Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Cardiology

Syncope as Unexpected Presentation of Takayasu's Arteritis - A Case Report and Literature Review

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DOI: 10.36347/sjmcr.2022.v10i03.020 | **Received:** 19.02.2022 | **Accepted:** 24.03.2022 | **Published:** 28.03.2022

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Abstract **Case Report**

Takayasu arteritis is an inflammatory disease that affects large vessels, especially the aorta and its branches. The clinical features of the disease depend on which arteries are affected.The syncope as uncommon presentation due to subclavian steal syndrome from Takayasu arteritis. We describe a case of a 36-year-old woman who presented with syncope and was found to have subclavian steal syndrome. We describe the patient's hospital course leading to the diagnosis of TA, which is a rare form of vasculitis. Then, we discuss phases of Takayasu arteritis, explain the mechanism of syncope in this vasculitis, and outline we outline the mainstay of treatment for Takayasu's arteritis.

Key words: Takayasu arteritis, subclavian steal syndrome, syncope, pulmonary hypertension, vasculitis, case report.

INTRODUCTION

Takayasu's arteritis is a chronic inflammatory disease mainly involving the large and medium-sized vessels with a predilection for the aorta and its branches. Clinical manifestations depend on which arteries are affected. Pulmonary arteritis is very uncommon in Takayasu's arteritis. We describe a case of woman with Takayasu arteritis, pulmonary artery involvement, who primarily presented with pulmonary hypertension.

CLINICAL OBSERVATION

The patient was a 36-year-old woman without history of surgeries, drug allergies, or significant medical problems within her family. The patient denied smoking, alcohol use, or recreational drug use. She was diagnosed hypertensive four years ago for which she was currently on treatment with a calcium channel blocker. She had also dyspnoea for 4 years. It was gradual in onset, slowly progressive to dyspnoea on routine activities. Patient also had fatigue on exertion. There was no history of orthopnea or paroxysmal nocturnal dyspnoea. Simultaneously she started having claudication and paresthesis of the right upper limb which also progressed gradually. The patient presented three episodes of syncope three days before her admission. No report of urinary or fecal incontinence, seizure-like activity, or post-event confusion was noted. There was no history of traumatism or any other symptoms specially chest pain.

Physical examination revealed Blood pressure readings on the left and right arms were 115/70 and 88/70 mmHg, respectively; pulse was 90 beats/min, respiratory rate 16 breaths/min, temperature 37.4°C, and oxygen saturation was 98% on room air. The blood glucose by finger stick analysis was 101 mg/dL. Mental status was normal. Right upper limb was pulseless while the left was weak. A left carotid bruit was heard. The patient had equal femoral pulses bilaterally. Harzer sign was positif. First heart sound was normal, 2nd heart sound showed narrow split with loud pulmonary component. There was pan systolic murmur in tricuspid area, which increased on inspiration. The rest of the examination was unremarkable.

Investigations

The complete blood count revealed hemoglobin of 15 g/dL, leucocyte count of 13.900 /mm^3, and a platelet count of 412.000 /mm^3. The patient's chemistries, coagulation profiles, liver function tests, thyroid-stimulating hormone level, study were normal. The initial troponin-T was 4 (99th percentile negative range being (0-13) ng/mL.The patient's erythrocyte sedimentation rate was 72 (normal

0 –10), and her C-reactive protein was negative. The electrocardiogram showed QRS right axis, right bundle block, right ventricular hypertrophy and P pulmonale. The chest X ray revealed Cardiomegaly with right ventricular enlargement, prominent main pulmonary artery and right atrial enlargement.

The patient's echocardiography noted left ventricle normal in size, and good contraction with ejection fraction at 65%. Right atrial and ventricular were enlarged, RV free wall was hypertrophic and contractility was decreased. TAPSE was 14mm. main pulmonary artery was dilated. There was severe tricuspid regurgitation with gradient estimated pulmonary artery pressure to be 160mmHg. Pulmonary function testing noted a mild restrictif lung disease whilst pulmonary scintigraphy objectivated right lung perfusion was scarcely visible in a ventilation-perfusion scintigraphy scan. Her arterial duplex ultrasound of upper limb et supra-aortic vessels showed increasing of the wall thickness circumferentially with inflammatory structure that seems chronic interesting right brachiocephalic trunk. The common carotid arteries were in pre-occluded phase whereas the right subclavian, axillar and vertebral arteries and were totally occluded Figure 1.

