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A RARE CASE STUDY, TAKAYASU'S ARTERITIS PRESENTATION WITH ALOPECIA AREATA

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Takayasu's arteritis is a rare, idiopathic, stenotic, chronic granulomatous vasculitis with strong predilection for aorta and its major branches. This report is about a 30 -year-old lady who presented with headache, body aches, bilateral progressive vision loss, hair loss, pain abdomen and multiple skin ulcers. Her vascular findings suggested inflammatory vascular granulomatous disease. She would later be diagnosed as having Takayasu's arteritis.

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INTRODUCTION

Takayasu's arteritis is also known as aortic arch syndrome and pulseless disease. It's a rare idiopathic condition of granulomatous arteritis which affects primarily aorta and its medium- and large-sized branches which includes proximal portions of pulmonary, coronary and renal arteries as well.

Takayasu's arteritis is a pan arteritis with initial mononuclear cell infiltrations including giant cells in the adventitia. This is followed by marked proliferation in intima which leads to granulomas formation in the media followed by disruption of the elastin layer and subsequent medial and intimal fibrosis and vascularisation of media. This process results in segmental luminal narrowing , thrombosis, occlusion and aneurysmal dilations in the affected vascular segments. There are diminished and absent pulses in around 90 percent of patients which manifests as limb claudication and blood pressure discrepancies between two arms/legs. These symptoms are secondary to the affected artery and could be life threatening [1]. Involvement of the carotid artery results in ophthalmic artery hypo perfusion and results in ocular ischemic syndrome [2].

This report is about a 30-year-old lady who presented with headache, body aches, bilateral progressive vision loss, hair loss, pain abdomen and multiple skin ulcers. She would later be diagnosed as a case of Takayasu's arteritis.

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CASE REPORT

A 30-year-old lady presented with gradually worsening headache and body aches over the past one year. Body aches were pronounced at shoulders, upper back and upper limbs. In addition, she reported a gradual decline in her vision in both eyes along with hair loss and pain lower abdomen. The patient medical history was significant for generalized fatigue, numbness and pain in the upper extremities particularly in the upper right arm that worsened on exertion. Brachial and radial arterial pulses of the right side were absent on palpation. However, on the left side, they were present but diminished. Blood pressure couldn't be recorded in either arm. Ophthalmic examination demonstrated visual acuity of counting finger in both right and left eyes at 2 meters. Retinal examination revealed microaneurysm, dot and blot haemorrhages, extensive area of ischemia in both eyes and neovascularization at optic disc in the right eye. Diagnosis of *bilateral ocular ischemic syndrome* was made by the ophthalmologist. Fundus flourescein angiogram couldn't be performed.

Dermatological examination confirmed the diagnosis of *alopecia areata* for the hair loss pronounced at late frontal area. Multiple skin ulcers, marked at both arms and upper chest, were labelled as *pyoderma gangrenosum* by the dermatologist. Later, they were confirmed by biopsy report.

Laboratory tests reported raised ESR at 87 mm/hr (normal value less than 20 mm/hr) and serum C-reactive protein of 12.95 mg/l (normal value is less than 5 mg/l). Rest of the laboratory tests including anti-nuclear antibodies (ANA) were insignificant except raised total leukocyte count and pus cells in the urine routine examination test which were attributed to the lower urinary tract infection.

Carotid Doppler ultrasonography revealed homogenous circumferential thickening of both, right and left common carotid artery, with marked occlusion on the right. CT Angiogram findings were circumferential wall thickening of aortic arch, its all branches and descending thoracic aorta. In addition, severe long segment stenosis of brachiocephalic trunk, bilateral common carotid, bilateral subclavian and right axillary artery was reported. Moreover, multiple dilated collateral vessels in posterior thoracic and anterior abdominal walls were observed which manifested as marked back ache and lower abdominal pain.

This patient was initially prescribed oral steroids which would be tapered. She might be considered for immunosuppressants such as Methotrexate if her symptoms do not improve with steroids therapy alone. Meanwhile, this patient is referred to the ophthalmologist for possible intervention for her retinal manifestations- retinal photocoagulation. This patient is consulted with vascular surgeon as well who will plan surgical intervention- carotid endarterectomy on the right side.

DISCUSSION

Takayasu's disease has a wide geographic distribution seen more commonly in Asia and Africa particularly in India and Japan. It is an autoimmune disease involving the arterial walls of medium and large arteries, causing pan arteritis.

Our patient, a 30-year-old lady was fulfilling five out of six criteria set by The American Rheumatological Society.

Takayasu's disease is predominantly a disease of young adults in their second and third decades of life. However, it is not uncommon in children and adults over 40-year-old [3,4,5]. Patients as young as 6-month-old and as old as 75-year-old have been reported [6]. Female-to-male ratio vary among patients in different geographical area ranging from 9:1 in Japan to 1.2:1 in Israel.

Takayasu's arteritis presents with nonspecific clinical manifestations. The clinical course of the disease is comprised by an early active inflammatory phase and late chronic phase. The active early inflammatory phase lasts for weeks to months and may have a remitting and relapsing course. It is characterized by systemic disease manifesting as constitutional symptoms such as fever, headache, malaise, night sweats, arthralgia along with dizziness and skin rashes. However, this phase is not present in all patients except in children with Takayasu's arteritis where constitutional symptoms are often seen.

Diagnosis of Takayasu's arteritis is rarely made in the early active phase. Evidence of vascular inflammation such as tenderness and bruit along the course of arteries may suggest diagnosis of Takayasu's arteritis

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in early phase of the disease process [3]. Late chronic phase present as stenosis and occlusion with subsequent ischemia of organs [3,7].

Our patient presented with constitutional symptoms along with rare dermatological presentation such as *Alopecia areata* in addition to usual findings such as *Pyoderma gangrenosum.* This patient also reported bilateral vision loss, a common presentation of the disease with abdominal pain attributed to the stenosis induced ischemia.

Imaging studies are required to confirm diagnosis in suspected cases of Takayasu's arteritis. With advances in imaging modalities, contrast angiography is no longer commonly used despite being a gold standard for the diagnosis. Instead, computed tomography angiography (CTA) and magnetic resonance angiography (MRA) is commonly used.

Treatment of Takayasu's arteritis is mainly based on steroids such as prednisone. In addition, other immunosuppressants such as methotrexate are effective in halting the disease progression and eliminating its inflammatory activity. Adjunctive therapy with anti-tumor necrosis factors such as etenarcept and infliximab have also shown efficacy in active relapsing disease [8].

In the presence of manifestations of stenosis and occlusion, endovascular revascularization procedures are considered. This varies from bypass graft to endarterectomy and percutaneous transluminal angioplasty to stent placement. However, a cautious approach is advised as Takayasu's arteritis is associated with high failure rates relating to surgical and endovascular procedures especially if these procedures are undertaken during inflammatory phase of the disease [9,10,11].

Like other causes of ocular ischemic syndromes, prompt attention is warranted in ophthalmological complications of Takayasu's arteritis to avoid long term irreversible effects such as permanent blindness.

Inflammatory nodules, Erythema nodosum and pyoderma gangrenosum being the most common skin manifestations of Takayasu's arteritis. Alopecia areata is a very presenting sign of Takayasu's arteritis. These lesions relieve upon therapy with steroids and immunosuppressants [12].

CONCLUSION

Skin conditions such as pyoderma gangrenosum and alopecia areata in a young lady should requires high index of suspicion for Takayasu's arteritis in the presence of constitutional symptoms provided all other causes for the above conditions are ruled out.

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