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Amaurosis Fugax and an Incidental Asymptomatic Pulselessness are Cardinal Features of Takayasu Arteritis - Case Report

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Abstract **Case Report**

Background: Takayasu Arteritis (TA) is a rare, chronic large-vessel vasculitis primarily affecting the aorta and its branches. Because the initial phase often presents with non-specific systemic symptoms, early diagnosis remains difficult. Amaurosis fugax (transient monocular visual loss) can serve as a critical sentinel sign of large-vessel involvement, warranting immediate vascular assessment. ***Case Presentation:*** We report the case of a 40-year-old female presenting with a three-month history of recurrent amaurosis fugax in the left eye, exertional dyspnea, and upper limb claudication. Physical examination revealed weak to absent radial pulses and a marked discrepancy in blood pressure between the upper and lower extremities ("pulseless disease"). Initial laboratory investigations revealed only a mild elevation in C-reactive protein (2.11 mg/dL), and a temporal artery biopsy was negative for Giant Cell Arteritis. ***Diagnosis and Management:*** Despite the lack of specific serological markers, CT Angiography (CTA) provided definitive evidence of TA. Imaging demonstrated severe attenuation of the left subclavian artery and diffuse irregular mural thickening of the left common carotid artery. This angiographic pattern differentiated the condition from Fibromuscular Dysplasia. The patient was started on an immunosuppressive regimen comprising prednisolone and methotrexate. At the 20-day follow-up, she reported complete resolution of visual disturbances and claudication. ***Conclusion:*** This case highlights that Takayasu Arteritis should be a primary differential diagnosis in young adults presenting with transient visual loss and pulse deficits. It underscores the necessity of relying on clinical and angiographic evidence for diagnosis, particularly when inflammatory markers are inconclusive and small-vessel biopsies are negative.

Keywords: Takayasu Arteritis, Amaurosis Fugax, Vasculitis, CT Angiography, Pulse Deficit.

1. INTRODUCTION

Transient Monocular Visual Loss (TMVL), also known as amaurosis fugax, is a critical and frequently recurrent presenting symptom that necessitates an urgent workup to rule out serious pathology [1]. In middle-aged patients, this symptom often prompts investigation into common causes such as atherosclerotic carotid disease or cardiac embolic phenomena. However, amaurosis fugax can also be a sentinel sign of less common, severe systemic conditions, notably large-vessel vasculitis [2].

Takayasu Arteritis (TA) is a rare, chronic, granulomatous vasculitis of unknown etiology, primarily affecting the aorta and its main branches. Early diagnosis is inherently challenging, as the initial phase is often characterized by non-specific systemic symptoms (e.g., claudication, dyspnea, chest pain). The disease progresses to cause stenosis, occlusion, or aneurysm formation in the large arteries, frequently leading to pulse deficits and significant blood pressure discrepancies between the upper and lower limbs [3]. We

present a compelling case of a 40-year-old female who presented with recurrent transient visual loss and symptoms of upper limb ischemia. The clinical findings, specifically the weak upper limb pulses, profound differential blood pressure, and characteristic multi-arterial stenosis revealed on CT angiography, led to a diagnosis of TA. This report is significant as it highlights the reliance on angiographic evidence and the distinctive hemodynamic consequences of the disease (pulseless presentation) for diagnosis, even when initial immunological screening and superficial artery biopsy results are negative.

2. CASE PRESENTATION

A 40-year-old female presented with a 3-month history of recurrent, transient episodes of blurry vision and complete vision loss lasting from a few seconds up to several minutes. Her presentation was prompted by a sudden, complete loss of vision in her left eye, which recovered spontaneously. The patient also reported systemic symptoms, including claudication, a painful, taut feeling along the venous tracks over her limbs, recurrent chest pain and tightness, and exertional dyspnea

Her past medical history was significant for an episode of Supraventricular Tachycardia (SVT) during a peripartum period. There was no history of skin or joint involvement.

