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EC NEUROLOGY
Case Report

32 Years Old Female with Pulseless Carotid

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Abstract

Takayasu arteritis (TA) is a chronic inflammatory arteritis affecting large vessels, predominantly the aorta and its main branches. Wall thickening, stenosis, fibrosis and thrombus formations is the hallmark of the vessel's inflammation seen in TA. Aneurysmal formation occurs in response to acute inflammation leading to arterial media destruction. Both sexes can be affected however it is predominately seen in female.

TA has a broad clinical spectrum of presentation varying form asymptomatic disease to devastating neurological impairments. The diagnosis of TA remains to be a challenge. The clinical presentation, negative vasculitis markers and classical radiological manifestations are all mandatory to aid to the diagnosis.

TA is a rare disease most seen in Japan, South East Asia, India and Mexico. Despite the increasing identification of young adult with TA, reports of disease in young population are still mingy practically at our part of the world. Her we report a 32 years old female from the middle east with TA.

Keywords: *Takayasu Arteritis; Vasculitis; Stenosis; Thrombus; Aneurysm; Diagnosis; Stroke; Immunosuppressant Agents*

Introduction

Takayasu arteritis (TA) is a granulomatous large vessel vasculitis that involves the aorta, its major branches, pulmonary renal and coronary arteries. It is also known as pulseless disease [1] thromboaortopathy or Martorell syndrome [2].

Panarteritis is the histological hallmark of this disease. Where all the arterial wall layers are involved, including intimal fibrous thickening and or typical atheromatous lesions. Destruction of medial smooth muscles and elastic layers, cellular infiltration and collagenous fibrosis in the media and thickened adventitia with cellular infiltration around vasa vosurum [3,4].

TA causes board spectrum of clinical presentation ranging from a symptomatic to devastating ischemic presentations including blindness and severe neurological deficits [5-12]. Diagnosis of TA remains challenging due to the non-specific symptoms, its systemic nature, multiorgan involvement and disease morbidity related to the cardiovascular, central nervous and renal systems.

Case Report

32-year old woman, referred to stroke clinic following two neurological episodes. The first episode was sudden onset slurry speech lasted for 24 hours and followed by complete recovery. A month later she had a dense left upper limb weakness that improved gradually but with residual deficit. She gave no other symptoms apart from significant unintentional weight loss (15 kg/last 6 months) and nonspecific bilateral lower limbs joints pain. Her past medical and medications history were both negative.

Her clinical examination revealed; thin built woman, BP 154/95 mmHg, HR: 91/min regular, RR 20/min, temperature of 36.9°C.

Pulseless right carotid artery, week left carotid artery pulse with significant bruit. Weak pulses of left brachial artery and bilateral popliteal arteries. Her neurological was unremarkable except for Left upper limb hemiparesis with power of 4+/5 (proximally as well as distally). Otherwise she had normal carinal nerves, sensory and cerebellar exam. Systemic examination was unremarkable otherwise.

Laboratory investigations showed C-reactive protein (CRP) of 13.7 mg/L and erythrocyte sedimentation rate (ESR) 26 mm/h. Normal complete blood count (CBC), renal and liver function tests. Her bone profile was also normal. Vasculitis work up including (ANA: anti-nuclear antibodies, ANCA: Anti-Neutrophil-cytoplasmic antibodies, antiphospholipid antibodies, rheumatoid factor, hepatitis B&C, ACCP IgG antibodies, ENA: Extractable nuclear antigens and Quantifer on TB gold test) were all negative. In addition, the thrombophilia screening was negative.

Initial brain CT: Showed recent cerebral infarct along the territory of the right middle cerebral artery (mainly lenico-striate arteries) with total right internal carotid occlusion. Carotid Doppler: right common carotid artery (CCA) and right internal carotid artery (ICA) have intraluminal echogenicity with absence of color flow. Left CCA shows increase intima media thickness at its proximal part with 50% stenosis.

