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Case Report

Langerhans Cell Histiocytosis with Sclerosing Cholangitis: A Radiological Perspective

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ABSTRACT

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by proliferation of CD207+ and CD1a+ dendritic cells. Liver involvement occurs in approximately 10-18% of pediatric LCH cases, with sclerosing cholangitis being an uncommon but serious complication found in only 1.3-6% of children with LCH.

We present a case of a 2-year-5-month-old male child who initially presented with persistent cough for 15 days and failure to thrive since 1½ years of age. The child had a history of recurrent respiratory infections, progressive abdominal distension and constipation. Radiological investigations revealed hepatomegaly with characteristic features of sclerosing cholangitis. Further evaluation confirmed the diagnosis of Langerhans cell histiocytosis with liver involvement.

Keywords: Langerhans cell histiocytosis; Sclerosing cholangitis; Pediatric hepatobiliary imaging; Liver involvement; Failure to thrive; MRCP, Ultrasound; CT Imaging, Pediatric radiology

Introduction

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by the abnormal proliferation and accumulation of dendritic cells resembling epidermal Langerhans cells[2]. The incidence in children is estimated at 2-9 cases per million per year, with the majority of cases occurring before five years of age. The clinical presentation of LCH is highly variable, ranging from isolated bone lesions or skin rashes to multisystem involvement with significant morbidity and mortality[3].

Liver involvement occurs in approximately 10-18% of pediatric patients with LCH and represents a high-risk feature of the disease. Sclerosing cholangitis, characterized by inflammation, fibrosis and multiple strictures of the bile ducts, is a particularly severe form of hepatic LCH found in only 1.3-6% of affected children. This complication is associated with poor outcomes, with a reported 5-year survival rate of only 25%[3].

The diagnosis of hepatic LCH with sclerosing cholangitis can be challenging due to its nonspecific clinical presentation and the requirement for specialized imaging and histopathological evaluation[4]. Radiological assessment plays a crucial role in identifying liver involvement, evaluating the extent of biliary disease and monitoring response to treatment.

We present a rare case of a toddler with LCH and sclerosing cholangitis, focusing on the radiological findings that contributed to diagnosis and management.

Cases Presentation

A 2-year-5-month-old male child was brought to the paediatric outpatient department by his father with complaints of persistent cough for 15 days and poor weight gain since 1½ years of age. The child was reportedly well until 1½ years of age when the mother first expressed concern about inadequate weight gain.

Over the past year, the child had experienced multiple episodes of fever, cough and cold, which were managed by a local practitioner with symptomatic treatment. Notably, the child developed progressive abdominal distension 2-3 months prior to presentation. There was documented weight loss of 2 kg (from 9 to 7 kg) over the last four months. The father reported intermittent disturbed sleep but denied any history of frequent vomiting, jaundice, hematemesis or swelling. There were no bony deformities, lethargy, pallor, cyanosis or visual defects noted.

The patient had no significant family history, no known tuberculosis contacts and no known drug allergies. Birth history revealed a full-term normal delivery with no complications or NICU stay.

Physical examination revealed a pale, undernourished child with significant hepatomegaly. Hyperpigmented papules with scaling over the palm were also seen. Based on the clinical presentation, a comprehensive radiological evaluation was initiated **(Figure 1)**.

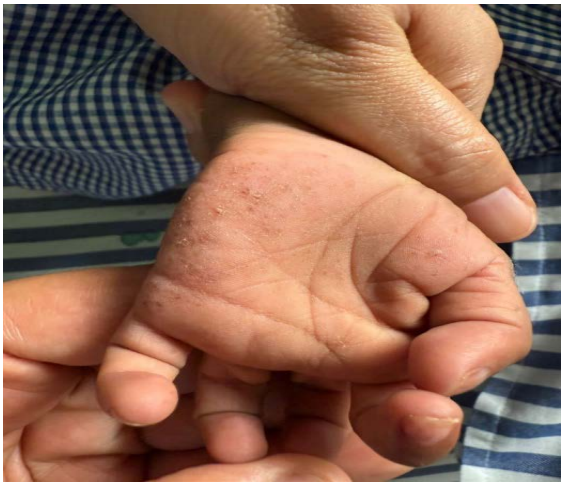

Figure 1: Hyperpigmented papules with scaling over the palm.

A chest radiograph performed showed reticulonodular opacities scattered over both lung fields suggesting the possibility of an infective aetiology **(Figure 2)**.

Ultrasound of the abdomen demonstrated hepatomegaly with increased periportal echogenicity and a heterogeneous parenchymal pattern. A few focal hypoechoic lesions were seen scattered in the liver parenchyma **(Figure 3)**.

