TAKAYASU ARTERITIS : A RARE PULSELESS DISEASE

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ABSTRACT
Takayasu arteritis was formerly known as a "pulseless" disease and is a chronic idiopathic vasculitis affecting the large vessels in the body most commonly the aorta. Takayasu arteritis is a rare condition and its acute phase presentation is similar to other conditions making diagnosis difficult.There is a possible relationship that exists between Takayasu arteritis and tuberculosis as both diseases have similar chronic inflammatory lesions and occasionally granulomas on the arterial walls. We present a rare case of a 15 yr old female patient with Takayasu arteritis with tuberculosis .

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Key words: Takayasu arteritis, pulseless disease, tuberculosis

INTRODUCTION
Takayasu's arteritis (TA) is a disease of unknown etiology, characterized histologically by an inflammatory cell infiltrate that affects all the layers of the arterial wall, especially the aorta and its major branches. Its incidence varies between 1.2 and 2.3 cases per million per year, and it is more common in Asians than in other racial groups (1). The etiology of TA is not clear. A number of features suggest an autoimmune base while others raise the question that the aortitis may be an expression of tuberculin sensitization. A causal relationship between TA and tuberculosis (TB) had been suggested. Both diseases show similar pathological changes in the form of chronic inflammatory lesions and, occasionally, granulomas on the arterial walls (2). The genetic relationship between these two diseases has not been reported; however, both diseases have been associated with human leukocyte antigen (HLA) alleles, cold agglutinins and cryoglobulins during the acute phase of the illness (3-6).

CASE PRESENTATION
A fifteen year old young female patient came to a private clinic located in Indira Nagar Lucknow and presented with complains of fever and pain in the abdomen for 15 days, patient had a history of intermittent and low grade fever for the past 6 months. On careful examination by a senior consultant the following observations were made,

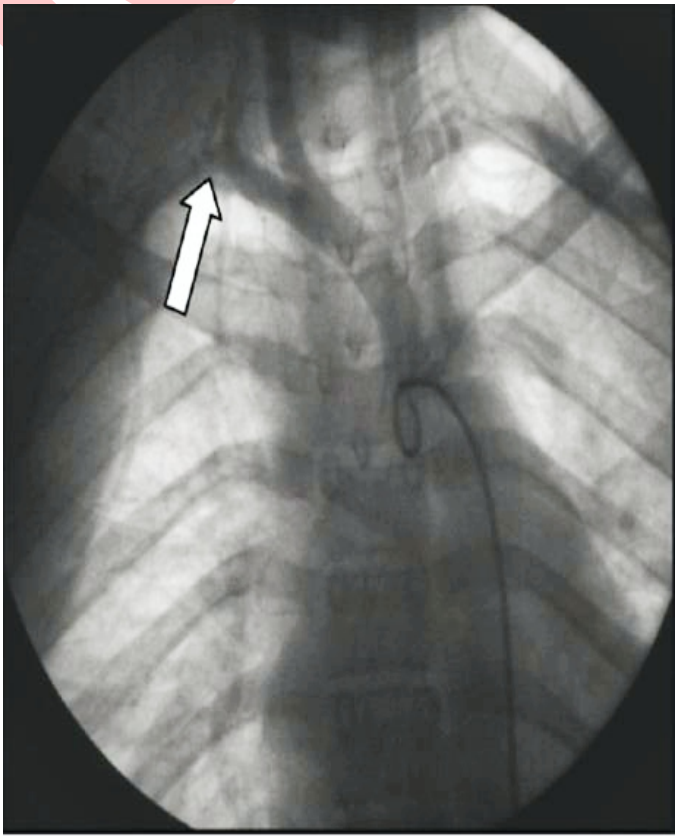

Fig.1: Occlusion of subclavian artery

presence of few enlarged left cervical lymph nodes l ess than 1cm in size, one right supraclavicular and one submandibular enlarged lymph node both nearly 1cm in size. The Brachial and Radial pulses were absent and only lower limb pulse was palpable.