CT angiography of neck vessels: Minimal wall thickening of aortic arch extending to the origin of brachiocephalic trunk and origin of left common carotid. Circumferential wall thickening and complete luminal occlusion of the right internal carotid artery (green arrow). Circumferential wall thickening and significant luminal obstruction of the left internal carotid artery (blue arrow).

Diagnosis: TA based on clinical presentation, negative vasculitis markers and classical radiological manifestations.

Treatment: Patient was started was on aspirin and anti-lipids agents and methylprednisolone. Reviewed by rheumatology team, subsequently she was kept on prednisolone and Mycophenolate Mofetil. Patient was referred to Neuro-vascular surgeon for possible further intervention. However, no surgery was planned.

Follow up: She continued to follow up with neurology, rheumatology, and vascular surgery clinics. Her last follow up with stroke clinic was few weeks ago through a phone call. She lives withing an hour and quarter of an hour by plane. Given the COVID-19 pandemic circumstances were all domestic flights are on hold. She reported no new symptoms.

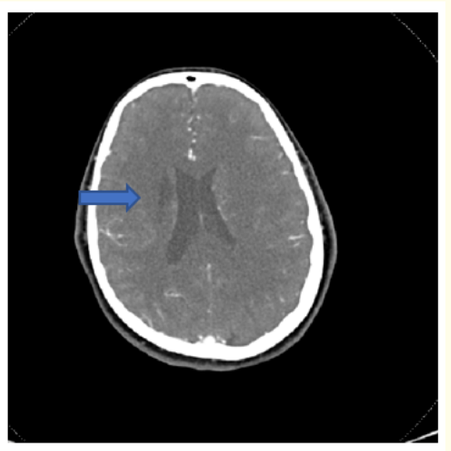

Figure 1: CT head: *showing cerebral infarct along the territory of the right middle cerebral artery (mainly lenico-striate arteries)*

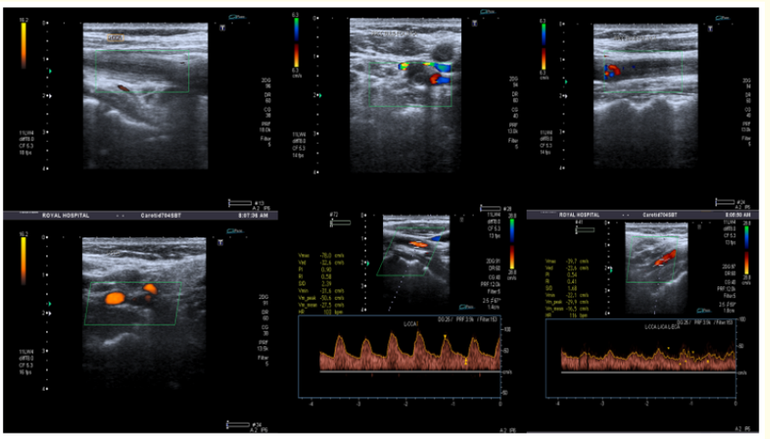

Figure 2: Carotid Doppler: *right common carotid artery (CCA) and right internal carotid artery (ICA) have intraluminal echogenicity with absence of color flow. Left CCA shows increase intima media thickness at its proximal part with 50% stenosis*

