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CASE REPORT

Takayasu Arteritis: A 4-year-old girl presenting with Heart Failure -Case Report and Literature Review

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Abstract:

Takayasu arteritis (TA) is a chronic idiopathic large vessel vasculitis. TA during childhood remains a diagnostic challenge due to the non-specific symptoms. Here we report a four year- old girl presenting with features of heart failure and later diagnosed as a case of childhood TA along with associated myocarditis. Though TA is rare in children, it must be considered in the differential diagnoses of children who present with heart failure.

Keywords: Takayasu arteritis, vasculitis, heart failure

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Introduction

Takayasu arteritis is a rare chronic idiopathic large vessel vasculitis involving the aorta, its branches and pulmonary arteries. The disease is associated with a high incidence of morbidity and a significant risk of early death [1-6]. It is predominantly a disease of young adults in the second and third decades of life [2]. The onset of illness may be earlier, including childhood, and rarely in infancy [1,3-6]. The clinical manifestations in children are less specific than in adults, and children usually present with fever, headache, hypertension, weight loss, and arthritis⁶. Diagnosis of TA during childhood remains challenging due to the non-specific symptoms, which probably account for higher mortality (33%) in paediatric patient than in adults [7]. Till date, few studies on childhood TA have been published [1, 3-6].

Here, we report a case of takayasu arteritis in a four-year-old girl who presented with features of heart failure. This case highlights that heart failure may be the presenting feature of TA and also it highlights the importance of modern day imaging in the diagnosis.

Case report

A four-year-old female child born out of non-consanguineous marriage presented with complaints of fever, weight loss and easy fatigability for 1 month and shortness of breath for 4 days. Fever was intermittent in nature, while shortness of breath was gradually increasing. There was no significant past or family history. No history of contact with any tuberculosis patient was found.



Figure 1. Chest X-ray showing cardiomegaly with left ventricular apex

On physical examination, her body weight was 13.5 kg (10-25th percentile) and height was 101 cm (10-25th percentile). Clinical examination revealed impalpable peripheral pulses in both the lower limbs. Blood pressure was 144/98 mmHg and 136/90 mmHg in the left and right upper extremities, respectively, and not recordable in both the lower limbs. There was tender hepatomegaly, cardiomegaly and raised JVP. The neurologic and respiratory system examinations were unremarkable, except for tachypnoea.

Blood counts showed mild anaemia, leukocytosis and normal platelet counts. ESR (132 in 1st hr) and C-reactive protein level (101 mg/dl) were raised. Antinuclear Antibody (ANA), antistreptolysin O (ASO) titre and Rheumatoid factor were negative. Serum electrolytes, liver and kidney function tests, thyroid function tests, and urinalysis were all within normal limits. Viral markers were negative for hepatitis B and C virus and HIV. Investigations for tuberculosis revealed negative results. There was no evidence of retinopathy or iridocyclitis on ophthalmological examination.

Chest X-ray revealed cardiomegaly (Figure 1). Abdominal sonography showed hepatomegaly, but kidney size and echotexture were normal bilaterally.



Figure 2. CT Angiography showing irregular narrowing of mid-thoracic aorta

Doppler sonography was done to exclude renal artery stenosis. Echocardiography revealed left ventricular ejection fraction of 35%, grade 2 mitral regurgitation and biventricular hypokinesia suggestive of myocarditis. CT angiography (CTA) showed irregular narrowing of mid thoracic aorta, involving a segment of 10 cm length, with maximum narrowing of 80% (Figure 2).

The patient's clinical findings and results of noninvasive imaging studies were compatible with the diagnosis of childhood TA according to the final EULAR/PRINTO/PRES (European League Against Rheumatism/Paediatric Rheumatology International Trials Organisation/Paediatric Rheumatology European Society) childhood TA criteria⁵. The diagnosis was further confirmed by angiography and angioplasty was done at the same sitting to dilate the narrowed portion of aorta (Figure 3). Pulses



Figure 3. Angiography showing angioplastic dilatation of the narrowed segment of aorta

returned to lower limb immediately after that and blood pressure became recordable.

Oral corticosteroid (prednisone, 2 mg/kg/day) and acetyl-salicylic acid were instituted. Blood pressure normalized gradually with oral amlodipine and losartan. She became afebrile and acute phase reactants normalized; oral prednisone was tapered gradually. On 9 months follow up, she was in remission without any relapse.

Discussion

Takayasu arteritis is a rare large vessels vasculitis with an incidence of 2.6/1,000,000. The etiology and precise pathogenesis of TA are still unknown. Since large vessel biopsies are most often not possible, diagnosis of TA depends on a history of constitutional symptoms suggesting a systemic illness, clinical findings, typical angiographic morphology, and the

exclusion of other conditions causing aortitis [8]. In 2008, EULAR/PRINTO/PRES proposed final validated classification criteria for childhood TA [5]. Diagnosis of our patient was based on these criteria and the characteristic angiographic findings.

