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CASE REPORT

Case Report: Takayasu arteritis in a male patient [version 1; peer review: 2 approved with reservations, 1 not approved]

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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

Open Peer Review

Approval Status ? ? X

	1	2	3
version 1	?	?	X
25 Mar 2019	view	view	view

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2. **Enrico Tombetti** , University of Milan, Milan, Italy
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Any reports and responses or comments on the article can be found at the end of the article.

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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 arteries¹. 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^{1,2}. Furthermore, the signs and symptoms exist due to systemic inflammation or ischemia of an organ or limb, and encompass angiodynia, claudication, peripheral pulselessness, murmurs, ischemic stroke, myocardial infarction, severe systemic arterial hypertension, etc¹. 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 Israel³. 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 complications⁴. 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 cardiovascular 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 $7.5 \times 10^9/L$ (normal: $4\text{--}10 \times 10^9/L$) and platelets were $180 \times 10^9/L$ (normal, $150\text{--}400 \times 10^9/L$); 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 thickness 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^{1,3}. 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 (6) narrowing at the aorta or its primary branches on arteriogram^{2,3}. Our patient met at least five of the above criteria. He was a 22-year-old male, and literature^{2,5} supports the fact that it is common in second and third decades. Moreover, in adults, almost 80% of the patients are women².

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)^{2,3}.

Our patient presented primarily with symptoms of the aortic branches, such as malaise, absent pulse on the right upper extremity and headache.

Table 1. Clinical presentation of Temporal Arteritis based on arterial location.

Vessels involved	Clinical features
1. Aortic branches	Malaise, decreased or absent pulse of upper extremities, dysfunction of upper extremities, headaches, dizziness, vision and orientation disturbances, syncope.
2. Aortic arch	Congestive heart failure, aortic valve insufficiency, arterial hypertension.
3. Coronary arteries	Ischemic heart disease, myocardial infarction.
4. Pulmonary arteries	Chest pain, dyspnea, coughing, hemoptysis, congestive heart failure.
5. Abdominal aorta or celiac trunk	Ischemia of the stomach and intestines, abdominal pain, nausea, vomiting
6. Renal arteries	Arterial hypertension, chronic renal failure

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 vasculitis⁶. 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 angiography or magnetic resonance angiography². 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)^{2,3,6}. 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 improvement³. 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 considered⁶. 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^{3,6}. Furthermore, despite the ongoing medical treatment, 50% of patients with TA progress to a stage that requires one or more surgical procedures⁶. 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.

Grant information

The author(s) declared that no grants were involved in supporting this work.

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Open Peer Review

Current Peer Review Status: ? ? X

Version 1

Reviewer Report 07 January 2021

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Tetsuro Yokokawa

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Authors demonstrated a young male patient of Takayasu arteritis. This is a rare case report diagnosed by using ultrasound imaging. This reviewer thinks this report has lack of novelty and have some concerns as shown below.

1. Some patients with Takayasu arteritis are male. Ultrasound imaging is usual examination for the diagnosis for vascular diseases. What is the novelty of this case report?
2. Takayasu arteritis should be diagnosed with imaging for inflammation such as positron emission tomography (PET). PET imaging is useful to monitor of immunosuppressive treatment. Did this patient undergo PET?
3. Authors should show the ultrasound images as Figures.
4. Authors should show laboratory data including liver and kidney function in a new Table.
5. This patient had elevated C-reactive protein. How did authors exclude infectious diseases?
6. This patient underwent endarterectomy. Was the pathological finding from the endarterectomy sample compatible for Takayasu arteritis? Please provide the pathological image as a Figure.

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: Cardiovascular diseases

I confirm that I have read this submission and believe that I have an appropriate level of expertise to state that I do not consider it to be of an acceptable scientific standard, for reasons outlined above.

Reviewer Report 07 January 2021

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Enrico Tombetti

Department of Biomedical and Clinical Sciences-L. Sacco, University of Milan, Milan, Italy

The author report a young male with neurological symptoms that was diagnosed of TA.

The neurological presentation is interesting, but I have some comments.

Major

- Treatment: the patients opt for single therapy with steroids. This is unusual, since TA has frequently a polyphasic course, as the authors disclose. Considering the risk of recurrences and the severity of presentation, I would have used upfront combination therapy with steroids & immunosuppressive agents (e.g.: methotrexate). If the authors wants to presents this case, they should discuss better therapy, explain why they have not considered immunosuppressive agents, and acknowledge that this would not have been the preferred therapeutic choice in most centers.
- Disease characterisation. I agree that with US data there is enough evidence to diagnose TA. However, I think that this patient deserve further disease characterisation, given the severity of presentation and considering that the patient underwent endoarterectomy, I would have studied him with antio-MRI or angio-CT. Has this been done? If yes, I would describe findings. if not, this should be discussed highlighting that optimal management should include, if possible CT or MR, unless in case of burn-out mild disease.

- Outcome of management: The authors simply report that the symptoms have improved. This is too vague. I would add a) postoperative results on carotid disease, b) clinical outcomes, and which symptoms have improved and the degree of improvement. In the case of follow-up data, discussion of the outcome may be performed at follow-up.
- Follow-up. I wonder if the authors have already performed the first follow-up assessment or not. If yes, please describe results. Otherwise, please describe how you plan to follow the patient.

Minor

- Past medical history should be better described.
- The authors correctly avoided to perform surgery without medical therapy. This point deserves further discussion in the light of literature data.
- "However, central nervous system examination revealed problems" this sentence should be rephrased.
- "with normal flow and no flow was seen in the PSV," I cannot understand this sentence. In general US findings are difficult to be read, and I suggest to rephrase this section.

References

1. Tombetti E, Mason J: Takayasu arteritis: advanced understanding is leading to new horizons. *Rheumatology*. 2019; **58** (2): 206-219 [Publisher Full 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?

Yes

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

Partly

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: 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.

Reviewer Report 17 May 2019

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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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