

CASE REPORT

A Rare Case of Childhood Arteritis

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SUMMARY

Takayasu's arteritis (TA) is a chronic, progressive, autoimmune, idiopathic, large-vessel vasculitis that usually affects young adult females. The disease has been reported to occur in all races and ethnicities. The diffuse nature of this disease can affect multiple-organ systems to varying degrees. Herein, we report the case of a nine year old child who presented with diarrhea, vomiting, loss of weight and weakness. Due to her initial presentation and lab investigations, she was started on antituberculous therapy. During further investigations she was diagnosed to be suffering from Takayasu's arteritis. After one year of ongoing treatment and follow-up, she displayed marked resolution in disease process affecting abdominal aorta and its main terminal branches. We discuss a possible relationship between Takayasu's arteritis and both latent and active tuberculosis which was first pointed out in 1948 by Shimizu and Sano.

Keywords: Takayasu's arteritis, Classification, Complications, Diagnosis, Therapy, Radiography,

INTRODUCTION

Takayasu's arteritis is an autoimmune, large-vessel vasculitis usually affecting young adults. The disease is named for the Japanese ophthalmologist, who in 1908, described a young woman with peculiar retinal arteriovenous anastomoses caused by retinal ischemia from large vessel vasculitis^{1,2}. This disease can affect both sexes and all races but most often involves young East Asian women with median age of 25 yr. The incidence of TA in USA is estimated to be 2.5 in 1000,000 and 1.3 in 1000,000 people in Europe³.

Takayasu's arteritis is a disorder of unknown etiology. On histological examination, the acute vascular lesions of TA show granulomatous infiltrative process with mononuclear macrophages and lymphocytes in the vessel wall. To manage a patient with TA is a great challenge for physician because treatment options vary depending on the stage of the disease process at the time of its diagnosis. Here, we report a rare case of a nine year old female diagnosed with TA, treated with antituberculous therapy and followed up.

CASE REPORT

A nine year old female was referred for ultrasound in February 2012 with complaints of diarrhoea and vomiting for last 2 weeks. She also had on and off fever, loss of weight and weakness for one year. On examination she appeared pale, dehydrated and tachypneic. Her height and weight were below 5th percentile. Laboratory finding revealed raised erythrocyte sedimentation rate (ESR) of 46 mm/h

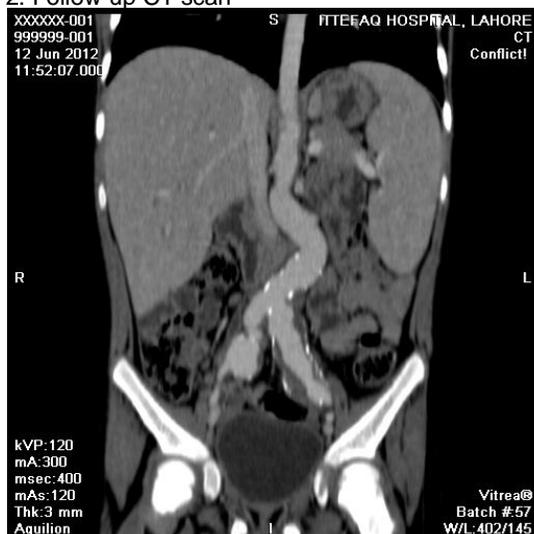
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(normal value <20mm/1 h) and serum C-reactive protein level of 1.03 mg/dl (normal value <0.5 mg/dl). Her serum calcium and potassium were also low. Other laboratory tests were normal. On duplex ultrasound thickening of vessel wall involving middle abdominal aorta and common iliac arteries extending into external & internal iliac vessels was noted. She was advised CT angiography for further evaluation which confirmed ultrasound findings. It showed diffuse intimal thickening and calcinosis of abdominal aorta and iliac branches and fusiform dilatation of left renal artery. The left internal iliac artery was completely occluded. So impression of aortoarteritis involving abdominal aorta & iliac branches highly indicative of paediatric Takayasu arteritis was made with differential of infective arteritis. She was offered antituberculous therapy by her physician and was followed up. Repeat CT scan after 3 months revealed marked resolution in thickening and patency of vessel lumen (Figs 1&2).

Fig. 1: Initial CT scan



Fig. 2: Follow-up CT scan



DISCUSSION

Takayasu's arteritis is an autoimmune chronic granulomatous necrotizing vasculitis mainly affecting large vessels; the aorta and its branches.⁴ The exact pathogenesis of the TA is still unknown, however tuberculosis, streptococcal infections, rheumatoid arthritis and other collagen vascular diseases have been argued as its etiology. Recently an immunological cause has been given more emphasis in its etiology^{5,6}. Manifestations are very polymorphous with presentations ranging from asymptomatic to neurologic catastrophes. According to American Rheumatologic Society three out of the six criteria is necessary to make definite diagnosis of Takayasu's disease⁷: (1) Onset before 40 years, (2) claudication of the extremities, (3) decrease in the brachial pulse in one or both arms, (4) difference of 10 mm Hg or more in blood pressure measured in both arms, (5) audible bruit on auscultation of the aorta or subclavian artery and (6) narrowing at the aorta or its primary branches on arteriogram. The disease is divided into two phases⁸ early/pre-pulseless/systemic phase, late/pulseless phase.

