Case Report

Unique case of takayasu arteritis with bilateral carotid stenosis

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ABSTRACT

Takayasu's arteritis (TAK) is a chronic, multifactorial vasculitis of the large vessels of the body, sometimes affecting the medium vessels. Thus, the most commonly afflicted vessel is the aorta and its branches. It is typically encountered in the young to middle-aged population, with a higher tendency for females. In this case report, we have a post-menopausal South Asian female, who presented with left hemiparesis following a fall. Magnetic resonance imaging of the brain did not reveal any parenchymal involvement but magnetic resonance angiography revealed severe stenosis in bilateral common carotid arteries (CCA), later confirmed by digital subtraction angiography. Imaging supplemented by clinical features and elevated inflammatory markers confirmed our diagnosis of TAK of the CCAs but without aortic infliction. This case underscores the importance of high clinical suspicion, early recognition, and aggressive treatment of TAK to prevent adverse events and improve patient outcomes. Regular follow-up and monitoring of inflammatory markers and imaging studies are crucial in managing this chronic condition.

Key words: Aortitis syndrome, Arteritis, Inflammatory vasculitis, Large vessel vasculitis, Takayasu's arteritis

Takayasu's arteritis (TAK) is a rare, idiopathic chronic systemic inflammatory disease affecting large arteries of the human vasculature, predominantly the aorta, its major branches, and the pulmonary arteries [1]. Also known as "pulseless disease," this large vessel vasculitis has a female sex preference, most often presenting under the age of 40 years [2]. The pathogenesis of this disease is believed to be autoimmunity against certain vascular antigens and resulting inflammation of the affected vasculature [3-5]. The site and extent of the inflammation and resulting segmental stenosis, occlusion, dilatation, and aneurysm formation [6] present as a myriad of symptoms, ranging from pains and limb aches to limb weakness and palsy, syncopal attacks, headaches, and uneven blood pressure. Constitutional symptoms of ongoing inflammation such as fever, weight loss, and fatigue are often encountered first in these patients.

In our case, a 55-year-old South Asian female, presenting with atypical features of gradually evolving left-sided hemiparesis, with no comorbidities or typical signs of any vasculitis but fulfilling radiological and clinical criteria for TAK on further evaluation, way after its median age of presentation in the second decade, prompted us to report this interesting and rather unusual case.

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CASE REPORT

A 55-year-old post-menopausal female of South Asian origin presented with left hemiparesis. She endorsed a history of sudden fall 15 days back, with no significant comorbidities. Subsequently, the patient experienced left-sided weakness and soreness. She did not report to the hospital, attributing those concerns to her fall. Two weeks after the fall, slurring of speech developed prompting her to present at the neurology clinic. There was no history of fever, muscle ache, pain on physical exertion, loss of consciousness, or changes in vision.

Physical examination revealed significant left-sided motor weakness. Standard labs reported a leukocyte count of 9800 cells/ mm^3 cube and hemoglobin of 11.8 g/dL. As per stroke protocol, prompt magnetic resonance imaging (MRI) and magnetic resonance angiography of the brain were ordered. MRI revealed the presence of non-ischemic subacute infarcts in fronto-parietal lobes and centrum semiovale. Magnetic resonance angio revealed severe short segmental narrowing of both common carotid arteries (CCA) approximately 20 mm on both sides. A transthoracic echocardiogram was done to look for embolic foci, the results were normal and the ejection fraction was 60%.

Aspirin 150 mg, clopidogrel 75 mg, and rosuvastatin 20 mg were administered. After this, a digital subtraction angiography was done which showed 80–90% of luminal narrowing in both

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CCAs. She was then referred to the Rheumatology department owing to a very high suspicion of large vessel vasculitis.

At the rheumatology department, a physical examination revealed high blood pressure of 140/90 mmHg and there was no blood pressure difference between both arms. Pulses were reduced at the neck on both sides and brachial, radial, and pedal pulses were normal. Jaw and temporal tenderness were absent. Significant bruits were heard on both sides over the common carotid but there were no abdominal bruits. Pulmonary and abdominal examinations were within normal limits.

Table 1 shows the raised inflammatory markers of the patient which favor the diagnosis of vasculitis in our patient. Improvement in inflammatory markers post-therapy suggests the effectiveness of treatment. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were ordered and reported values of 46 mm/h and 7.90 mg/L, respectively. Computed tomography aortogram revealed diffuse circumferential smooth wall thickening of bilateral proximal CCA with 5–6 mm of origin of CCA on the left side and 1.5 cm on the right side, which gave the impression of possible arteritis (Fig. 1).