In TA, aortic involvement, aortic aneurysms and branch vessel disease of the subclavian, innominate, renal, common carotid, vertebral, and mesenteric arteries are highly prevalent (in decreasing order of frequency) [8]. Based on angiographic findings Takayasu's arteritis is divided into type I (involving aortic arch and its branches), type II (thoracoabdominal aorta and its branches) and type III (involving lesions of both type I & II). Involvement of pulmonary arteries in addition to any of the above types is grouped as type IV. Thoracoabdominal aortic involvement is commoner (type II/III) in children [9] and our patient also showed involvement of the thoracic aorta (type II).

TA is typically more common in women of child-bearing age. Our patient is relatively younger than the previously reported patients with TA. Early disease manifestations are often nonspecific, like fever, malaise, weight loss, headache, hypertension, myalgias, arthralgias, dizziness etc. ("pre-pulseless" stage), whereas later manifestations include diminished pulses, asymmetric blood pressures, claudication, Raynaud phenomenon, renal failure, splenomegaly, and symptoms of pulmonary or cardiac ischemia or stroke [10].

However, the salient clinical features are markedly lower blood pressure and weaker pulses in the upper extremities with coldness or numbness of the fingers [11]. This is why TA is also called "Pulseless Disease". Visual disturbances like visual defects, retinal hemorrhages and total blindness are also common in TA [12]. Neurological deficits and cerebral aneurysm have been reported in children with TA [13]. It is also the most common cause of renovascular hypertension in India [14]. Bicakcigil, et al. (2009) studied 248 cases of TA [15] and came across the following cardinal features - constitutional symptoms (66%),

diminished pulses (88%), bruits (77%), hypertension (43%), aortic regurgitation (33%), renal artery stenosis (26%), pain in extremities (69%), claudication (48%), cerebrovascular accidents (18%) and pulmonary hypertension (12%).

In the study by Talwar et al, myocardial involvement including myocarditis is one of the common presenting features in TA and may precipitate heart failure in these patients [16], as is seen in this case. Congestive cardiac failure can also be associated with hypertension, aortic regurgitation, and dilated cardiomyopathy [17]. TA very rarely manifests with hepatosplenomegaly in its pre-pulseless stage, but in our patient, though hepatomegaly was there at presentation, it was considered as a clinical feature of cardiac failure due to myocarditis.

Laboratory findings are often non-specific, including raised acute phase reactants (e.g. ESR, CRP etc.) and other markers of chronic inflammation (e.g. anemia of chronic disease, leukocytosis, thrombocytosis and hypergammaglobulinemia) [10]. Autoantibodies are not useful in diagnosing TA except to help exclude other autoimmune diseases.

Angiography has traditionally been the gold standard diagnostic test. Aortography reveals focal, smooth, symmetric narrowing of the aorta and multiple branch-vessel stenosis or occlusion. Stenosis is the most common finding, although arterial dilatation and aneurysms are often found.

Magnetic Resonance angiography (MRA) is preferred for long-term follow-up, as it visualizes vessel wall inflammation and mural thickening (oedema representing active inflammation), and provides information relating to activity of the disease. CTA provides similar information to MRA, albeit with higher radiation. Sonography might help to establish the early stage of TA in a pre-stenotic phase in the extracranial vessels.

As the disease has its prodrome fairly similar with many other commoner diseases, early diagnosis is definitely a challenge. On the other

hand, early identification and early institution of specific treatment dramatically improves the prognosis, with lesser complications and lesser need for more invasive surgical procedures or more toxic medications. So, here comes the utility of modern-day imaging, which of course, should be preceded by a lenient clinical suspicion.

Corticosteroids are still the mainstay of treatment in childhood TA [6]. Remission is achieved in 60% of patients treated with glucocorticoids alone, although relapses may occur with dose reduction [6,10]. Cyclophosphamide and methotrexate can be added, if the patient is unresponsive to glucocorticoids alone [18]. Despite the extent and severity of vascular lesions, patients can benefit from surgical interventions, which include bypass surgery, the use of interposition grafts and percutaneous transluminal angioplasty [2, 6, 19]. The outcome of TA depends on the vessel involvement and severity of hypertension. The mortality rate in children was reported to be as high as 35-40% by five years [1].

Conclusion

TA can present in wide variety of ways, many with a typical history of other conditions. Although rare, childhood TA must be considered in children presenting with non-specific systemic symptoms, hypertension and increased acute phase reactants.

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