A case of 'node first Kawasaki disease'

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Introduction

Kawasaki disease (KD) is a systemic vasculitis predominantly involving medium size arteries[1]. Around 50% of individuals with KD develop lymphadenopathy. If they have only lymphadenopathy associated with fever, it is called 'node first KD' (NFKD), which is a rare presentation[2]. We report a patient with NFKD presenting at a very young age, which is even rare in this age group.

Case report

A two year and six month old boy was admitted with fever for two days and right sided cervical lymphadenopathy for one day duration. Despite administration of broad-spectrum intravenous antibiotics for five days, the high grade fever and lymphadenopathy persisted. He was ill looking and irritable. Cervical lymphadenopathy was prominent on the right side. He had a 2cm tender hepatomegaly. Rest of the system examination was normal. Pedal erythema and oedema developed on day seven of the illness.

White blood cell count (WBC) was 26,800/cu mm with 80% neutrophils. Platelet count rose from 310,000/cu mm to 522,000/ cu mm on day 10. Peripheral blood smear showed band forms and toxic granules in neutrophils. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were 118mm/1st hour and 151mg/dL respectively. Serum albumin was 2.8g/dL. Urine full report did not reveal sterile pyuria. There was biochemical evidence of cholestatic hepatitis and ultrasonic evidence of acute hepatitis. Ultrasound scan of the neck showed bilateral cervical lymphadenopathy without evidence of suppuration.

According to above clinical and investigation findings KD was diagnosed. Child was given intravenous immunoglobulin (IVIG) 2g/kg on day seven of the illness and was started on an anti-inflammatory dose of aspirin (80mg/kg/day). With the commencement of treatment, fever settled within 12 hours. There was a dramatic improvement in the general condition of the child. Aspirin dose was reduced to anti thrombotic dose on day 10 of the illness, after 72 hours of fever free period. There was no evidence of coronary involvement in initial and subsequent 2D echocardiograms. Even during the course of the illness rest of the principal clinical criteria were not noted. Child was discharged on day 10 of the illness without permanent sequelae of the disease.

Discussion

Incidence of KD is highest under five years of age. KD can resolve without treatment and go undiagnosed, as generally it is a self-limiting disorder[3]. In all patients with KD, initial manifestations are acute onset high fever, poor general wellbeing and irritability[2]. Principal clinical criteria develop later. Lymphadenopathy (>1.5 cm) is the least common clinical feature[2]. It is non-suppurative and usually unilateral. However, bilateral lymph node involvement is also not rare[3]. Approximately 80-90% of patients develop the other four criteria; generalized pleomorphic rash, palmoplantar erythema, non-purulent conjunctivitis and mucosal enanthema[2]. Our patient did not fulfill criteria to diagnose typical KD. But the patient had the criteria to diagnose incomplete KD. High fever for more than five days, lymphadenopathy and pedal oedema associated with high ESR, CRP and four abnormal laboratory findings (platelet count ≥450,000/cu mm after day seven, elevated alanine transferase (ALT), serum albumin ≤3g/dL and WBC count ≥15,000/cu mm) were the criteria to diagnose incomplete KD[2,4]. Our patient presented only with fever and cervical lymphadenopathy, suggestive of NFKD. It is a rare presentation and even rarer in the younger age group[5].

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NFKD is usually misdiagnosed as bacterial cervical lymphadenitis (BCL) and delay in establishing the diagnosis may lead to permanent coronary complications. Comparison between typical KD with and without lymphadenopathy, NFKD and BCL have been done in several studies[5,6]. Majority of individuals with NFKD were older than typical KD[5]. When compared with BCL, NFKD had lower WBC, platelet count and haemoglobin levels. However, NFKD showed higher absolute band count, ESR, CRP and ALT suggestive of hepatobiliary inflammation[6]. Yanagi S, *et al* have suggested a scoring system which has four features to discriminate NFKD from BCL to prevent unnecessary delay in diagnosing NFKD[7]. These are age >5 years, absolute neutrophil count $>10\times10^9$ cells/L, aspartate aminotransferase >30 IU/L and CRP >7mg/dL[7]. Except the typical age, our patient fulfilled the other three criteria in the scoring system. Radiological features also help in differentiating KD from BCL. BCL is characterized by a single dominant lymph node with appreciable features of suppuration. Multiple lymph nodes with limited inflammation are characteristic features of KD[6].

There are a few case reports in the literature where NFKD was complicated with coronary aneurysms including giant coronary aneurysms[8,9]. As the clinical picture of NFKD is almost similar to BCL, delay in establishing the diagnosis and delay in commencing the definitive treatment are commonly seen.

NFKD and typical KD with lymphadenopathy are believed to have higher degree of inflammation compared to KD without lymphadenopathy and NFKD carries higher risk of developing coronary aneurysms and immunoglobulin resistance[5,6]. We believe that our patient benefited from early commencement of IVIG. According to predicting risk of IVIG resistance in KD our patient scored three. A score of ≤3 is considered as low risk to develop resistance to IVIG[3]. Being a male and serum albumin <3.5g/dL were the only poor prognostic factors we identified in our child.

NFKD being treated as BCL is a common scenario. NFKD needs to be excluded, when there is poor response to intravenous antibiotics without suppuration in cervical lymphadenopathy. Delayed diagnosis and delayed commencement of IVIG are major risk factors for coronary complications. High degree of clinical suspicion is required to prevent an unfavourable outcome.

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