Physical Examination:

On physical examination, the most striking findings were related to arterial flow:

1. **Pulses: Weak or absent radial and brachial pulses** were noted, with the right side specifically reported as weak.
2. **Blood Pressure Discrepancy:** A significant difference in BP was observed:

- Right arm BP: Irreconcilable
- Left arm BP: 90/50 mmHg
- Right leg BP: 150/90 mmHg
- Right leg BP: 150/90 mmHg
- Left leg BP: 110/70 mmHg

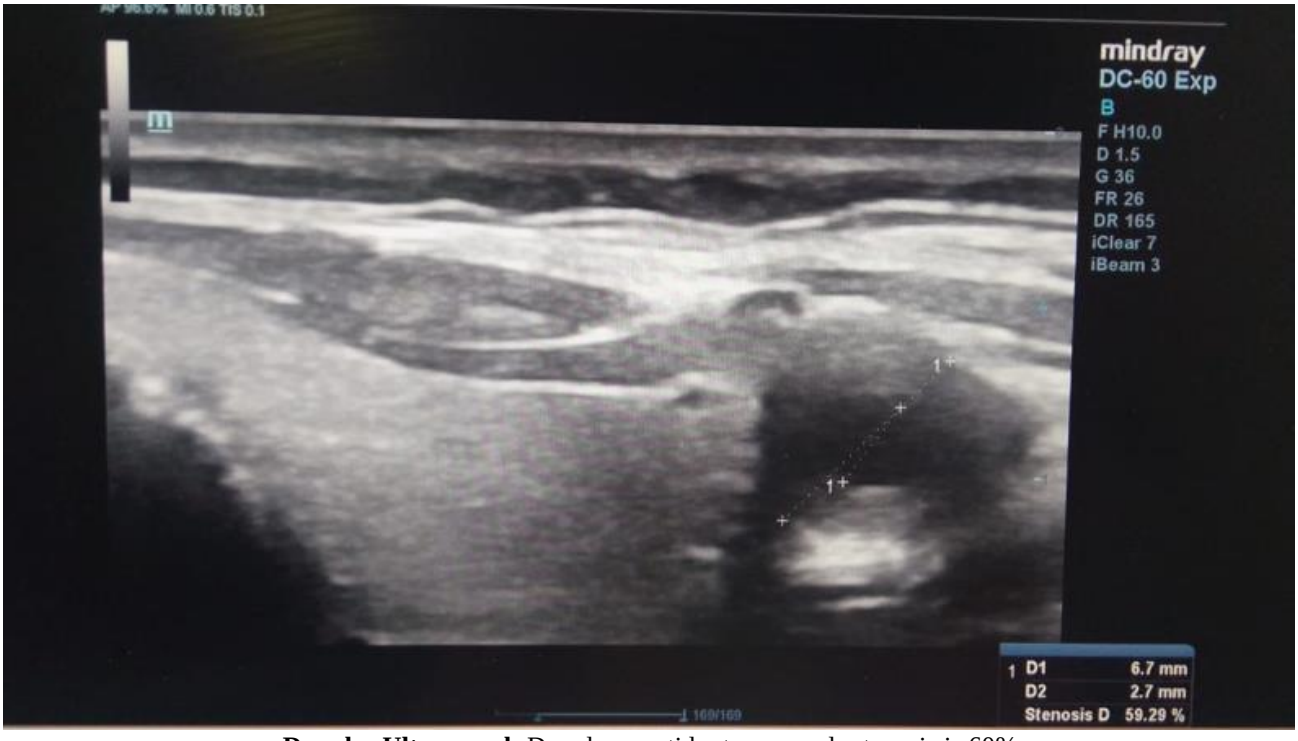
3. **Other Systems:** Eye examination and fundoscopy were normal. Cardiac echocardiogram and enzymes were normal.

