

Histiocytosis X Presenting as Chronic Discharging Ears

Malignant histiocytosis was first described in 1939 by Scott and Smith as histiocytic medullary reticulosis(1). We report a case of Histiocytosis X in a 2-year-old by presenting with chronic discharging ears, which is a rare manifestation of the disease.

A 2-year-old boy was admitted with complaints of chronic ear discharge and fever of 1 month duration. He had recurrent vague pain abdomen and lethargy for almost same period. There was loss of appetite and significant weight loss. There was history of mild abdominal distension during this period. On direct interrogation, there was history of polyuria and polydipsia. There was no history of cough, respiratory distress, rash, bleeding manifestation, or convulsion.

On examination, he was malnourished, irritable and pale. Blood pressure and upper segment/lower segment ratios were normal. There was no lymphnode enlargement or rash. There was serous discharge from both ears and prominent abdomen with dilated veins. Liver was enlarged 3 cm below costal margin with a smooth surface and sharp margin and spleen palpable 8 cm, firm in consistency. There was no ascites. There were 5-6 circular punched out lesions over skull which measured 1-1.5 cm in diameter. On the 4th day of admission, petechial rashes appeared over the scalp and gluteal region. Examination of other systems were normal.

Investigations showed: hemoglobin-7.9 g/dl, TLC-7900/cu mm, N-70%, E-2%, ESR 57 mm in first hour, peripheral smear—hypochromic microcytic anemia;

and platelet count 1.5 lakhs / cu mm. Urine routine examination was normal with specific gravity of 1.000 and specific gravity after dehydration was the same. Osmolality could not be done. Other investigations were blood urea 22 mg/dl. Total serum bilirubin 0.9 mg/dl, SGOT-50 IU, SGPT-37 IU, alkaline phosphates-416 units/L, LDH-599 U/L, reticulocyte count 1.2%, mantoux test-negative. On X-ray skull, multiple osteolytic lesions were seen and on X-ray pelvis one osteolytic lesion was visualized on both sides. X-ray mastoids, chest, bone marrow aspiration, liver biopsy and ultrasound abdomen were normal. Biopsy from skull lesions showed bony fragments interspaced by histiocytes lymphocytes, plasma cells, eosinophils and polymorphs. Many multinucleated giant cells were seen. The cytologic diagnosis was histiocytosis X. The child was treated with corticosteroids and vincristine and was discharged at request on day three of admission.

This case of histiocytosis X had all the classical features except lymphadenopathy was not present. The presentation with chronic discharging ears is a rare manifestation which prompted this report.

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REFERENCE

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