and the availability of noninvasive vascular imaging. However, it may be performed simultaneously with angioplasty. Here we present a case of endovascular aortic biopsy conducted to detect active inflammation in the aortic wall.

Keywords: Takayasu arteritis, aortic biopsy, endovascular

Introduction

Takayasu arteritis, or "pulseless disease," is a rare chronic inflammatory arteriopathy affecting large vessels, such as aorta, its branches, and the main pulmonary artery. Classic triphasic pattern of clinical presentation is likely to occur only in a minority of patients. It preferentially affects women younger than 50 years. The disease progression varies, ranging from a rapid progression to quiescence reached within 2 years (1). The disease activity is evaluated by biochemical markers, but sometimes, there is a discrepancy between the clinical picture and biochemical markers, and further evaluation is required. Here we present a case of endovascular aortic biopsy during angioplasty, conducted to detect active inflammation in the aortic wall.

Case Presentation

A 20-year-old female of Asian descent presented with complaints of decreased appetite, nausea, and vomiting that lasted for 2 weeks. Clinically, she was found to be hypertensive (170/80 mm Hg at right arm). Bilateral upper and lower limb pulses were normal. On further work-up for young-onset hypertension, the patient had elevated serum creatinine levels (4.5 mg/dL), C-reactive protein (9.77 mg/dL), and ESR (>55 mm/hour). There was no past history of tuberculosis.

Ultrasound revealed different kidney sizes (right kidney, 10 cm; left kidney, 8.7 cm). Renal Doppler showed a parvus tardus pattern bilaterally with an increased acceleration time. A magnetic resonance angiography revealed circumferential narrowing of the descending thoracic aorta and suprarenal abdominal aorta. The branches of the suprarenal aorta, celiac axis, and superior mesenteric artery (SMA) also showed more than 90% stenosis with collateral arcades between SMA and IMA. A bilateral renal artery stenosis was present (right side 90% and left side 80%) each extending for a length of 12 mm in the main renal artery. Infrarenal aorta was completely normal. Dialysis was conducted, and the patient was put on a quadruple antihypertensive regimen. As her renal function was deteriorating rapidly, she was referred for endovascular management. On DSA, the left renal artery was diffusely narrowed from the origin. The right renal artery showed an ostial narrowing, which was crossed using the 0.014 microwire followed by angioplasty and stenting. Good flow was obtained in the right renal artery after stenting.

Post stenting, the patient was maintained on a standard double antiplatelet regime. The blood pressure was controlled with a lesser dose of antihypertensive drugs, and the serum creatinine level was also stabilized to 1.1 mg/dL. The patient was started on prednisolone (0.75 mg/Kg body weight) and was discharged.

Six months later, the patient again presented with uncontrolled hypertension (180/100 mm Hg at the right arm). On examination, the lower limbs pulse was feeble with features of vascular claudication. Bilateral femoral, popliteal, posterior tibial, and dorsalis pedis arterial pulses were absent. A biochemical evaluation of the acute-phase reactants showed a raised ESR (40 mm/hour), despite immunosuppression. The CRP



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was within the normal range (<0.3). Because disease progression was suspected, oral methotrexate was added to prednisolone. A CT angiogram of the abdominal aorta was done, revealing significant thickening of the wall of the abdominal aorta with luminal narrowing, and there was a focal dissection flap in the aorta near the renal artery ostia site (Figure 1). Renal Doppler revealed a normal flow in the right renal artery stent. In view of clinical progression and discrepancy in the biochemical markers, vascular biopsy was planned along with aortic angioplasty.

The patient was reassessed with repeat angiographic examination. A 6 Fr arterial sheath was placed in the right common femoral artery. 5000 U of heparin were given. A right renal angiogram showed the patent stent with good distal flow in the right renal artery. An aortogram was done, and it showed smooth tapering of the thoracic aorta and abdominal supra-renal aorta (Figure 2a-c). There was a significant pressure gradient between the thoracic and the infrarenal abdominal aorta. Endovascular biopsy was done from the narrowed segment of the suprarenal aorta using the endomyocardial biopsy forceps, jaws (Argon medical devices) (Figure 2d, e). A 1.5 mmx50cm fenestrated jaw biopsy forceps was used and was placed through the access introducer 5F x 45cm. Multiple passes were done, and significant tissue was obtained for histopathology. A post-biopsy angiogram did not reveal any dissection from the biopsy site, and there was no contrast extravasation to suggest perforation. Following biopsy, angioplasty of the suprarenal abdominal aorta and thoracic aorta was done using an 8x40 mm balloon followed by a 12x40 mm balloon. Post angioplasty, there was a decrease in the pressure gradient and a visible angiographic result. A histopathologic examination revealed dense collagenous tissue with occasional benign fibroblasts and inflammatory infiltrates in the biopsy specimen (Figure 3).