3. INVESTIGATIONS

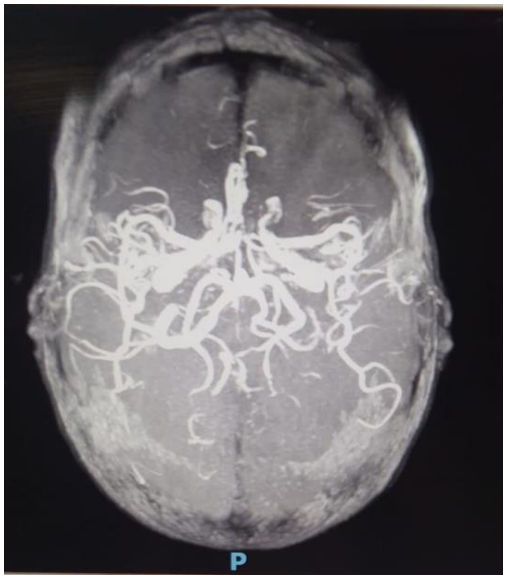
Laboratory and Histology

Initial laboratory evaluation revealed a mildly elevated C-Reactive Protein (CRP) of $2.11 mg/dL. Comprehensive serological screening, including Anti-Cardiolipin Antibody IgG-IgM, ANA, HIV Ag/Ab, and Hepatitis B/C screens, were all negative/non-reactive

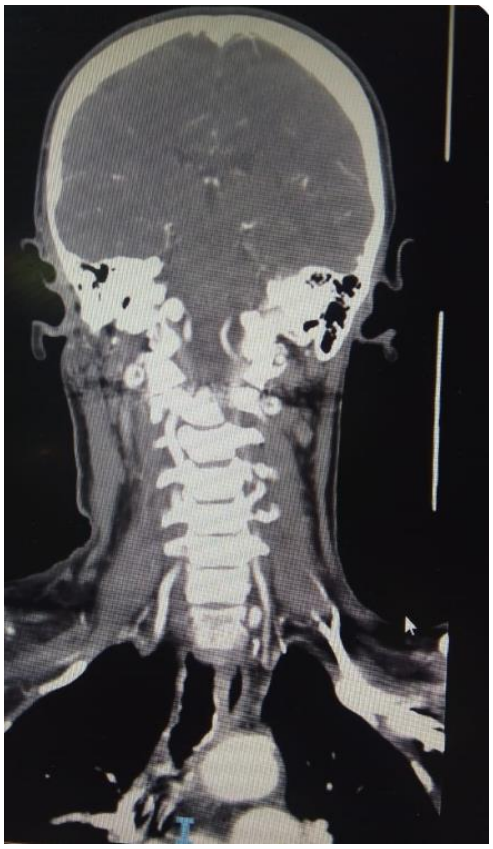
A biopsy taken from the left superficial temporal artery showed No Abnormality Detected (NAD) and no evidence of Giant Cell Arteritis, ruling out GCA.



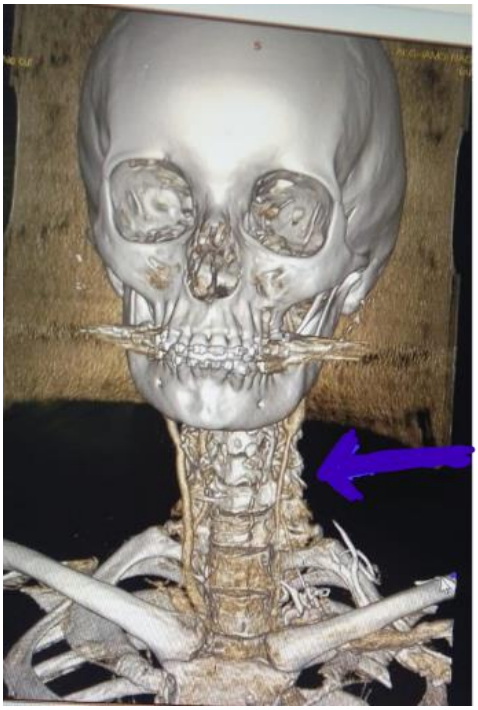
Doppler Ultrasound: Doppler carotid artery reveals stenosis is 60%



MRI/MRV Brain: Demonstrated a normal appearance of the Circle of Willis and no evidence of aneurysm or acute cerebral parenchymal lesions