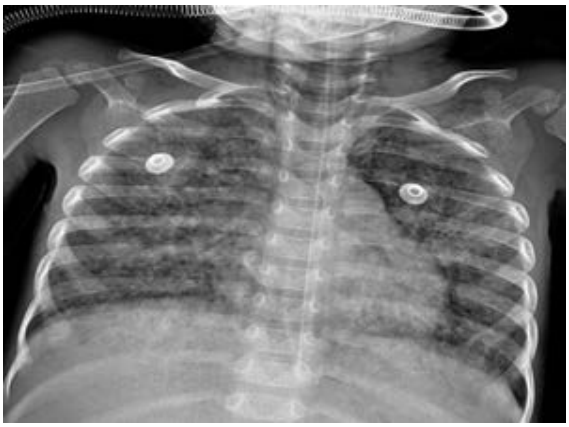

Figure 2: Ultrasound of the abdomen demonstrated hepatomegaly with increased periportal echogenicity.

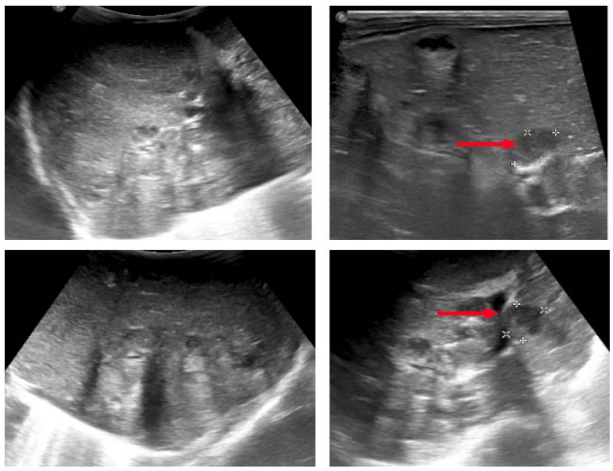

Figure 3: USG images of the liver showing dilated and irregular intra-hepatic biliary radicals.

Few hypoechoic areas are also seen in the liver parenchyma.

Contrast-enhanced computed tomography (CECT) of the abdomen showed hepatomegaly with periportal hypodensity and dilatation of the intrahepatic biliary system **(Figures 4 and 5)**. The extrahepatic biliary system appeared normal. Few focal hypoechoic areas were seen in the liver parenchyma around the peripheral bile ducts suggestive of cholangitic abscesses.

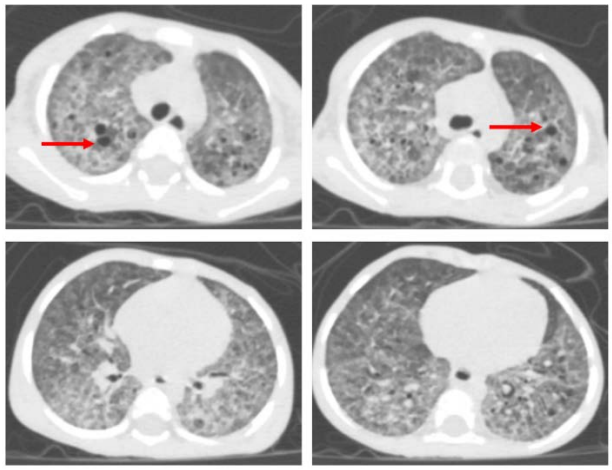

Figure 4: Diffuse interlobular septal thickening is noted in all lobes of both lungs with subpleural sparing. Multiple sub centimeter thin walled intraparenchymal cysts are seen scattered over both lungs (arrow). Diffuse ground glass opacities are also noted over both lungs.

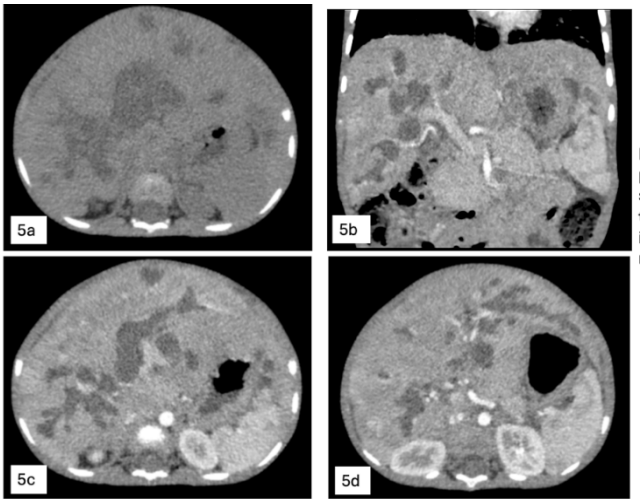

Figure 5: (a) axial plain, (b)Coronal portal, (c) & (d) axial portal images showing moderate dilatation of the central and peripheral intrahepatic biliary radicals.

