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Case Report

An unusual case of axillary lymphadenopathy

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Introduction

Lymphadenopathy is a common presenting complaint in pediatric age group.^{1,2} Common causes for lymphadenopathy include viral illnesses, tuberculosis, malignancies. Rarely, lymphadenopathy may be the presenting complaint in autoimmune illnesses and vasculitis syndromes. We recently had a 12-year-old girl who presented with a swelling in the right axilla, the evaluation of which revealed an unusual diagnosis.

Case report

Twelve-year-old girl, a resident of Assam, presented with a single complaint of swelling in right axilla of one year duration

which had gradually increased in size to reach the size of 3 cm × 3 cm. There was no history of contact (Tuberculosis/animals), breathing difficulty, weight loss, bone pains, fever, rash, loss of appetite or distension of abdomen. She had been vaccinated as per UIP and BCG scar was present over left deltoid. Growth parameters were within normal limits. Examination revealed a well-preserved adolescent girl with single large, non-adherent non-tender lymph node in right axilla with normal overlying skin. She also had absent pulsations in the lower limbs, upper limb hypertension (210/110 mm Hg in right upper limb, 168/96 mm Hg in left upper limb, 124/76 mm Hg in right lower limb and 118/80 in left lower limb) and continuous murmur over back. There was no organomegaly. Investigations revealed anemia (Hb 10.5 g/dL, PCV – 28%) and peripheral smear showed normocytic normochromic picture with normal white cell count and platelets. ESR was elevated (46 mm fall in 1st hour) and C reactive protein was positive. Liver and renal function tests were within normal limits. Chest X-ray was normal and USS abdomen did not reveal any significant finding. Mantoux test showed 25 × 25 mm induration with blistering. The swelling in right axilla was subjected to fine needle aspiration and later excision biopsy both of which showed non-specific lymphadenitis. 2D Echo revealed normal biventricular function, mild concentric left ventricular hypertrophy, and normal heart valves and reduced peak systolic velocity in abdominal aorta. Doppler study of renal vessels was normal. Computerized tomographic angiography revealed classic features of Takayasu vasculitis in the form of diffuse concentric wall thickening of ascending and thoracic aorta causing lumen narrowing with two discrete areas of focal stenosis at D8

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(4.2 mm dia, 15 mm length) & D10 (3.9 mm dia, 10 mm length) levels along with well formed collateral vessels over both scapular and infrascapular regions (Figs. 1 and 2). There was no evidence of aortic dilatation and/or dissection. Other major vessels and their branches were normal in caliber and course. She was managed with antihypertensives, oral steroids and salt restricted diet. She is on regular follow up with serial ESR monitoring and is planned for angioplasty of thoracic aorta and left subclavian artery once her vasculitis comes under control.

Discussion

Takayasu vasculitis/Aortoarteritis is the commonest large vessel vasculitis seen in children and is more commonly seen in females.³ The disease is more common in South–East Asian countries as compared to the rest of the world. According to EULAR classification criteria of 2005,⁴ diagnosis of Takayasu vasculitis requires angiographic abnormalities (conventional, CT or MRI) of the aorta or its main branches (Table 1). Although non-specific symptoms like fever, rash, arthralgia/arthritis, myalgia, often precede specific symptoms, localized lymphadenopathy alone as a presenting feature is rare and finds a mention in standard textbooks and some case reports. Localized lymphadenopathy is a well-defined feature of some medium and small vessel vasculitis (eg Kawasaki disease, Wegner's granulomatosis) and is also included in diagnostic criteria for some (eg unilateral cervical lymphadenopathy in Kawasaki disease) but no specific pattern of lymphadenopathy is described in cases of large vessel vasculitis.^{4–6} The systolic murmur/bruit heard over the back was most likely

Fig. 1 – CT angiograph (Reconstructed view) showing two areas of significant stenosis in thoracic aorta at D8 & D10 levels (solid arrows) and stenosis of left subclavian artery (red arrow).

Fig. 2 – CT angiograph (Axial view) showing area of significant concentric stenosis in thoracic aorta (solid arrow) causing lumen narrowing and multiple collaterals in the right infrascapular region (yellow arrows).

due to presence of significant stenosis of thoracic aorta. Our patient had all the criteria present at the time of diagnosis without having any clinical symptoms of claudication.⁴ Positive mantoux reaction has been described in cases of Takayasu vasculitis as a result of altered immunological response in these cases.^{7,8} Tuberculosis and Takayasu vasculitis are both relatively common in South–East Asian countries as compared to western countries, so it is very important to look for presence of tuberculosis in a case of Takayasu vasculitis. Though our patient had elevated ESR and a strongly positive Mantoux reaction, we did not find a focus of tuberculosis despite detailed investigations for the same. Immunosuppression forms the mainstay of treatment for these cases along with anticoagulation followed by surgical/endovascular management.^{3,9,10} Corticosteroids, methotrexate and cyclophosphamide are generally used for immunosuppression and ESR/C reactive protein used for monitoring the disease activity. The disease has a relapsing, remitting course with evidence of histological inflammation in cases in clinical remission. Once the disease is in remission, endovascular intervention/surgery can be carried out. Limb claudication, heart failure, and valvular insufficiency are the main causes of morbidity in cases with Takayasu vasculitis while myocardial infarction and aneurysmal rupture are the leading causes of death. Overall mortality rates are between 10 and

Table 1 – EULAR classification criteria for Takayasu arteritis.

Mandatory criteria

Angiographic abnormalities (conventional, CT, or MR) of the aorta or its main branches
Plus one of these four criteria

1. Pulse deficit and/or claudication of the extremities
2. Blood pressure discrepancy in four limbs (>10 mm Hg)
3. Bruit over the aorta and/or its major branches
4. Systolic/diastolic hypertension (>95th centile for height)

CT, computed tomography; MR, magnetic resonance.

30% with higher rates in South–East Asian countries. The case highlights the need of detailed examination and evaluation of all cases of lymphadenopathy at unusual sites.

Conflicts of interest

All authors have none to declare.

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