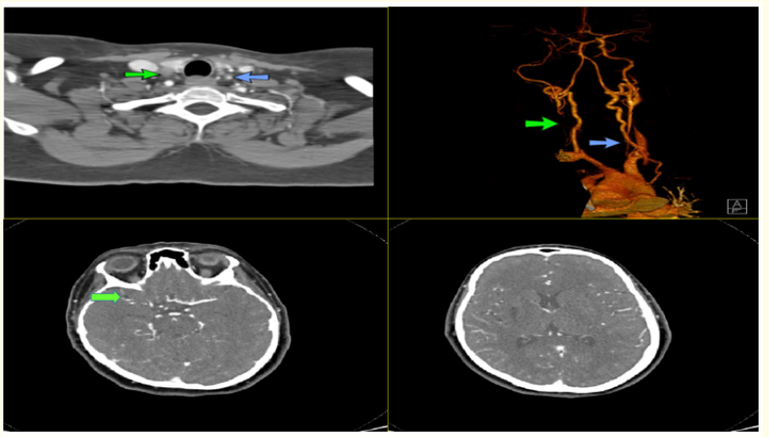

Figure 3: Cerebral CT Angiogram

Minimal wall thickening of aortic arch extending to the origin of brachiocephalic trunk and origin of left common carotid. Circumferential wall thickening and complete luminal occlusion of the right internal carotid artery (green arrow) Circumferential wall thickening and significant luminal obstruction of the left internal carotid artery (blue arrow).

Discussion

Although TA symptoms and sings may present as early as the second or third decade of life. The clinical variations of this disease presentations have led in most of the cases to significant delay in diagnosis establishment. The time interval between the onset of the very first symptom of TA and the diagnosis can vary form months to years [2,8]. It took around 12 months to diagnosis our patient as she was seen before referral to us; at two other hospitals. One outside the country and two others in Oman.

Fever, malaise, myalgia, weight loss and mild anaemia are the most common nonspecific features of the this disease [5]. However diminished or absent pulses, vascular carotid. subclavian or abdominal vessels bruits, aortic regurgitation. Takayasu retinopathy, congestive cardiac failure, dilated cardiomyopathy, seizures, stroke and other more are the main characteristic features of the disease [2,5,6,8,9,13].

In 1990 the American College of Rheumatology (ACR) defined specific criteria for the diagnosis of TA [14]. Some of these features can mimic other diagnosis. Therefore, it is especially important to exclude other differential diagnosis such as tuberculosis, syphilis, fibromuscular dysplasia, and giant cell arteritis [8,15]. The diagnosis of TA requires companying the clinical presentation, negative vasculitis markers and classical radiological manifestations. Angiography remains the gold standard for diagnosis.

The treatment of the TA can be divided into medical and surgical. The medical arm of the treatment includes steroids [5,7,9], cyclophosphamide, azathioprine methotrexate and mycophenolate [10,12,16-18]. The main goal of the treatment is to control the disease activity and preserve vascular competence. At least 50% of patients treated with steroids will response. The treatment with other immunosuppressant agents demonstrated variable response based on studies. There are not enough studies to favour one over the other. The decision should be tailored based on the tolerance of side effect profile of each drug. The response can be judged by the clinical response as well the decline in a follow up inflammatory markers.

There is also a necessity to treat stroke when it happens in patients with TA. This would include offering thrombolysis and thrombectomy in the hyperacute phase of ischemic stroke. TA is not a contraindication for such treatment [19-21]. The other options will be dual antiplatelet agents initially when presenting late for thrombolysis and or thrombectomy therapy. Later the use of single antiplatelet agent and ant lipid agents will help maximize the risk of re-stroking. Controlling hypertension is also mandatory although it might be challenging since the use of steroids.

Our patient was referred to our outpatient stroke clinic few months after the onset of her neurological symptoms and sings. She was admitted at the stroke unit for evaluation of stroke in the young. The evaluation lead to the diagnosis of TA and the initiation of the appropriate treatment and follow up visits.

Surgical treatment

The surgical intervention is considered mainly cerebrovascular disease or critical stenosis of three or more cerebral vessels to prevent stroke [22,23]. Other indication is hypertension and renal artery stenosis [23]. Our patient was referred for evaluation of possible anastomoses by neurosurgery, however neurosurgeon decided against that.

Conclusion

This case illustrates a severe presentation of a rare disease worldwide and particularly in our region. TA can present with an uncommon complaint in young adult and can progress to a point where the long-term survival is in jeopardy. Our patient's CT angiography was

particularly impressive for the amount of stenosis and collateralization required to maintain blood supply to the brain. It is important to suspect TA in young women who present with focal weakness or cryptogenic stroke. Early recognition. treatment and radiological intervention and regular follow up can alert the course the disease. In the case of aggressive disease. absence of follow up or non-compliance, the disease would most likely prove to be fatal.