Lab Investigations revealed that the RBC'S were microcytic and hypochromic, a WBC count revealed lymphopenia and neutrophilia. An LDHI2 level of 449.47 was found to be much higher than the normal values ranging between135-225. An X-Ray chest revealed superior mediastinal widening with a prominent right hilum. A USG whole abdomen revealed retroperitoneal lymphadenopathy and mild peritoneal collection, the echotexture of ovaries showed it to be hypoechoic. A CT aortography was done and the observations of the radiologist were as follows, the second and third part of the bilateral subclavian arteries and bilateral axillary arteries were not visualized. Branches of the first part of bilateral subclavian arteries were prominent with a collateral flow to bilateral proximal upper limits through internal mammary and suprascapular branches of thyrocervical arteries (fig.1). A marked narrowing of the superior mesenteric artery was seen from the origin. A large collateral filling of superior mesenteric artery was noted arising from the inferior mesenteric artery. The ascending aorta, arch of aorta, descending thoracic and abdominal aorta were found to be normal. The findings were thus suggestive of Takayasu arteritis. The CT scan of thorax revealed an area of increased lung attenuation, fibro-consolidatory changes were noted in the right apical segment, right middle lobe, bilateral lower zones and along right oblique fissure. Multiple lymph nodes with central necrotic changes and few of them showing matting in pretracheal, paratracheal, subcarinal, bilateral hilar, prevascular, para aortic, aortocaval and mesenteric regions, the largest measuring 2.7x3 cm in size . The lymph nodes were seen to causing compression of the right pulmonary artery. The findings were thus suggestive of tuberculosis.

DISCUSSION

Several workers from India and Japan have suggested hypersensitivity to Mycobacterium tuberculosis as a possible factor in the pathogenesis. Sen et al. reported the presence of tuberculosis in 71% of their patients(7) while Lupi –Hererra found prevalence of tuberculosis in 48% cases (8). All the data regarding the link to tuberculosis have been generated from countries where tuberculosis was also highly prevalent at that time. While India still has a high prevalence of tuberculosis and TA, the Japanese continue to report substantial incidence of TA, though that of tuberculosis has gone down. Thus, there seems to be an association with M. tuberculosis but the evidence regarding its role as the cause of takayasu arteritis is not very strong. The evaluation of disease activity in patients with TA is a challenge. It was initially believed that the disease had three phases: a pre-inflammatory or systemic phase, followed by vascular inflammation, and ending in a “burnt-out” fibrotic, stenotic phase. However, this is too simplistic a sequence. The absence of systemic clinical features does not exclude ongoing vascular inflammation nor does the presence of ischemic symptoms always suggest active inflammation.

The ability to measure disease activity in TA is limited by the absence of any definite laboratory test for this purpose. The presence of constitutional symptoms along with raised levels of acute-phase reactants have usually been used to diagnose the disease. In 50% of patients, the onset of disease is heralded by constitutional symptoms such as fever, anorexia, arthralgia, signs and symptoms of local limb ischemia, hypertension and raised ESR; in the other 50%, there is no history suggestive of an acute phase and these patients present with advanced obstructive lesions (8). Fever is present in 20%–42.5% and arthralgia in 10.9%–55% in various series (8). In our case the patient presented with a debilitating fever and abdominal pain with absence of upper limb pulse (Brachial or Radial).

Noninvasive radiological techniques aimed at detecting arterial involvement are being evaluated. High-resolution B-mode ultrasonography is useful in showing obstructive involvement of the blood vessels, and can also detect intima– medial thickening in the absence of obstructive lesions, which may be a sign of active disease. One study demonstrated circumferential thickening of the common carotid arteries in 19 of 23 TA patients, while angiography revealed stenotic lesions in only 13 of these patients (9). The characteristic finding was circumferential arterial wall thickening of one or both common carotid arteries in the form of macaroni-like, diffusely thickened intima–medial complex. As ultrasonography cannot pick up pulmonary or coronary involvement, magnetic resonance imaging (MRI) and computerized tomography (CT) have also been evaluated as diagnostic techniques. In the early stages of the disease, subtle inflammatory wall thickening may be the only abnormality and MRI may pick up concentric wall thickening of the vessels. T2-weighted images may show bright signals of edema in and around the inflamed vessel. Contrast-enhanced MRI, showing enhanced vessel walls even in the chronic stage, may

suggest activity. Intravascular ultrasound (IVUS) studies of the aorta have shown thickening and altered echogenicity of the media, adventitia and peri-arterial tissues. This was seen even in some portions of the aorta, which looked normal on angiography (10). The technique used in our case was a low dose of AP scan aortography was taken and 100ml of contrast was administered. Scans were acquired in arterial and delayed phases. Reformation was done in sagittal, coronal and volume rendered projection.

CONCLUSION

TA is a systemic disease that might progress to cause vital organ ischemia although the exact cause is not known,it is possible that an infection triggers Takayasu arteritis in people with a genetic predisposition. The detection of the disease in its active phase remains a challenge however techniques such as ultrasound, Computed tomography seem to be promising.

CONFLICT OF INTEREST

The authors declare that they have no competing interests.

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