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Takayasu Arteritis: A Rare Clinical Entity

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Abstract:

Takayasu's arteritis is an inflammatory disease often affecting the ascending aorta and aortic arch, causing obstruction of the aorta and its major arteries. The disease commonly presents in the 2nd or 3rd decade of life, often with a delayed diagnosis. The disease is progressive and there is no definitive therapy. Glucocorticoids and immunosuppressive agents have been reported to be effective in some patients during the active phase. We report a case of Takayasu arteritis who responded well to glucocorticoids and methotrexate.

Key words: Aorta, Glucocorticoids, Immunosuppressive Agents, Methotrexate, Takayasu Arteritis.

Introduction

Takayasu's arteritis (TA) is a rare chronic inflammatory arteritis affecting the large vessels in the body predominantly the aorta and its main branches. The disease results from an attack by the body's own immune system, causing inflammation in the walls of arteries. The inflammation leads to narrowing of the arteries, and this can compromise blood flow to the affected organ or part of the body. TA can result in a weak pulse or loss of pulse in arms, legs and organs. For this reason, people used to refer to the illness as "pulseless disease".

Case Report

A 32 year old female was referred to our hospital with complaints of recurrent episodes of dizziness and vertigo for last 2 years. This was accompanied by blurring of vision, black outs and recurrent syncopal attacks. She had also history of weakness and pain on exertion in both upper limbs. On examination peripheral pulses were weakly palpable in lower limbs but not palpable in upper limbs with unrecordable blood pressure. Both carotids were weakly palpable with audible bruit.

On investigation, patient had mild anaemia, with elevated CRP and ESR. MR angiogram revealed non-visualization of bilateral carotid, subclavian arteries and innominate arteries. Based on the presenting symptoms, clinical findings, investigations and abnormal angiogram a diagnosis of Takayasu arteritis (type 1) was entertained. FDGPET scan was done to look for the activity and extent of the disease that also revealed active inflammatory disease involving arch of aorta, bilateral common

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carotid and bilateral subclavian arteries. Patient was put on oral methotrexate 15 mg weekly along with oral prednisolone 40 mg per day. The methotrexate dose was gradually increased upto 25 mg weekly. With treatment, patient showed marked improvement in her symptoms.

Discussion

TA is a chronic inflammatory disease of unknown etiology affecting medium and large vessel arteries. Although TA has been described worldwide, it occurs most commonly in Japan, China, India, and Southeast Asia. The first case of TA was described in 1908 by Dr. Mikito Takayasu (Japanese ophthalmologist) as a wreathlike appearance of blood vessels in retina. TA affects 3rd-4th decade of life. Females are affected eight times more frequent than men. TA is characterized by granulomatous inflammation of the aorta and its major branches, leading to stenosis, thrombosis and aneurysm formation. The lesions of TA are segmental, patchy and involving all three layers of vessels.

Although the presenting manifestations of TA are protean, the vast majority of patients present with symptoms and signs of vascular insufficiency (from stenosis, occlusion, or aneurysm), systemic inflammation, or both. The most common presenting vascular symptoms are claudication (35%), reduced or absent pulse (25%), carotid bruit (20%), hypertension (20%), carotidynia (20%) light-headedness (20%), and asymmetrical arm blood pressures (15%). Stroke, aortic regurgitation, and visual abnormalities are present at onset in less than 10% of patients [1].

The 1990 ACR criterion for the classification of TA [Table 1] remains the most widely applied [2]. Ishikawa defined clinical groups based on the natural history and complications of the disease [Table 2]. The four most important complications were defined as Takayasu retinopathy, secondary hypertension, aortic regurgitation, and aneurysm formation, each being graded as mild/moderate or severe at the time of diagnosis [3]. New angiographic classification of Takayasu arteritis [Table 3], that allows a comparison of patient characteristics according to the vessels involved and is helpful in planning surgery, but offers little by way of prognosis [4].

Diseases which can mimic TA are rheumatic (giant cell arteritis, Cogan's syndrome, relapsing polychrondritis, ankylosing spondylitis, rheumatoid arthritis, systemic lupus erythematosus, Buerger's disease, Behçet's disease), infectious (syphilis, tuberculosis) and others (atherosclerosis, ergotism, radiation-induced damage, retroperitoneal fibrosis, inflammatory bowel disease, sarcoidosis, neurofibromatosis, congenital coarctation or Marfan's syndrome).

Table 1:1990 American College of Rheumatology criteria for the classification of TA [2]

1	Age at onset ≤ 40 years
2	Limb claudication
3	Diminished brachial pulse
4	Difference of > 10 mmHg systolic pressure between arms
5	Bruit over the subclavian artery or aorta
6	Abnormal angiogram
For diagnosis ≥ 3 criteria should be present (Sensitivity: 90.5%, Specificity: 97.8 %)	

Table 2: Ishikawa clinical classification of Takayasu arteritis [3]

Groups	*Clinical features*
Group I	Uncomplicated disease, with or without pulmonary artery involvement
Group IIA	Mild/moderate single complication together with uncomplicated disease
Group IIB	Severe single complication together with uncomplicated disease
Group III	Two or more complications together with uncomplicated disease

Table 3: New angiographic classification of Takayasu Arteritis, Takayasu conference 1994 [4]

Type	*Vessel involvement*
Type I	Branches from the aortic arch
Type IIA	Ascending aorta, aortic arch and its branches
Type IIB	Ascending aorta, aortic arch and its branches, thoracic descending aorta
Type III	Thoracic descending aorta, abdominal aorta, and/or renal arteries
Type IV	Abdominal aorta and/or renal arteries
Type V	Combined features of types IIB and IV
Involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+), respectively.	

Diagnosis is mainly based on physician awareness, along with a high index of suspicion. If TA is suspected, it is essential to palpate peripheral pulses, listen for bruits, and measure blood pressure in all four limbs. Patient should be evaluated for evidence of an acute phase response (elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) and normocytic-normochromic anaemia [5], and the diagnosis is confirmed on imaging. Conventional arteriography is gold standard but invasive. CT and MR angiography have largely replaced the arteriography because of noninvasive technique, useful in early stage and follow up. 18F-Fluorodeoxyglucose (18F-FDG) PET imaging is increasingly being used in patients with large vessel vasculitis, allowing more precise anatomic location of metabolic activity with enhanced sensitivity, particularly in the event of moderate FDG accumulation. The principal advantage of FDG PETCT is the diagnosis of early pre-stenotic disease, an event that can be missed by intra-arterial angiography [6].

Majority of the patients have good initial response to steroids (oral prednisolone 1 mg/kg/day) with 80-90% remission but about half of the patients relapse when the drug is tapered or stopped, despite good initial response [7]. Immunosuppressants (methotrexate, azathioprine, MMF, etanercept and infliximab) are used in these patients that enable the dose of corticosteroids, reduce their adverse effects and increase the rate of sustained remission.

The indications for considering intervention include uncontrolled hypertension due to renal artery stenosis, severe symptomatic coronary artery or cerebrovascular disease, severe aortic regurgitation, stenotic or occlusive lesions resulting in critical limb ischemia, and aneurysms at risk of

rupture [8]. Common surgical interventions are angioplasty and reconstructive surgery.

Conclusion

TA is a rare clinical entity, can present in wide variety of ways, many with a typical history of other conditions. The use of steroids along with immunosuppressant like methotrexate is a cost effective regimen and has a good initial response. However, chances of relapse should be kept in mind while treating the patient. Regular follow up is necessary for the assessment of the disease progression.

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