Bibliography

1. Shimizui K and Sano K. "Pulseless disease". *Neuropathology and Clinical Neurology* 1 (1951): 37-47.
2. Lupi-Herrera E., *et al.* "Takayasu arteritis. Clinical study of 107 cases". *American Heart Journal* 93 (1977): 94-103.
3. Koide K. "Takayasu arteritis in Japan". *Heart Vessels* 7 (1992): 48-54.
4. Numano F. "Takayasu's arteritis". *Lancet* 356 (2000): 1023-1025.
5. Hall S., *et al.* "Takayasu arteritis. A study of 32 North American patients". *Medicine* 64 (1985): 89-99.
6. Ishikawa K. "Natural history and classification of occlusive thromboaortopathy (Takayasu's disease)". *Circulation* 57 (1978): 27-35.
7. Shelhamer JH., *et al.* "Takayasu's arteritis and its therapy". *Annals of Internal Medicine* 103 (1985): 121-126.
8. Subramanyan R., *et al.* "Natural history of aortoarteritis (Takayasu's disease)". *Circulation* 80 (1989): 429-437.
9. Kerr GS., *et al.* "Takayasu arteritis". *Annals of Internal Medicine* 120 (1994): 919-929.
10. Moriwaki R., *et al.* "Clinical manifestations of Takayasu arteritis in India and Japan-new classification of angiographic findings". *Angiology* 48 (1997): 369-379.
11. Jain S., *et al.* "Takayasu arteritis in children and young Indians". *International Journal of Cardiology* 75 (2000): S153-S157.
12. Sato EI., *et al.* "Takayasu arteritis. Treatment and prognosis in a University Center in Brazil". *International Journal of Cardiology* 75 (2000): S163-S166.
13. Sharma S., *et al.* "The incidence and patterns of pulmonary artery involvement in Takayasu's arteritis". *Clinical Radiology* 42 (1990): 177-182.
14. Arend WP., *et al.* "The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis". *Arthritis and Rheumatology* 33 (1990): 1129-1134.
15. Michel BA., *et al.* "Clinical differentiation between giant cell (temporal) arteritis and Takayasu's arteritis". *The Journal of Rheumatology* 23 (1996): 106-111.
16. Hoffman GS., *et al.* "Treatment of Takayasu's arteritis (TA) with methotrexate (MTX)". *Arthritis and Rheumatology* 34 (1991): S74.
17. Hoffmann GS., *et al.* "Treatment of glucocorticoid resistant or relapsing Takayasu arteritis with methotrexate". *Arthritis and Rheumatology* 37 (1994): 578-582.
18. Daina E., *et al.* "Mycophenolate mofetil for the treatment of Takayasu arteritis: report of three cases". *Annals of Internal Medicine* 130 (1999): 422-426.
19. The National Institute of Neurological Disorders and Stroke rt-PA Stroke Study Group. "Tissue plasminogen activator for acute ischemic stroke". *The New England Journal of Medicine* 333 (1995): 1581-1587.
20. Werner Hacke., *et al.* "Thrombolysis with Alteplase 3 to 4.5 Hours after Acute Ischemic Stroke". *The New England Journal of Medicine* 359 (2008): 13.
21. William J Powers., *et al.* "Guidelines for the Early Management of Patients With Acute Ischemic Stroke: A Guideline for Healthcare Professionals From the American Heart Association/American Stroke Association (2018).
22. Giordano JM., *et al.* "Experience with surgical treatment of Takayasu's disease". *Surgery* 109 (1991): 252-258.
23. Giordano JM. "Surgical treatment of Takayasu's arteritis". *International Journal of Cardiology* 75 (2000): S123-S128.

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