Case Report

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Takayasu Arteritis Presented with Unilateral Vision Loss : A Case Report

Syed Md Jabed[1*] Shaila Sharmin[2] Md Ferdous[3] Puja Saha[4]

ABSTRACT

Background: *Takayasu Arteritis (TA) is a rare chronic granulomatous inflammation of the aorta or its branches and is prevalent all around the world. It causes stenosis of large arteries and ischaemic damage to target organs. There is usually a delay in recognising TA because of the rarity and unfamiliarity with the disease, unspecific early symptoms and lack of diagnostic equipment for early diagnosis. The purpose of the study to reminding readers of this issue and emphasize the necessity for further research on how to deal with in the best way.*

Case Report: *We report a case of 30 years old male presented on 22nd August 2023 at Institute of Applied Health Sciences (IAHS) Chattogram with gradual loss of vision of left eye over a month accompanied by headache and later on diagnosed as a case of Takayasu's Arteritis.*

Conclusion: *Visual loss is the most common ocular symptom in Takayasu arteritis. Since a variety of causes may result in reduced vision in Takayasu arteritis. A careful history is very important to ascertain the cause of visual loss in Takayasu arteritis patients to reduce unnessary investigations and referrals.*

Key words : Ischemic vision loss; Large vessel vasculitis; Takayasu Arteritis (TA).

Introduction

Takayasu Arteritis (TA) often known as pulseless disease, is chronic inflammatory arteritis characterized by damage to the medium and large arteries as well as their branches. The aorta and its primary branches, particularly the renal, carotid, and subclavian arteries, are commonly affected, resulting in stenosis, occlusions or aneurysmal degeneration of these large vessels.[1] Despite the fact that the disease is widespread throughout the world, the Asian population is thought to be far more affected. In Japan, the greatest known prevalence of Takayasu arteritis was assessed to be 40 per million while in the United States, the lowest known frequency was reported to be 0.9 per million.[2] The typical onset of symptoms occurs between 10 and 45 years of age.[3,4] The pathophysiology is not fully understood but involves T-cell dominated infiltration of the arterial wall tissue leading to vascular damage, stenosis and even aneurysmatic dilatation.[3,5] Depending on the quality and location of lesions, the disease may manifest with a wide variety of symptoms, laboratory findings and organ involvement. During the disease's acute phase, patients may also have symptoms such as limb weakness or pain, headaches, syncopal attacks and uneven blood pressure. Fever, weight loss and exhaustion are examples of constitutional symptoms that may occur first.[6] The purpose of the study to disseminated our knowledge and experience of clinical characteristics, presentation and treatment about this issue for the readers as future references.

1. Professor (cc) of Medicine
Institute of Applied Health Sciences (IAHS) Chattogram.
2. Associate Consultant of Medical Retina
Chittagong Eye Infirmary and Training Complex (CEITC) Chattogram.
3. Assistant Professor of Medicine
Institute of Applied Health Sciences (IAHS) Chattogram.
4. Medical Officer of Medicine
Bangabandhu Memorial Hospital (BBMH) Chattogram.

*Correspondence : **Dr. Syed Md Jabed**
Cell : +88 01717 16 03 88
Email : bappycmc@gmail.com

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Case Report

A 30-years-old smoker, non-diabetic male presented on 22nd August 2023 at Institute of Applied Health Sciences (IAHS) Chattogram with gradual loss of vision in his left eye over one month.He has a history of dull, persistent headache,occasional blackouts, early fatigue during activities mostly while using hands and a low grade intermittent fever for last 3 months. On examination ,he was mildly anemic, no oedema, thyroid not palpable, all pulses absent in both upper limbs, blood pressure was found160/100 when measured at thigh, temperature 99^0 F. His visual acuity fall to Perception of Light (PL) in the left eye, apex beat not shifted, no murmur, bruit present at the neck, abdominal or renal bruit was absent, kidney not ballotable. Fundoscopy revealed early optic atrophy on left eye. Takayasu disease was suspected. On routine

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Investigation ESR – 90 mm in 1st hour, Hb%- 9 g/dl, S. creatinine- 0.9 mg/dl, Urine R/E – normal, CXR – Cardiomegaly. CT-Angiogram revealed wall thickening and luminal narrowing at the origin of brachiocephalic trunk, left common carotid and left subclavian arteries suggesting large vessel vasculitis-Takayasu Arteritis. Patient was treated with iv methylprednisolone for 3 days followed by oral prednisolone combined with methotrexate for disease control and amlodipine for blood pressure and aspirin as antiplatelet. His headache, fatigue and feverish feelings were improved but there was no immediate visual improvement. He was counselled and discharged on 27th August 2023 with advise to follow up.