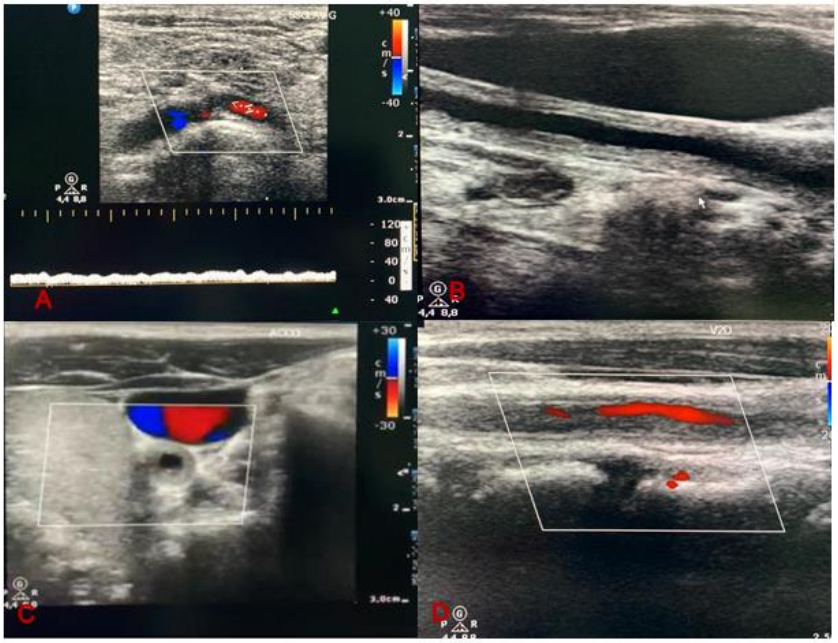

Fig-1: A: Longitudinal section color-duplex vascular sonogram from the patient showing slow flow of the left subclavian artery B: Longitudinal section B-mode showing right common carotid artery with homogeneous, midechoic, circumferential wall thickening ("macaroni sign") with luminal stenosis. Intima-media thickness is 0.22 cm, C:Transverse section B-mode view of right common carotid artery showing concentric intima-media complex thickening ("macaroni sign")and dilatation of the internal jugular vein, D: Longitudinal section color duplex sonogram from the patient showing occlusion of the right vertebral artery.

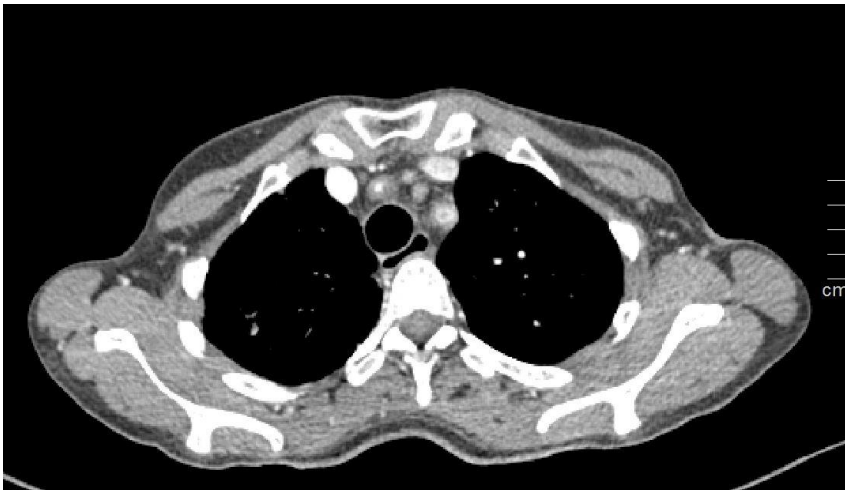

Fig-2: Axial section computed tomography angiography of the supra-aortic vessels of the patient with homogenous, hypodense, circumferential increased vascular wall thickness leading to severe stenosis.

The patient computed tomography angiography of supra-aortic vessels, confirmed the results of duplex ultrasound Figure 2. Whilst the computed tomography angiography of the chest and abdomen revealed a dilatation of the right cardiac cavities, enlargement of the pulmonary artery trunck, an occlusion of the right pulmonary artery Figure 3 and exclude a stenosis of the renal arteries and the abdominal aorta and its branches.

