Disseminated Langerhans histiocytosis presenting as cholestatic jaundice

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Jaundice in Langerhan cell histiocytosis is a manifestation of liver disease; which may be attributed to hepatocellular dysfunction or hemolysis. Cholestasis may result from lymph nodes obstructing the porta hepatis. This report describes a case of type II histiocytosis X with obstructive cholestasis and pulmonary involvement in the form of cysts without significant lymphadenopathy at the porta. A 2 year old boy with progressive abdominal distention, right ear discharge and seborrhoic dermatitis at since 6 months of age was studied. He also had a one month history of fever, jaundice and skin lesions on his scalp for an uncertain duration. He had firm hepatosplenomegaly and shifting dullness. Laboratory findings revealed mild anemia. Liver chemistries revealed AST of 119 U/l, ALT 50 U/l, ALP of 4045U/L and GGT of 924IU/l and a total serum bilirubin of 10.0 mg/dl with direct fraction of 8.9 mg%. Bone marrow examination revealed mild plasmacytosis, dyserythropoiesis consistent with hepatic disease, but no evidence of metastasis. Contrast enhanced tomography of the chest revealed multiple centrilobular nodules in bilateral lung field diffusely exhibiting ground glass opacity, with cystic changes in the upper lobe. Skeletal survey showed multiple osteolytic punched out lesions in the parietal areas of the skull bilaterally and right iliac bones. The skin biopsy of the scalp was showed CD1a positive and S100 positive histiocytic cells. He completed chemotherapy as per protocol and showed progressive improvement throughout his course. Although there is multisystem involvement in LCH, health practitioners must be cognizant that LCH can present as cholestatic jaundice without significant porta hepatis lymphadenopathy.

Biography
Rohit Kapoor has completed his MD (Postgraduation) in Pediatrics at the age of 26 years from Vardhman Mahavir Medical College and Safdarjung Hospital, New Delhi, India. He is currently employed as a Medical Officer under the Government of India. He has completed his thesis during Post-graduation on "Retinopathy of prematurity in infants with birth weight between 1500 grams to 1800 grams". He has cleared the USMLE examinations and wishes to specialize in combined Internal Medicine and Pediatrics residency or in Pediatrics separately.

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