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Involvement of BCG Scar in Kawasaki Disease

Kawasaki disease (KD) is an acute, generalized, self-limiting vasculitis of small and medium sized arteries of unknown etiology mostly affecting children less than 8 years age. As there is no diagnostic test for KD, the diagnosis is based on clinical criteria and exclusion of other diseases. An important clinical sign although not included in the diagnostic criteria, which helps in the diagnosis is involvement of the BCG scar erythema and induration(2).

We report two cases of KD with involvement of BCG scar, which presented to our hospital.

Case 1: An 11-month-old female infant, immunized for age was referred to our department with history of fever, cough and skin rash of 10 days duration. On examination, child was irritable and febrile. Physical examination showed exfoliation in both axilla and over the back, fissuring of the lips, erythematous oral cavity, strawberry tongue and conjunctival injection. There was remarkable inflammation and induration involving the BCG scar. Systemic examination was normal.

Baseline investigations showed Hb 8.4 g/ dL, TLC 13,800/cumm, DC-P72%, L-28%, ESR 70 mm/hr, Platelets 320,000/cumm, CRP-24 mg/L, Urine Micro 1-2 WBC/HPF. CSF study normal, S. Widal and ASO titer were negative; cultures (Blood, Urine, CSF) RFT, LFT, S. electrolytes were all normal. RA factor, ANA and dsDNA were negative. As conical picture was suggestive of KD, a 2-D ECHO study was done and reported as : abnormally dilated LMCA & LCD, consistent with KD.

Case 2: A 1-year -old fully immunized female child presented with history of fever and cough of one-week duration. At the onset of the illness, there was inflammation of the BCG scar as observed by the mother. On examination, child was irritable, had bilateral conjunctival injection, erythematous lips and oral mucosa. Systemic examination was normal.

Investigations were as follows: Hb 9.8 g/ dL, TLC 14,200/cumm, DC-P64%, L-46%, ESR 115 mm/hr, Platelets 345,000/cumm, CRP 30 mg/L. Other investigations as done for the previous case were inconclusive. 2-D ECHO showed coronary artery dilatation/ ectasia.

Both the children were started on IV Immunoglobulin (2 gm/kg over 12 hours), Tab. Aspirin (70 mg/kg/day) in divided doses and supportive care. Fever subsided dramatically within 24 hrs of Initiation of treatment. Subsequent blood investigations done during the third week of illness showed thrombocytosis and a fall in ESR. The convalescent period was unremarkable.

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Local inflammatory reactivation of a previous BCG inoculation site is a specific and early manifestation of KD (3). This phenomenon was hypothetically ascribed to cross-reactivity between mycobacterial Heat Shock Protein (HSP) 65 and its human homologue HSP 63(4). Subsequently, studies have revealed that T-cells obtained from the peripheral blood of KD patients, recognize an epitope from HSP 65 and cross-reacted with the corresponding peptide sequence of human HSP 63(3,4). Immunofluorescence studies on biopsy specimen from skin lesions and reactivated BCG Inoculation site in KD patients have also shown striking similarities(6). These findings may suggest that cross-reactivity between specific epitopes of mycobacterial and human HSP could play a role in the development of the tissue damage characteristic of KD.

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Cystic Nephroma

A 13-month-old male child presented with asymptomatic left sided abdominal mass incidentally discovered by his mother. On examination, a 10×10 cm retroperiotoneal mass was felt in the left upper abdomen. Abdominal ultrasonography revealed a multiloculated cystic retroperitoneal mass suggestive of hydatid cyst of the left kidney. CT scan revealed a well-defined circumscribed exophytic mass arising from outer border of left kidney (*Fig. 1*). The renal parenchyma was only minimally compressed; the pelvicalyceal system and function were normal.

Patient was managed by nephrectomy. Histopathological examination revealed multilocular lesion with definite epithelial lining of loculi. There was no solid tissue identified within the cystic tumor. Neither mature nephrons nor malignant blastemal cells were present within the septae of the cystic lesion. Loculi were neither communicating with each other, nor with renal pelvis.

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