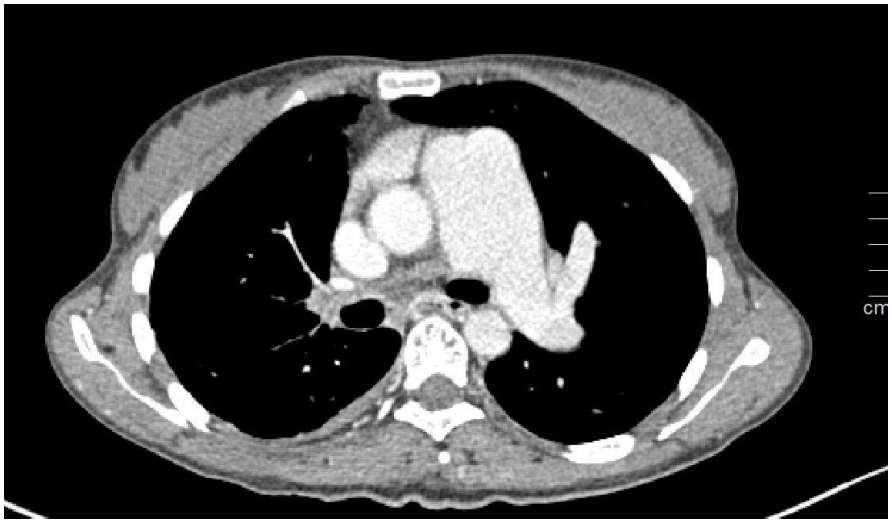

Fig-3: Axial section pulmonary computed tomography angiography of the patient showing the enlargement of the pulmonary artery trunck with occlusion of the right pulmonary artery.

The patient was thus diagnosed with subclavian steal syndrome and was found to have multiple lesions involving the major blood vessels. She was diagnosed with Takayasu's arteritis as per the criteria laid out by the American College of Rheumatology and was started on prednisone. She had an unremarkable hospital course, and was discharged home on prednisone pers os et bolus de tocilizumab, and atorvastatin with appropriate follow-up. At the time of this writing, the patient was doing well.

DISCUSSION

TA is a chronic inflammatory disease of unknown etiology, primarily affecting the aorta and its main branches [1-3].The disease commonly affects women—especially those younger than 40 years of age.

The course of the disease runs in 2 phases: the first is the preischemic phase, characterized by systemic symptoms, such as a general feeling of discomfort, fever, asthenia, sweating, and aching joints; the second, the ischemic phase, commonly appears years later and leads to ischemia in the region supplied by the affected artery [4]. The ischemic stage is characterized by decreased or absent pulse, cardiac murmur, systemic arterial hypertension, aortic valve insufficiency, neurological involvement, and pulmonary hypertension. There are no laboratory findings specific to Takayasu arteritis and a confirmed diagnosis is based on the histopathology of the vessel affected.

The subclavian artery (SA) is the most commonly affected artery in TA [2, 5]. Subclavian steal syndrome results when there is a lesion in the SA proximal to the origin of the vertebral artery (VA). The lesion causes reduction in pressure distal to the lesion [5, 6]. With exercise of the upper extremity on the affected side, blood from the SA, instead of coursing cranially to supply the basilar territory of the brain, flows caudally in a retrograde fashion, draining blood away from the posterior fossa into the upper extremity [5, 6]. Blood thus flows from the contralateral VA via the basilar artery into the ipsilateral VA and then into the SA, compromising posterior circulation. This phenomenon is called subclavian steal syndrome and results in symptoms of compromised posterior circulation such as vertigo and syncope [6].

Treatment of TA involves glucocorticoids. Disease- modifying anti-rheumatic drugs such as methotrexate and tocilizumab has also been used. Control of comorbidities such as hypertension and hyperlipidemia is paramount, as they lead to atherosclerosis, which contributes to the development of subclavian steal syndrome. Surgical options are used when medical management fails. Surgical modalities may include angioplasty, stenting, bypass grafting, and endarterectomy [3, 6, 9].

CONCLUSION

The presented patient had a prior history of a syncopal episode and a chronic history of left arm claudication. The rarity of Takayasu's arteritis and the presentation with non-specific symptoms may contribute to the difficulty in diagnosing the disease earlier [1, 3, 9]. This case report may familiarize health care providers with the rare condition and facilitate prompt diagnosis and treatment of TA.

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