Unusual imaging presentation of infantile atypical Kawasaki disease

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Abstract

Kawasaki disease is a systemic medium vessel vasculitis of unknown etiology affecting children under 5 years of age. There are no specific diagnostic tests, and thus, the diagnosis of the disease is primarily made on the basis of clinical criteria. Unusual presentations of Kawasaki disease have been variably reported from different parts of the world. However, presentation of the disease in the form of peripheral thromboembolism and florid non-coronary aneurysms has rarely been described. This report describes the imaging findings in infantile atypical Kawasaki disease with aneurysms of multiple medium-sized arteries, including coronary arteries, emphasizing the detection of clinically silent aneurysms in the disease.

Key words: Coronary artery aneurysm; infantile atypical Kawasaki disease; Kawasaki disease

Introduction

Kawasaki disease, first described by Tomisaku Kawasaki in Japan in 1967, is an acute systemic vasculitis of unknown etiology involving the small and medium-sized vessels with predilection for coronary artery involvement. The disease affects infants and children under the age of 5 years with no racial predilection and a sex ratio of 1.5:1 in favor of male children. Typically, the disease is a multisystem febrile vasculitis with predominant cardiovascular manifestations. If untreated, it can lead to development of coronary artery aneurysm in nearly 25% of patients and systemic vascular aneurysms in less than 2% cases.

Case Report

A 6-month-old male infant was brought to the emergency department with complaints of fever for 1 month, passage of dark stools for 1 week, and discoloration of upper extremity for 2 days. Parents also volunteered the history of fleeting maculo-papular rash. The child was referred for Doppler imaging (Philips HD 11 XE ultrasound machine, PHILIPS USA) of the upper extremity and sonography of the abdomen on emergent basis. Doppler USG showed partially thrombosed fusiform aneurysm of right subclavian artery, axillary artery, brachial artery, and non-thrombosed aneurysm of the right subclavian artery [Figure 1A and B]. Subsequently, ultrafast low-dose CT angiography (Philips Brilliance 40 Slice CT scanner; PHILIPS USA) was done in emergency settings which confirmed the color Doppler findings and additionally showed aneurysms of bilateral common carotid, extracranial internal and external carotid, and bilateral vertebral arteries [Figure 2]. There were aneurysms involving multiple coronary arteries, with giant fusiform aneurysm of left anterior descending artery measuring 13 mm [Figures 3 and 4]. CT angiography of thoraco-abdominal aorta revealed small saccular aneurysm at the origin of celiac artery measuring 29 × 25 mm, with fusiform dilatation of superior mesenteric artery [Figure 5A and B]. There were non-thrombosed fusiform aneurysms of bilateral common iliac artery and common femoral artery without the involvement of the main aortic trunk [Figure 6]. The caliber of thoraco-abdominal aortic trunk was normal with distal smooth tapering.

Based on the imaging features, a possibility of infantile panvasculitis was considered. Review of the clinical parameters revealed normochromic-normocytic anemia,
leukocytosis, and elevated erythrocyte sedimentation rate (ESR; 43 mm in first hour) and C-reactive protein (34 mg/l). The blood cultures and peripheral smears were negative for microbes and had no malarial parasites [Table 1]. Based on the clinical history of conjunctivitis, fleeting rash, swelling of extremities, and multiple aneurysms of medium to small-sized vessels, a diagnosis of infantile atypical Kawasaki disease was made. Apart from the supportive management, the infant was started on intravenous immunoglobulin, but succumbed to death on the third day of admission.

**Discussion**

Kawasaki disease or mucocutaneous lymph node syndrome is a multisystem idiopathic vasculitis affecting medium-sized vessels. The diagnosis is made based on the amalgam of clinical criteria which occur in a sequential and fleeting progression, thus proving a diagnostic challenge. The defining criteria for the diagnosis of Kawasaki disease include fever for at least 5 days and four of the following five principal features: Conjunctivitis, mucositis, cervical lymphadenopathy, truncal rash, and edema of the extremities.[4] Atypical forms of the disease are not rare and are defined as the disease with three principal features or coronary artery involvement with less than three principle diagnostic criteria.[5]

The higher incidence of the atypical form of the disease in infantile age group and the unusual presentations lead to a delay in diagnosis and management, consequently increasing the complication rates. Coronary artery aneurysms, the
Kumar, et al.: Imaging of infantile atypical Kawasaki disease

most serious complication of the disease, occur in up to 25%, whereas other systemic aneurysms occur in less than 2% cases. Majority of the mortality in Kawasaki disease is attributed to cardiovascular events secondary to these aneurysms.

Aneurysm formation is a major cause of morbidity and mortality in children with Kawasaki disease with increased risk in patients with delayed diagnosis, age less than 6 months or more than 9 years, male gender, prolonged (>14 days) or persistent fever despite treatment, leukocytosis, elevated ESR, low albumin, hyponatremia (<135 meq/l), and hematocrit of <35%.\(^\text{[2]}\)

Thus, the diagnosis of Kawasaki disease is suspected on the basis of clinical features and an exclusion of other possibilities. The possible differential diagnoses in the given case of infantile vasculitis consisted of Takayasu disease, fibromuscular dysplasia, infective vasculitis, and Kawasaki disease.

Takayasu’s arteritis is a panarteritis of young adults with involvement of aorta and its main branches with the possible involvement of coronary or pulmonary vasculature. Dilatation and aneurysms are less frequent features than stenosis and occlusion. Aneurysms and dilatation in aorto-arteritis commonly involve the ascending aorta and aortic arch. Fibromuscular dysplasia is a congenital non-inflammatory angiogenic dysplasia with predominant involvement of the renal vasculature in young females and classically shows string of beads appearance of angiography.\(^\text{[6]}\) The negative results of microbiological tests excluded an infective etiology.

**Conclusion**

To conclude, this report emphasizes the role of a radiologist in emergency settings in suspecting the diagnosis and the role of imaging in diagnosis of clinically silent aneurysms in infantile atypical Kawasaki disease.

**References**

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Cite this article as: Kumar N, Mittal MK, Sinha M, Gupta A, Thukral BB. Unusual imaging presentation of infantile atypical Kawasaki disease. Indian J Radiol Imaging 2016;26:373-6.

Source of Support: Nil, Conflict of Interest: None declared.