Discussion

Takayasu arteritis is an idiopathic inflammatory vascular disorder that may involve the thoraco-abdominal aorta and its branches. Women comprise 80% to 90% of patients with Takayasu arteritis, mostly in the second to third decade. Although the etiology is uncertain, many theories have been postulated, including autoimmune mechanisms and infection with *Mycobacterium tuberculosis* (2). Clinically, these patients present with non-specific symptoms such as fever, weight loss, and fatigue during the early stages (3). With progression of the disease, vascular symptoms dominate the clinical picture. These include reduced blood pressure

with weakened pulses in the upper extremities, ocular disturbances, blindness, pulmonary artery hypertension, and myocardial infarction.

The criteria proposed by Sharma et al. (4) in diagnosing Takayasu arteritis consists of three major criteria, including the left and right mid-subclavian artery lesions and characteristic signs and symptoms lasting for at least 1 month and 10 minor criteria: a high ESR, carotid artery tenderness, hypertension, aortic regurgitation or

annuloaortic ectasia, pulmonary artery lesion, left mid-common carotid lesion, distal brachiocephalic trunk lesion, descending thoracic aorta lesion, abdominal aorta lesion, and coronary artery lesion. The presence of two major or one major and two minor criteria or four minor criteria suggests a high probability of Takayasu arteritis. When applied to 106 Indian patients with angiographically proven Takayasu arteritis and 20 control subjects, it had a sensitivity of 92.5% and specificity of 95% (4). Apart from changes

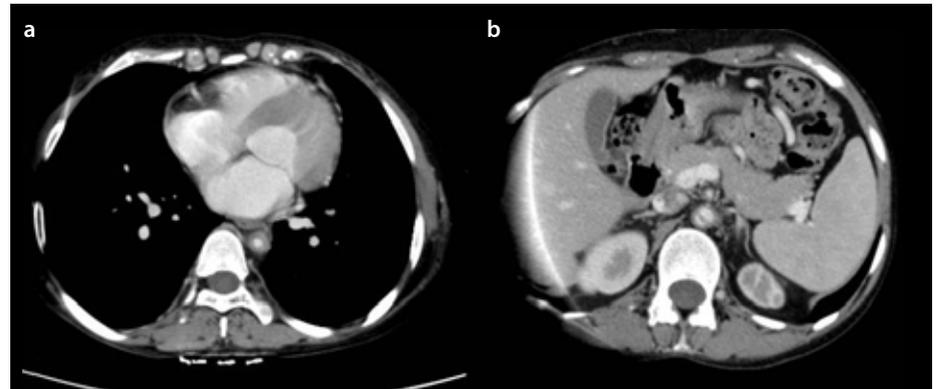


Figure 1. a, b. A computed tomography angiogram shows a diffuse circumferential thickening of the descending thoracic and abdominal aorta with severe luminal narrowing (a); a dissection flap is noted in the suprarenal abdominal aorta, which is seen to separate the false lumen (posterior) from the true lumen (anterior) (b)



Figure 2. a-e. Digital subtraction angiography in the same patient shows a luminal narrowing and irregularity of the aortic wall extending from the level of thoracic aorta to the abdominal aorta (a); a thin dissecting flap is seen within the abdominal aorta just above the right renal artery ostia (b); there is evidence of a good flow through the stent placed in the right renal artery (c); a 1.5mmx50cm fenestrated jaw biopsy forcep within the abdominal aorta with jaws open (d); jaws closed (e)

