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A CASE REPORT ON TAKAYASU'S ARTERITIS

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Abstract:
It is a type of primary systemic vasculitis mainly affecting the medium and large arteries. It is a pulseless disease and chronic inflammatory arteritis affecting the aorta and its main branches it is rare condition affecting in women in the 2nd and 3rd decades of life. The signs and symptoms are due to inflammation or ischemia of an organ and include vascular pain, claudication, peripheral pulselessness, murmurs, myocardial infarction and severe systemic arterial hypertension. The disease more affect in women than men. We here by discuss an interesting case of a young lady presenting complaints pain over right side of the neck over arterial aspect since 7 months, throbbing type gradually progressive radiating to right side of the face, chest pain, breathlessness, abdomen pain, fever, loss of appetite, blurring of vision. Color Doppler ultra sound was sufficient for the diagnosis of TA after which we started the patient on medical patient with corticosteroids and immunosuppressive agents have been reported to the effective in some patients during the active phase and this is probably youngest case in hospital.

Keywords: *Vascular disease, Aorta, Corticosteroids, Hypertension, Auto immune*

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INTRODUCTION:

Takayasu arteritis is a rare chronic inflammatory arteritis affecting the large vessels in body predominantly the aorta and its main branches. It is a granulomatous of large arteries. 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). Occlusive thromboaortapathy and martorell syndrome (vessels inflammation leads to wall thickening fibrosis, stenosis and thrombus formation). TA appears to have an acute early phase with nonspecific symptoms such as Hypertension, headache, fever, muscle pain, arthralgia, night sweats, and weight loss. Due to non-specific symptoms and the absence of lab parameters the disease is unrecognized. If it is untreated during the next phase the disease affects the aorta and its branches. Vessel wall infection leads to concentric wall thickening, fibrosis and thrombus formation. Affected vessels way become stenotic and at this stage commonly reflect end-organ ischemia such as renal infarction and stroke.

Conventional and digital subtraction angiography has been used for the evaluation for arterial steno occlusive changes or aneurysm in T4. Novel therapeutics approaches such the use of TNF-ALPHA and drug eluting arterial stents for improving the prognosis in severe disease (5). 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 in Mexico to 1:2 in Israel (3). 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 complications (4).

CASE REPORT:

A 24years old woman presented in our hospital with chief complaints of progressive shortness of breath, chest pain since 3 months, low backache since 1 month, pain in abdomen since 1 month, pain over right side of the neck over arterial aspect since 7 months, throbbing type gradually progressive radiating to right side of the face, right and fore arm and right scapular area palpitations only on exertion and with onset of pain lasting for about 10 minutes and relieved with rest. She had a fever since 1 weak ago which is intermittent, loss of appetite since 2 months and blurring of vision. There was no family history of stroke and any inherited disorders and heart related problems. In this general examination the systemic blood pressure was 70/50mm of Hg on 1st day of examination at supine position. On 2nd day the blood pressure found to be 90/60mm of Hg and 3rd day of examination the blood pressure was found to be 110/80mm of Hg.

Laboratory findings were showed that an elevated ESR of 32mm/hr and serum C - reactive protein level of 0.9mm/dl. On ophthalmic examination visual activity was 6/36 in the right eye and counting finger at 2 meters in the left eye. 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. The treatment was given oral steroids on a tapered basis and consulted the department of surgery for a possible surgical intervention.

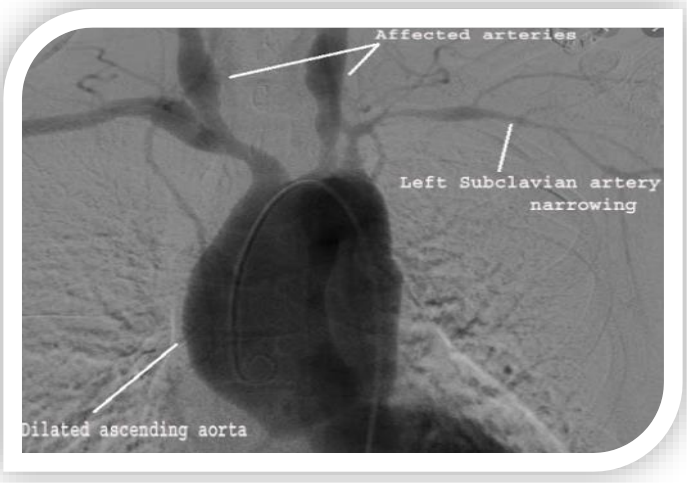

Fig: Dilated aorta

Epidemiology and Etiology:

- Worldwide incidence: 2.6 cases per million per year
- Japanese patients with Takayasu arteritis- Higher incidence of aorta arch involvement
- In contrast, series from India report higher incidences of abdominal involvement
- Most frequencies in Asian countries like- Japan, Korea, and China
- Exact etiology is unknown
- Underlying pathologic process is inflammatory
- Several etiologic factors have been proposed
- Spirochetes, Mycobacterium tuberculosis, Streptococcal organisms
- Genetic factors may play a major role in pathogenesis
- Raised ESR, Leucocytosis, arthralgia and anti-aorta antibodies (7)

DISCUSSION:

TA is one of the common causes of renal vascular hypertension in young Asian females if often related to renal artery stenosis. It is also known as "PULSELESS DISEASE" or "AORTOARTERITIS". 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). Imaging plays a major role in diagnosis of TA. Sonography used for the initial evaluation of patients with suspected one. The four most complications of TA-retinopathy, secondary hypertension, aortic regurgitation and aneurysm.

TA is characterized by granulomatous inflammation of the aorta and its major branches leading to stenosis, thrombosis, and aneurysm formation (6). The lesions of TA are segmental patchy and involving all three layers of vessels. 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 (Table-1). The clinical illustrations can be different based on the location of arterial lesions (2, 3).

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 vasculitis (4). 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 angiography (2). 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) (4). 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, immunosuppressant's such as prednisone and/or methotrexate can lead to significant improvement and consulted the department of surgery for a possible surgical intervention.

CONCLUSION:

TA is a rare disease that is both diagnostically and therapeutically challenging to physicians. Early diagnosis is very important in this disease and improves the outcomes. The role of patient gender and prognosis is concerned, the literature is sparse,

more studies should be conducted. Further, although medical treatment is considered as the mainstay for TA and it is imperative to apprehend both indications and the available options of surgical interventions.

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