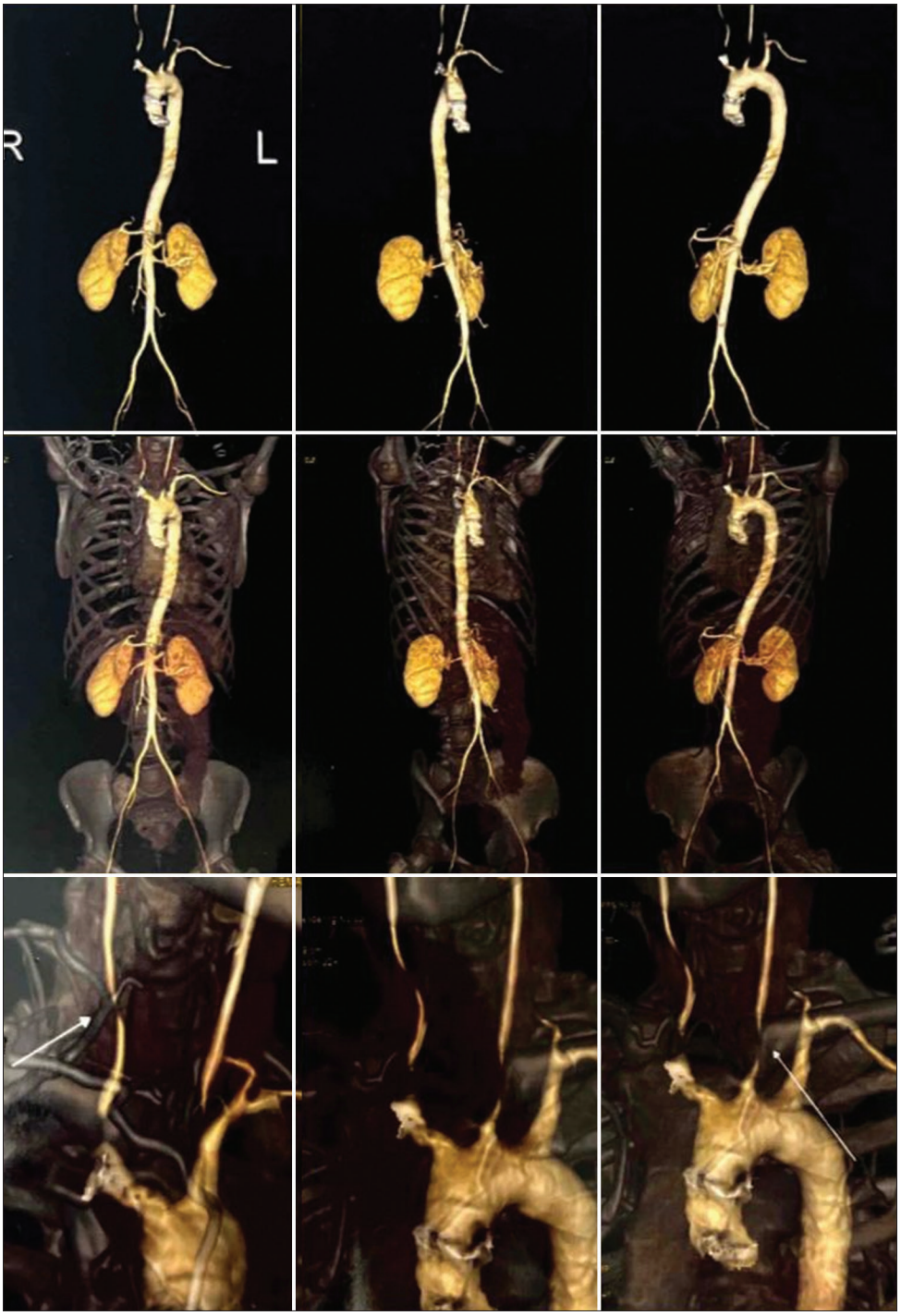

Figure 1: Computed tomography aortogram showing normal aorta and bilateral narrowing of common carotid artery lumen (bottom row arrow)