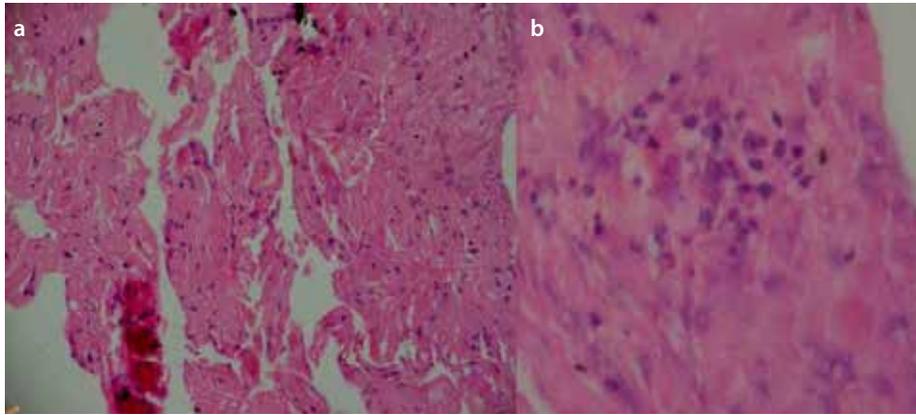


Figure 3. a, b. A histopathologic specimen (H&E) shows dense fibrocollagenous tissue with the presence of benign fibroblasts and hemorrhage (a); a high resolution image shows the presence of neutrophils and plasma cells-mixed inflammatory infiltrates (b)

in the acute-phase reactant and imaging, recently, the disease extent index for Takayasu arteritis and the Indian Takayasu Arteritis Score seem to be helpful in assessing the disease activity and damage in Takayasu arteritis. Contrast-enhanced MRA or CTA allow noninvasive imaging of the aorta and its major branches for the assessment of luminal status as stenoses and aneurysms. Vessel wall thickening, edema, and enhancement in venous phase with a low attenuation ring are features of disease activity in CT angiography. Ultrasound may show hypoechogenicity and mural thickening in active lesions, however more useful for superficial vessels as carotids (5, 6).

Contrast enhancement of aortic wall disappeared in 93.75% patients after glucocorticoid treatment in a study by Prieto-Gonzalez et al. (7); however the vessel wall thickening persisted in two-thirds of patients. The authors concluded that a longer follow-up is necessary to determine the clinical significance of persisting wall thickening and its relationship with relapses.

The role of 18F-FDG Positron emission tomography to detect an active large vessel vasculitis also has not been clearly established. Histopathology of the organs involved is the gold standard for analyzing disease activity in vasculitis and its management. However, being an invasive procedure, clinical, biochemical, and radiological correlation are routinely done in the follow-up assessment in Takayasu arteritis. An endovascular biopsy of the vessel wall is not done routinely as it requires interventional expertise and involves a certain degree of risk. Endomyocardial biopsy forceps are used generally for myocardial biopsy, but we used the same forceps for the biopsy of the aortic wall. The blunt end of the forceps protected the vessel wall from the injury.

Percutaneous endovascular biopsy is a minimally invasive procedure done with patient being awake and aware of the procedure. Not many indications and no randomized trial are there to access the efficacy of these biopsies, but there are case reports and small case series. A transvenous catheter biopsy of pulmonary artery sarcoma was performed by Winchester et al. (8) in a 51-year-old woman (8). Ronagi et al. (9) biopsied a primary aortic tumor in a 74-year-old woman. Most of the endovascular biopsies done were for vascular lesions suspicious for tumors. Only one case has been reported on endovascular aortic biopsy for the diagnosis of Takayasu arteritis (10). We have done an endovascular biopsy during aortic angioplasty to look for disease activity in a follow-up of a patient with vasculitis. Morrissey et al. (10) performed the biopsy in a 17-year-old woman who presented with lower-limb claudication and normal ESR. In MRA, the patient had stenosis of infrarenal aorta and the left renal artery. The biopsy specimen showed a thickened intima, disruption of the internal elastic lamina, giant cells, and inflammatory changes consistent with Takayasu arteritis. The patient was put on steroid therapy, and she improved. The authors concluded that the results of the biopsy can effectively guide management, thus avoiding unnecessary surgery in patients with active disease but normal ESR or in patients with a burnt-out disease and prolonged exposure to immunosuppressive agents. In our case, histopathology did not reveal definitive inflammatory infiltrates within the aortic wall. The patient was managed with angioplasty and immunosuppressive drugs and was discharged in a stable condition. Her last follow-up after 3 years; blood pressure was stabilized with two antihypertensive drugs.

Conclusion

Endovascular biopsy performed simultaneously with angioplasty in experienced hands may be helpful to assess the disease activity and patient management in Takayasu arteritis.

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