SPOT DIAGNOSIS (IMAGE GALLERY)



SEBORRHEIC DERMATITIS WITH OSTEOLYTIC LESIONS ON X-RAY SKULL

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A $2\frac{1}{2}$ year old child presented with complaints of failure to thrive, delayed milestones and skin lesions over scalp since $1\frac{1}{2}$ years. On examination, baby was severely malnourished with posterior cervical lymphadenopathy, marked hepatosplenomegaly and seborrhoeic dermatitis {fig 1,2}. The blood investigations showed hemoglobin of 8.7 g percent, total counts of 9100 cells/ cumm with neutrophils of 81 percent and lymphocytes of 19 percent, platelets 3.3 lakh/ cumm and ESR of 80 mm. Patient's HIV status was negative. X ray of the skull revealed multiple punched out osteolytic lesions {fig 3}.

What is the diagnosis?

Langerhans cell Histiocytosis {LCH}. In the child, skin biopsy findings were confirmatory of langerhans histiocytosis with langerhans cell with an excentric, "coffee-bean" nucleus in the background of granulomatous cells. LCH is a proliferative disorder of histiocytes characterized by infiltration of one or more organs by large mononuclear cells with bland appearing nuclei with a central groove. The incidence has been estimated to be 2-5 per million per year {1}. LCH can occur at all ages but the peak incidence is at 1-4 years of age with predilection for males {2}. Approximately 80 percent of patients have skeleton involvement, 60 percent will have lymphadenopathy, skin involvement and hepatosplenomegaly {2,3}. Accumulation of langerhans cell along with macrophages, lymphocytes and eosinophils together form granulomatous lesion. Cutaneous manifestations are common in LCH and may represent the earliest sign of the disease. The typical lesion is small yellow papule with scaling {3,4}. The skin lesions usually occur as scaly, erythamatous, papular, that are seen on the scalp, axilla, postauricular areas, hand and feet. The papules may coalesce to form an erythematous, weeping or crusted eruption, mimicking seborrehic dermatitis as in our case. If there is widespread involvement of the skin, eczema and hemorrhagic rash can be present. In some cases rash can be erosive and a risk factor for superinfection. Cutaneous lesions appear more or less in successive crops. Cutaneous lesions may be the presenting feature {4,5}. In 80 percent of patients lytic bony lesions will be present $\{1\}$. Bony lesions are more often single rather than multiple. They may be asymptomatic or associated with pain and swelling. Calvarium is the most commonly affected bone. Radiologically the lesions appear as "punched out" or sharply demarcated as shown in the skull x-ray of our child.

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