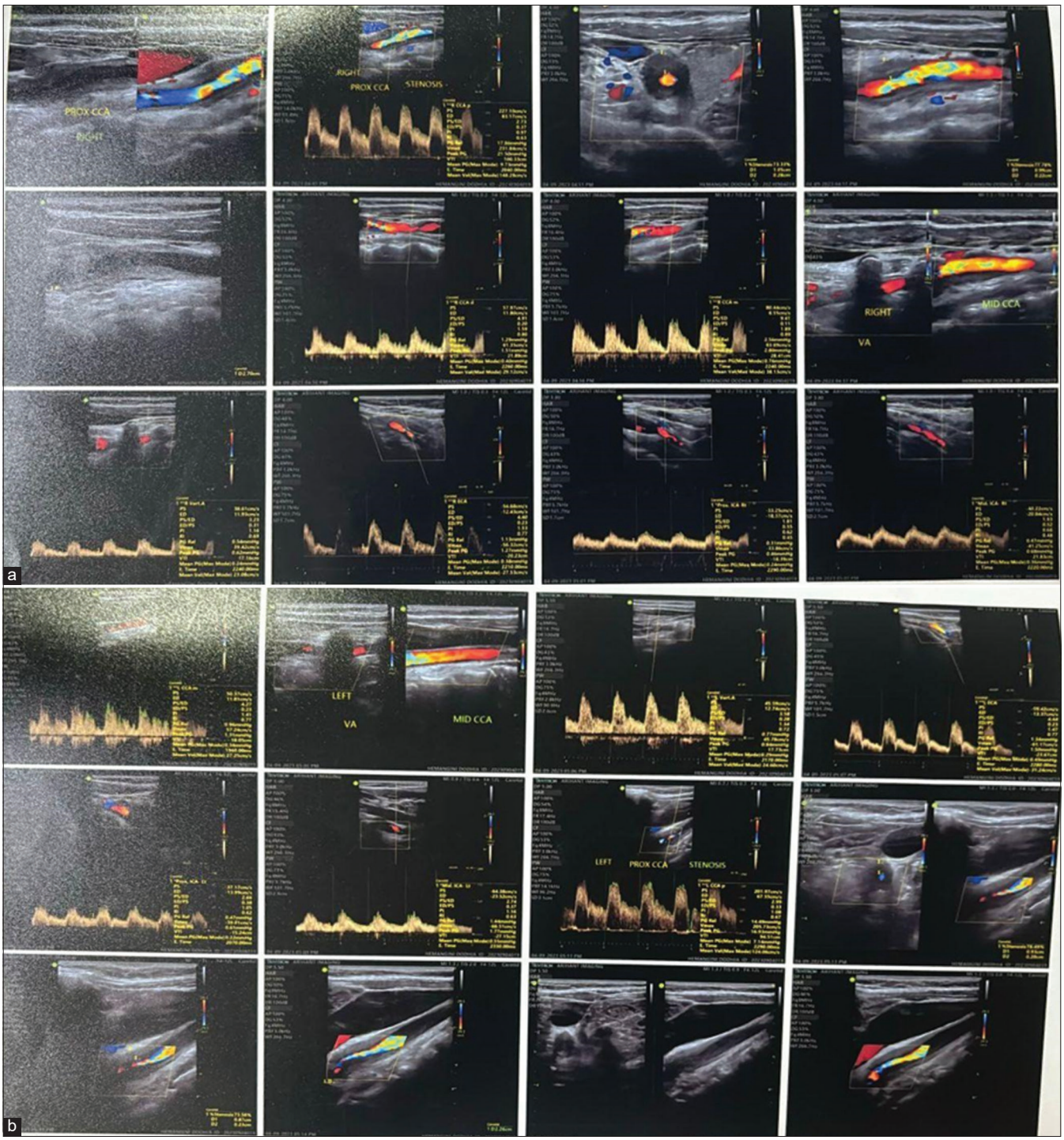

Figure 2: (a) color Doppler velocimetry of right common carotid artery showing reduced flow; (b) color Doppler velocimetry of left common carotid artery showing reduced flow

Table 1: Investigations of the patient

Test	September 11, 2023	June 08, 2024
Triglycerides	161.6	159
Non-high density lipid cholesterol	150	162
Low-density lipid cholesterol	117.68	118.4
Thyroid-stimulating hormone	8.9	7.5
Erythrocyte sedimentation rate	46	12
C-reactive protein	7.9	1.9
Ferritin	43.9	42.8

Due to high clinical suspicion, a diagnosis of TAK was established and she was started on a high dose of prednisone dose – 1 mg/kg body weight and a steroid-sparing agent methotrexate biweekly dose of 15 mg. Physiotherapy was suggested. At regular follow-up, she remained symptom-free with a reduction in ESR and CRP value. At 6 months, color Doppler was repeated which revealed significantly reduced thickening of vessel wall on both sides approximately 2.3 mm was reported (Fig. 2).

