CASE REPORT

Case Report: Takayasu arteritis in a male patient [version 1; peer review: 1 approved with reservations]

Syed Mohammad Mazhar Uddin¹, Aatera Haq¹, Zara Haq², Uzair Yaqoob ³, Osama Mohiuddin², Anosh Aslam Khan²

¹Department of Medicine, Civil Hospital Karachi, Karachi, Sindh, Pakistan
²Dow University of Health Sciences, Karachi, Sindh, Pakistan
³Department of Medicine, Jinnah Postgraduate Medical Centre, Karachi, Sindh, Pakistan

Abstract

Takayasu arteritis (TA) is a type of primary systemic vasculitis mainly affecting the medium and large arteries. The signs and symptoms are due to systemic inflammation or ischemia of an organ or limb and include angiodynia, claudication, peripheral pulselessness, murmurs, ischemic stroke, myocardial infarction and severe systemic arterial hypertension. The disease tends to affect more women than men. Here we present a case of TA in a 22-year-old male patient. Our patient presented with complaints of aphasia and right-sided weakness, with on-and-off symptoms of malaise, generalized weakness, unilateral headache, fatigue and shortness of breath lasting two years. Color Doppler ultrasound was sufficient for a diagnosis of TA, after which we started the patient on medical treatment and also consulted the department of vascular surgery. Overall, this case report highlights the importance of screening for TA in male patients so that the diagnosis is not overlooked, and also adds more data to the limited literature on male patients.

Keywords

Takayasu Arteritis, Autoimmune, autoimmune in males, vascular disease
Corresponding author: Uzair Yaqoob (ozair_91393@hotmail.com)

Author roles: Uddin SMM: Conceptualization, Data Curation, Formal Analysis, Funding Acquisition, Investigation, Methodology, Project Administration, Resources, Software, Supervision, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing; Haq A: Conceptualization, Data Curation, Formal Analysis, Funding Acquisition, Investigation, Methodology, Project Administration, Resources, Software, Supervision, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing; Yaqoob U: Conceptualization, Data Curation, Formal Analysis, Funding Acquisition, Investigation, Methodology, Project Administration, Resources, Software, Supervision, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing; Haq Z: Conceptualization, Data Curation, Formal Analysis, Funding Acquisition, Investigation, Methodology, Project Administration, Resources, Software, Supervision, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing; Mohiuddin O: Conceptualization, Data Curation, Formal Analysis, Funding Acquisition, Investigation, Methodology, Project Administration, Resources, Software, Supervision, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing; Khan AA: Conceptualization, Data Curation, Formal Analysis, Funding Acquisition, Investigation, Methodology, Project Administration, Resources, Software, Supervision, Validation, Visualization, Writing – Original Draft Preparation, Writing – Review & Editing

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Introduction

Takayasu arteritis (TA), also known as “pulseless disease”, is a type of primary systemic vasculitis affecting medium and large arteries, including the aorta and its branches, as well as the pulmonary and coronary arteries. It is a chronic inflammatory disease of unknown origin characterized by granulomatous vasculitis, leading to thickening, dilatation, stenosis, and/or aneurysm formation of the involved vessels. Furthermore, the signs and symptoms exist due to systemic inflammation or ischemia of an organ or limb, and encompass angioinflammation, claudication, peripheral pulselessness, murmurs, ischemic stroke, myocardial infarction, severe systemic arterial hypertension, etc. TA tends to affect females more than males, with 80% of patients being female. However, the female-to-male ratio varies, from 9:1 in Japan and 6.9:1 in Mexico to 1.2:1 in Israel. Furthermore, TA is associated with significant morbidity and can be life-threatening. Around 20% of patients experience monophasic and self-limited disease, whereas others can have a progressive or relapsing/remitting disease. Moreover, the overall 10-year survival rate for this disease is approximately 90% which can be reduced in the presence of major complications. Here we present a case of TA in a 22-year-old male.

Case report

A 22-year-old male college student, belonging to Interior Sindh province, with no known history of comorbid presented to the emergency department of civil hospital, Karachi, Pakistan, on 3rd January 2019, with an insidious onset of aphasia and right-sided weakness for six days. According to the patient’s brother, he was in his usual state of health six days back when he developed an altered level of consciousness, which led them to bring him to hospital. In the hospital, the patient regained consciousness on the day of admission but was unable to speak and move his right side of the body. There was no additional history of fever, fits, nausea, vomiting, urinary incontinence, fecal incontinence, as well as any visual symptoms. However, the patient had complained of malaise, generalized weakness, shortness of breath and fatigue for the last two years. Furthermore, the patient also complained of intermittent unilateral headache which was not relieved by over-the-counter pain medications. The patient had no other significant past medical, surgical and family history. On arrival, his blood pressure was 140/90 in the left arm but non-recordable in the right arm, pulse was 60 beats/minute (only recordable in the left arm), and temperature 36.7°C and respiratory rate 22 breaths/minute. On examination, his general physical, respiratory and abdominal examinations were unremarkable. However, central nervous system examination revealed problems, with aphasia in speech. Motor examination on the right side revealed reduced power in both upper and lower limbs as well as upgoing plantars on the right lower limb. The rest of the central nervous examination was normal. His cardiovasculard examination showed muffled heart sounds, and no murmurs were present, but he was positive for bilateral carotid bruit.

