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Clinical Image

A case of langerhans cell histiocytosis with cholangitis

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A one-year-old boy presented with a maculopapular, pruritic rash over his entire body for 1 month, fever for 3 weeks, distension of abdomen for 2 weeks, yellowish discoloration of skin and eyes associated with swelling of face and legs for 1 week, and passing dark colored urine since 1 day. On examination, the child had icterus, sparse scalp hair with seborrheic dermatitis [Figure 1a], and a maculopapular rash over the entire body mainly involving the trunk including face, palms, and sole [Figure 1b] with bilateral pedal edema and moderate hepatomegaly [Figure 1c]. He had bicytopenia (Hemoglobin – 7.6 g/dL, Platelet count – 49,000/mm³) with raised C-reactive protein (79 mg/dL), deranged liver function tests (Total Bilirubin – 12.8 mg%, Direct Bilirubin – 11.2 mg% Serum Glutamic Oxaloacetic Transaminase – 134 IU/L, Serum Glutamic Pyruvic Transaminase – 53 IU/L, Alkaline Phosphatase – 377 IU /L, Gamma-Glutamyl Transpeptidase – 378 IU/L), hypertriglyceridemia (597 mg%), and hyperferritinemia (3090 mg%). Ultrasound of the abdomen showed hepatomegaly with altered echotexture, prominent intra hepatic biliary radicles with echogenic content suggestive of cholangitis. Differentials of Langerhans cell histiocytosis (LCH) and hemophagocytic lymphohistiocytosis were considered and a bone marrow biopsy was done which showed diffuse sheets of histiocytes within marrow spaces [Figure 2a] with cells expressing CD 1a, [Figure 2b] S100 protein [Figure 2c] and strong diffuse expression of Langerin [Figure 2d] in 20–30% cells, confirming the diagnosis of LCH.^[1]



Figure 1: (a) Sparse scalp hair with seborrheic dermatitis, (b) Maculopapular rash over the palm, and (c) Maculopapular rash over the abdomen.

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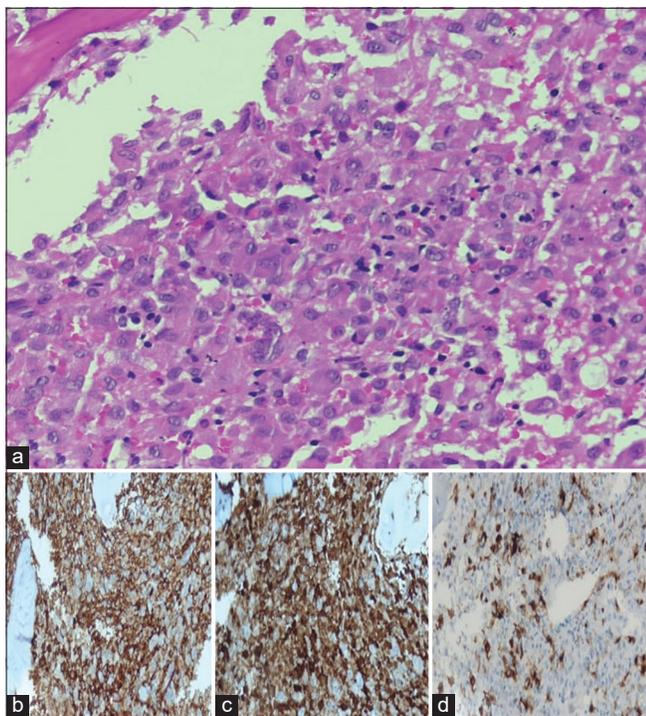


Figure 2: (a) Bone Marrow Biopsy showing diffuse sheets of histiocytes involving marrow spaces, (b) Immunohistochemistry staining positive for markers CD1a, (c) S100, and (d) Langerin.

Cholestasis and sclerosing cholangitis is seen in cases of LCH in children and has a poor prognosis.^[2,3]

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Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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