In the early phase of disease patient presents with diverse signs and symptoms such as fever, night sweats, weakness, arthralgia, myalgia, cough, chest and abdominal pain, skin rash, and anemia. Laboratory investigations show persistent elevation of the ESR and a positive C-reactive protein test. Although the diagnosis of early-phase TA is extremely difficult but early diagnosis is important because prognosis depends upon the time at which steroid therapy is initiated⁸. Radiological features of early phase TA demonstrates loss of sharp definition and a wavy or scalloped appearance of descending thoracic aorta on chest X ray. Such subtle findings in

a young female patient should alert the radiologist to the diagnosis of early-phase Takayasu arteritis⁸. CT and MRI angiography play an important role in early diagnosis of TA as the basic radiologic feature of early-phase Takayasu arteritis is aortic wall thickening.

In the late phase of disease, the systemic features subside, while arterial stenosis or occlusion predominate the clinical picture leading to variable ischemic signs and symptoms. Radiological features include an irregular, undulant outline and linear calcifications of the aorta, dilatation of the ascending aorta, cardiomegaly, decreased calibre of pulmonary vessels and rib notching. Calcification, in TA characteristically appears linear and involves the aortic arch and descending thoracic aorta, sparing the ascending aorta. Conversely, calcification of the ascending aorta predominates in syphilitic aortitis⁸. Aortography is the imaging modality of choice for the diagnosis of late-phase TA. The stenosing variety of late-phase Takayasu arteritis is known as atypical coarctation of the aorta. Diffuse ectatic change of the thoracic and upper abdominal aorta associated with tapered narrowing of the distal abdominal aorta produces characteristic "rat-tail" or comma-shaped configuration. There are four types of TA⁸ (i) Type I is classic pulseless disease (involvement of the branches of the aortic arch), (ii) type 2 is mixed type (combination of classic pulseless disease and the atypical coarctation type), (iii) type III is the atypical coarctation type (involvement of the thoracic or abdominal aorta and its branches) and (iv) type IV is the dilated type (extensive dilatation of the length of the aorta and its branches). Type II is the most common variety. Both CT and MRI are highly sensitive and successful modalities in demonstrating stenosis, dilatation, aneurysms, wall thickening and calcification. Faint calcification of the aorta is not visualized with MR but can easily be detected with CT.

Shimizu and Sano were the first to describe a possible relationship between Takayasu's arteritis and both latent and active tuberculosis⁹. They raised this hypothesis because the giant-cell granulomas found in the arterial specimen of Takayasu's arteritis patients were morphologically similar to those found in tuberculous lesions. Anecdotal cases of patients with tuberculosis and concomitant Takayasu's arteritis have been reported to show complete symptomatic remission after successful antituberculous treatment.¹⁰⁻¹² According to epidemiological data there is over representation of TA in patients with past or present tuberculous infection, with prevalence rate of 21.8% to 70%. In a study conducted in India, patients with TA were 46.6 times more likely to have active tuberculosis as

compared to general population.¹³ Data from Mexico showed that this ratio could be exceeded. In a case series study of Takayasu's arteritis from Mexico, 48% of patients had a previous tuberculous infection such as pulmonary involvement, lymphadenopathy and Bazin's erythema induratum; while the prevalence of active tuberculosis was reported to be 0.028% in the general population¹⁴.

Differential diagnosis includes, rheumatic, giant cell arteritis, systemic lupus erythematosus, Buerger's disease relapsing polychondritis, ankylosing spondylitis, rheumatoid arthritis, and Behçet's disease. Infectious arteritis includes; syphilis, tuberculosis and other like inflammatory bowel disease, sarcoidosis, congenital coarctation, Marfan's syndrome and Ehlers-Danlos syndrome. Complications include retinopathy, secondary hypertension, aortic regurgitation and aneurysm formation. Usual treatment is with steroid therapy.

CONCLUSION

Takayasu's arteritis is a disease of young females but can be seen in a child with wide variety of presentations, many with a typical history of other conditions. Modern imaging modalities such as CT, MRI and angiography play a vital role for a definitive diagnosis of TA. Although the use of steroids is cardinal to the acute medical treatment, it is not curative. The possible association between tuberculosis and TA needs to be studied more extensively in future.

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