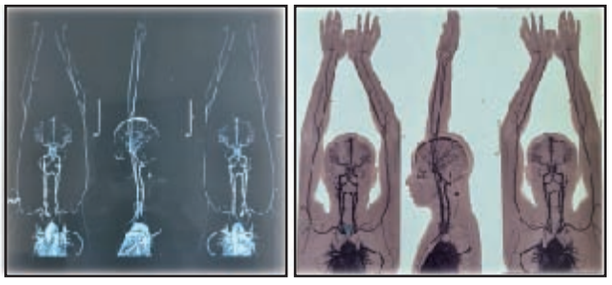

Figure 1, 2 CT angiogram showing narrowing at the commencement of brachiocephalic and left common carotid artery

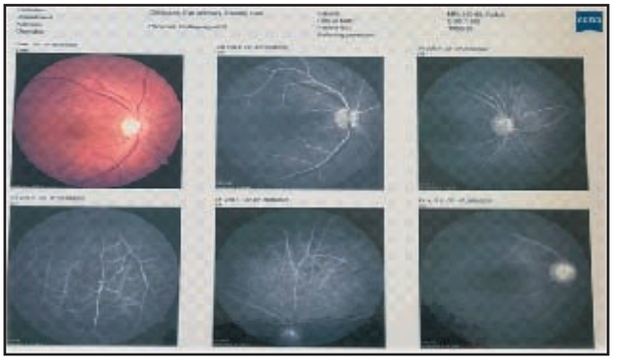

Figure 3 FFA left eye showing delay in choroidal and arteriovenous filling with an increase in foveal avascular zone due to capillary drop out

Discussion

Takayasu arteritis is a difficult condition to deal with. Early identification of this disease is difficult and necessitates clinical suspicion and vigilance. It is a granulomatous inflammatory vasculitis of the medium and large arteries that results in transmural fibrous thickening of the arterial walls, causing vascular blockages and ischemic changes and primarily affects aorta and its principal branches.[1] Women are affected in 80-90 % of cases with age of onset is usually between 10 to 40 years.[7,8] It has a worldwide prevalence with greatest prevalence in Asia.[9,10] Non-specific symptoms include fever, night sweats, tiredness, weight loss, arthralgia, myalgiaand mild anemia.[9] As the inflammation progresses, more stenoses form and the symptoms and effects become increasingly severe, leading to complications. Takayasu retinopathy, secondary hypertension, aortic regurgitation, and aneurysm formation were the four most common sequelae observed in Takayasu patients.[11] Early clinical signs of TA typically reflect a general inflammatory state, while advanced stages are characterized by findings related to vascular damage, such as neurological symptoms, peripheral pulselessness, bruits or blood pressure discrepancies.[3] Clinical manifestations, laboratory values, mainly inflammatory markers, and diagnostic imaging are used to diagnose Takayasu arteritis. Angiography is still the gold standard for diagnostic and treatment planning.. The use of Doppler ultrasound to assess vascular wall inflammation is a non-invasive approach. Histological diagnosis is frequently impractical due to the vessels involved and histological testing is limited to those patients undergoing revascularization procedures.[12] There are six criteria defined by the American College of Rheumatology to diagnose Takayasu arteritis, presence of three or more out of the six criteria is known to have a sensitivity of 90.5% and specificity of 97.8%.[8]

In this case, our patient presented with low grade fever for 3 months which reflects a chronic infection or inflammation. In a country like ours the first differential diagnosis of such presentation is tuberculosis although a recent review study showed fever was the most common presenting symptom of Takayasu arteritis (20.93%).[13] He also complained headache, not responding to conventional treatment, also a common presentation of this disease. (13.95%).[13] It might be due to high blood pressure found in our patient or due to vascular involvement. Exclusion of tubercular meningitis clinically is also necessary in our settings and was done in this patient. Occassional blackouts, early fatigue of upper limbs (Claudication) support vascular stenosis and was confirmed by absence of pulses in upper limbs.[8] The symptom that bring our patient to reach the diagnosis is his rapid deterioration of vision of left eye to near blindness within one month,which forced him to visit eye specialist who rightly suspected Takayasu arteritis having the ischemic retinopathy by fundoscopy and FFA (Figure 3) and thus referred. Although retinopathy is not uncommon in Takayasu arteritis and there are a few case reports regarding this, blindness is rather an uncommon presenting feature.

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Management of Takayasu arteritis aims at suppressing the inflammatory process and preservation of vascular capability. Corticosteroids and methotrexate are a commonly used anti-inflammatory therapy which we rightly used in this patient with immediate symptomatic improvement of fever, fatigue and headache but not the blindness.Its happen as approximately 20% of patients with Takayasu arteritis are resistant to any therapy and despite good adherence to immunosuppressive medications, remission occurs only in approximately half of cases. We advised the patient to follow up routinely so that we can monitor his response and titrate drugs accordingly.

Conclusions

The diagnosis of Takayasu arteritis in this case presented was reached after ocular ischemia-induced blindness had set in. Ocular ischemic syndrome is a hypoperfusive retinopathy resulting from chronic hypoperfusion of the ophthalmic artery secondary to carotid artery stenosis. Before blindness, he had 2 months of non-specific presentations of chronic illness without much attention. This case demonstrates the enigmatic character of Takayasu arteritis and echoes the importance of a high index of suspicion for early diagnosis.

Disclosure

All the authors declared no competing interests.

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