DISCUSSION

TAK is a progressively debilitating form of granulomatous inflammation of the large vessels and associated, first described by the Japanese ophthalmologist Dr. Mikito Takayasu in 1908 [7]. The largest vessel of the body - the aorta, its big branches, and coronary, and pulmonary vasculature are among the most often afflicted by the disease.

A recent population study conducted by Sanchez-Alvarez *et al*. concluded the incidence rate of TAK at 0.3–3.3/million new diagnoses new year and its prevalence in society at 4.7–360 cases/ million [8]. Several studies have examined the pathogenesis of Takayasu disease, suggesting a link to a mix of genetic factors, auto-immune response, and vascular remodeling [5,6]. This interplay of immune processes leads to inflammation of vasa vasorum followed by pan arteritis, aortic wall thickening, and occlusion [9,10]. Possible triggers identified include collagen in the vessel wall and cellular antigens such as anti-endothelial cell antigens [4]. Genetic studies on Japanese populations have shown the involvement of various loci associated with Takayasu disease, including immune-regulatory - RPS9/LILRB3, LILRA3, IL38 loci; and inflammatory cytokine genes- IL6 and IL12B loci; as well as the human leukocyte antigen-B52 allele, which are strongly upregulated in acute Takayasu flare-ups [11]. The disease has been associated with tuberculosis [5] and possibly COVID- 19 vaccination [11]. Our patient received three doses of the COVID-19 vaccine, the last dose being in 2022, which may have contributed to her developing Takayasu at the age of 53, possibly explaining the atypical nature of her age at presentation. Though the exact prevalence of TAK is unknown, it is noted to be 3–10× more in younger ages 21–40 than in patients above 40 years of age, and the incidence in pts >40 years of age increases in society as move toward the West [12].

The presentation of TAK is very heterogeneous. This disease goes through phases of blood vessel involvement which are pre-stenotic, stenotic, and post-stenotic. Symptoms are presented according to different stages. Sometimes at the beginning of the disease, the patient may be completely asymptomatic, leading to a delay in diagnosis. Constitutional symptoms are presented in about one-third of cases. Significant symptoms are reported at the occlusive or stenotic phase depending upon the arteries involved, stenosis of the carotid artery would lead to cerebrovascular accidents, renal artery involvement would lead to reduced renal function, and most commonly involvement of subclavian vessels would result in upper limb ischemic symptoms. Reliable physical findings in the occlusive phase include pulselessness and bruits in involved arteries [13]. This can also lead to accelerated atherosclerosis. The most common cause of death in Takayasu is still heart failure [14].

ESR has a sensitivity of 72% and specificity of 56% and reveals systemic inflammation. CRP has a sensitivity of 71.4 percent and a specificity of up to 100%, reflecting the burden of systemic inflammation [13]. ESR is most commonly higher than CRP in Takayasu patients with vascular inflammation. Leukocyte count may be normal or slightly elevated. Usually, in 60% of patients with Takayasu disease, stenosis is seen in the abdominal and thoracic descending aorta and the branches involved are subclavian, common arteries followed by renal arteries. Dilatation and aneurysm can also be found [13]. The case was made interesting by the fact that both CCAs were involved in a South Asian woman, with no involvement of the Aorta or other large vessels. Our patient had involvement of large which supports the diagnosis of Takayasu.

The mainstay of treatment for Takayasu includes steroids (a dose of 1 mg/KG daily for 3–6 months, followed by tapering from 4 to 6 weeks), conventional immunosuppressive agents, such as methotrexate, azathioprine, mycophenolate mofetil, leflunomide, cyclosporine, cyclophosphamide. Biological agents, such as anti-tumor necrosis factor alpha, ANTI IL6, ANTI CD20 monoclonal antibody, IL12/23 inhibitor, Janus kinase inhibitors. Supportive measures include antiplatelets and statins. Our patient was prescribed methotrexate (7.5 mg), prednisone (60 mg), aspirin (150 mg), rosuvastatin (40 mg), clopidogrel (75 mg), and folic acid (5 mg). After 6 months of treatment, our patient showed a significant reduction in wall thickening of the right common carotid artery (2.3 mm thickening) as compared to 20 mm seen 6 months back. Thus displaying the effectiveness of the treatment for our patient.

CONCLUSION

Clinicians should always bear in mind the possibility of large vessel vasculitides, such as TAK as possible differential diagnoses for the evaluation of unexplained neurological symptoms, especially with no active comorbidities and risk factors. Timely diagnostic confirmation and initiation of suitable therapy leads to prompt reduction of symptoms and thus better patient outcomes. This case suggests that TAK may be relatively underdiagnosed in the elderly in the Eastern world.

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