A coronal view of a CT Angiography (CTA) of the neck and chest inlet is a critical piece of visual evidence that should be included in the Investigations (Section 3) of your report, likely as Figure 7. This image visually confirms the advanced, large-vessel vasculitis of the aortic arch branches, which is pathognomonic for Takayasu Arteritis (TA). Specifically, the Coronal CTA imaging (Figure 7) visibly demonstrates the severe attenuation and occlusion of the left subclavian artery near its origin and illustrates the irregular contours and luminal narrowing of the left common carotid artery, providing direct anatomical support for the radiology reports detailing diffuse wall thickening and a beaded, stenosed lumen. This visual confirmation of the extent and location of the pathology is essential for justifying the clinical diagnosis of TA over other differential diagnoses, like FMD



The 3D reconstruction is exceptionally useful as it offers a superior spatial relationship of the affected vessels. The image visually confirms the pathology reported in the 2D slices and reports: the arrow points to the general area of the significant narrowing and irregularity of the left common carotid artery and the corresponding proximal involvement of the great vessels, which is highly characteristic of the Takayasu Arteritis that was diagnosed in the patient. This figure adds significant instructional value by showcasing the "beaded" appearance and the severity of the arterial wall thickening in the context of the patient's anatomy

4. DIFFERENTIAL DIAGNOSIS AND FINAL DIAGNOSIS

The constellation of transient monocular vision loss (amaurosis fugax), pulse deficits in the upper limbs, claudication, and a striking differential BP between limbs pointed strongly towards a disease affecting the large vessels. Primary considerations included Fibromuscular Dysplasia (FMD) and Large-Vessel Vasculitis (Takayasu Arteritis)

The angiographic findings ultimately supported the diagnosis of Takayasu Arteritis (TA). The characteristic features of diffuse wall thickening and occlusion of the proximal left subclavian artery and diffuse irregular mural wall thickening along the entire left CCA (Figure 4) satisfied the criteria for TA, despite the negative temporal artery biopsy (Figure 2) and low-level inflammatory markers

5. TREATMENT AND OUTCOME

The patient was referred to Rheumatology, and the case was clinically labeled as Takayasu Arteritis. The patient was immediately initiated on a definitive immunosuppressive regimen for active vasculitis:

Glucocorticoids: Prednisolone and Immunosuppressive Agent: Methotrexate.

At a follow-up 20 days post-treatment initiation, the patient reported a complete resolution of her symptoms: no further vision deterioration and no new attacks of transient visual loss or claudication were noted.

6. DISCUSSION AND CONCLUSION

This is the point we want to highlight, consider uncommon causes of amarousus. Through history and physical exam, supported by extensive radiological studies, this case emphasizes that a definitive diagnosis of Takayasu Arteritis requires a high index of suspicion and can be made based on clinical and angiographic evidence, even when traditional inflammatory markers (CRP) and small-vessel biopsy (Temporal Artery) are non-diagnostic [4]. The rapid clinical improvement following the initiation of immunosuppressive therapy further validates the diagnosis of an active inflammatory process.

The systemic nature of TA means that relying solely on serological findings can lead to significant diagnostic delays, as demonstrated by our patient's mildly elevated CRP 2.11 mg/dL and otherwise negative immunological screening (Figure 1), as well as the negative temporal artery biopsy [5].

The critical features leading to the TA diagnosis over FMD were the involvement and occlusion/diffuse wall thickening of the proximal large-vessels (subclavian

artery and entire common carotid artery) and the profound pulse and BP discrepancy indicative of significant flow limitation from the aortic arch branches.

In conclusion, recurrent transient visual loss with pulse deficits is a medical emergency requiring urgent imaging to evaluate the great vessels. Takayasu Arteritis remains a key differential diagnosis, and a lack of highly elevated inflammatory markers should not preclude a diagnosis when classic angiographic findings are present

The critical features leading to the TA diagnosis over Fibromuscular Dysplasia (FMD) the involvement and occlusion/diffuse wall thickening of the proximal large-vessels (subclavian artery and entire common carotid artery) [6].

7. ETHICAL CONSIDERATION

Written informed consent was obtained from the patient involved in this case report. The patient was fully informed about the nature of the research and agreed to the use of their clinical data without mentioning personal details.

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