Based on the history and examination, we ordered pertinent laboratory tests along with other specific tests. When his baseline laboratory values were ordered, complete blood count (CBC) showed hemoglobin (Hb) of 12.3 g/dL (normal: 11.1–14.5 g/dL), mean corpuscular volume of 77.5 fL (normal, 76–96 fL), WBC count of 7.5×10^9/L (normal: 4–10×10^9/L) and platelets were 180×10^9/L (normal, 150–400×10^9/L); however, C-reactive protein was 140 mg/L (normal <3 mg/L) and erythrocyte sedimentation rate was 80 mm/hour (normal, 0–22 mm/hour). As the patient had intermittent shortness of breath, we also performed a chest x-ray and echocardiography, both of which were normal. The patient’s coronary and renal angiogram was also normal. As the carotid bruit was audible bilaterally, we also performed carotid artery color Doppler imaging (evaluating the common carotid artery (CCA), internal carotid artery (ICA), external carotid artery (ECA) and vertebral artery (VA)). Findings showed diffuse homogenous intimal thickening involving the bilateral CCA, ICA, and ECA, demonstrating macaroni sign, causing marked luminal narrowing. Other findings include minimal flow and reduced peak systolic velocity (PSV) in the right CCA and ICA on power Doppler and no flow on color and power Doppler images in the right ECA. The right VA was dilated with normal flow and no flow was seen in the PSV, left proximal and mid CCA on power Doppler images (suggestive of complete occlusion). However, left ICA and ECA showed minimal flow and reduced PSV on power Doppler (possibly due to collateral supply), left VA shows no flow (suggestive of complete occlusion) and there was turbulent flow in the aortic arch. Overall, on the basis of clinical manifestations and carotid artery Doppler findings, a diagnosis of TA was made. We started the patient on oral prednisone (60 mg) once daily on a tapered basis and consulted the department of vascular surgery for a possible surgical intervention. The patient had an endarterectomy two weeks after admission which led to an improvement in his symptoms, after which he was discharged on 5th February 2019, and no follow-up has since been conducted.

Discussion

TA can be seen in a broad geographical area, but it is mainly found in Asia and Africa. The nature of the disease is autoimmune, involving arterial walls resulting in panarteritis. According to the American Rheumatological society, for diagnosing TA, out of the following six clinical findings, three are compulsory: (i) Onset before 40 years; (ii) Claudication of the extremities; (iii) Decrease in the brachial pulse in one or both arms; (iv) Difference of 10 mmHg or more in blood pressure measured in both arms; (v) Audible bruit on auscultation of the aorta or subclavian artery; and (vi) Narrowing at the aorta or its primary branches on arteriogram. Our patient met at least five of the above criteria. He was a 22-year-old male, and literature supports the fact that it is common in second and third decades. Moreover, in adults, almost 80% of the patients are women.

The disease progresses in two phases; the early active phase that lasts weeks to months, giving constitutional symptoms and can have relapse and remission, and the late chronic phase which is caused by arterial stenosis along with ischemia and occlusion of organs. The clinical illustrations can be different based on the location of arterial lesions (Table 1).

Our patient presented primarily with symptoms of the aortic branches, such as malaise, absent pulse on the right upper extremity and headache.
A diagnosis of TA is primarily based on clinical and radiological findings, as the results of biopsy are nonspecific as the histopathology may imitate other types of vasculitis. Suspected TA always warrants prompt vascular imaging, enabling earlier diagnosis and further decreasing the risk to the patient. Although angiography was considered to be the standard method for diagnosis of TA, it has been replaced by computed tomography angiography or magnetic resonance angiography. Furthermore, literature has shown that ultrasound with color Doppler flow imaging and angiography are highly useful for detecting and determining the severity of the disease (except for right brachiocephalic artery). However, in our setting, due to limited resources, we only conducted carotid artery Doppler imaging, and the findings were sufficient to achieve a diagnosis of TA. As far as treatment is concerned, immunosuppressants such as prednisone and/or methotrexate can lead to significant improvement. Cyclophosphamide is reserved for treatment-resistant cases; a prior study revealed that steroids and cyclophosphamide are effective in the early acute phase when surgical management is not considered. However, in the presence of symptomatic occlusive lesions (fibrotic phase), procedures such as bypass grafts, patch angioplasty, endarterectomy, percutaneous transluminal angioplasty, or stent placement should be considered. Furthermore, despite the ongoing medical treatment, 50% of patients with TA progress to a stage that requires one or more surgical procedures. In our case, we started the patient on 60 mg prednisone orally daily, but due to the stenotic lesions, department of vascular surgery was also consulted for a possible endarterectomy.

Conclusion
Overall, TA is a rare disease (especially in men) that is both diagnostically and therapeutically challenging to physicians. Early diagnosis is very important in this debilitating disease, in order to improve the outcomes. As far as the role of patient gender and prognosis is concerned, the literature is sparse, and more studies should be conducted. Furthermore, although medical treatment is considered as the mainstay for TA, it is imperative to apprehend both the indications and the available options of surgical interventions.

Data availability
All data underlying the results are available as part of the article and no additional source data are required.

Consent
Written informed consent for publication of their clinical details was obtained from the patient.

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References

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Samuel Katsuyuki Shinjo
Division of Rheumatology, Faculdade de Medicina FMUSP, Universidade de São Paulo, São Paulo, Brazil

• Please give us more information about a possible involvement of thoracic and abdominal aortic and its main branches.
• Was the patient treated only with prednisone?
• Table 1. Please correct the sentence ("Temporal arteritis").
• Discussion: should be more explored; There is no relevance to describe the "TA classification criteria" in the Discussion; the relevance of the present case report should be more explored throughout the text.

Is the background of the case's history and progression described in sufficient detail?
Partly

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?
Partly

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?
Partly

Is the case presented with sufficient detail to be useful for other practitioners?
Partly

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: Inflammatory myopathies; systemic vasculitis.
I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

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