Given the biliary abnormalities detected on initial imaging, magnetic resonance cholangiopancreatography (MRCP) was performed, which revealed irregular narrowing and beading of the intrahepatic bile ducts with areas of stricture alternating with slight dilatation **(Figure 6)**, classic features of sclerosing cholangitis. The common bile duct and extrahepatic ducts were relatively spared.

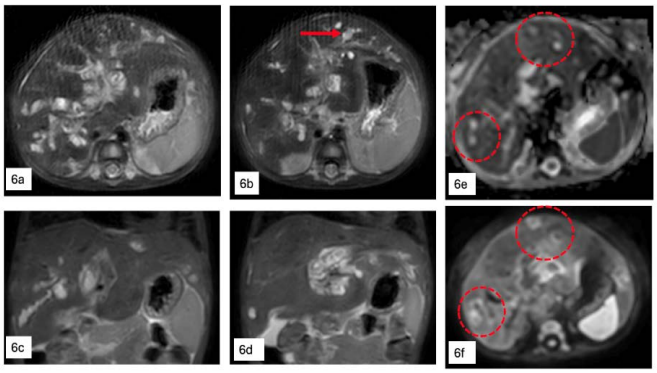

Figure 6: (a) and (b) axial T2WI; (c) and (d) coronal T2WI showing irregular appearance of the intra- hepatic biliary radicals with ill-defined focal lesions connected to the radicals (arrow) - these show a peripheral high signal on DWI (e) with mild signal loss on ADC maps (f).

Mural uptake was noted along the dilated biliary radicals. No skeletal lesion was seen on PET/ CT **(Figure 7)**.

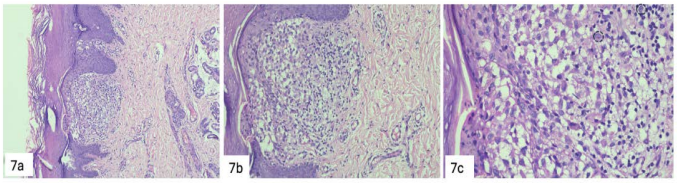

Figure 7: H & E images at (a) 4x, showing normal epidermis with collection, (b)10x: Normal skin with lesion between two rete ridges, (c) 20x: histiocytes and lymphocytes hugging the epidermis with cleaved lymphocytes.

Liver function tests showed a cholestatic pattern with elevated alkaline phosphatase and gamma-glutamyl transferase levels **(Figure 8)**.

Skin biopsy was done which was positive for CD1a-expressing cells confirming the diagnosis of LCH.

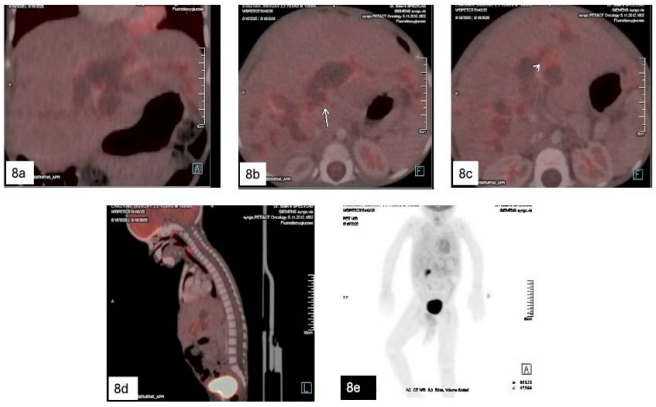

Figure 8: (a), (b) and (c) FDG PET images showing mural uptake along the dilated biliary radicals, (d) And (e) No FDG avid lytic skeletal deposits seen.

Discussion

Hepatic involvement in LCH represents a significant clinical challenge, with sclerosing cholangitis being one of the most severe manifestations[3]. Our case illustrates the typical radiological findings in a child with LCH-associated sclerosing cholangitis, highlighting the importance of multimodality imaging in establishing the diagnosis.

Ultrasound is typically the first imaging modality employed in the evaluation of pediatric hepatobiliary disorders. In hepatic LCH, ultrasound findings may include hepatomegaly, periportal hypo echogenicity and in some cases, multiple hypoechoic nodules within the liver parenchyma[2]. As seen in our patient, altered periportal echogenicity represent inflammatory and fibrotic changes in the portal tracts, which are characteristic of LCH-related biliary disease.

Computed tomography provides additional information regarding the liver parenchyma and can detect subtle abnormalities not apparent on ultrasound. Typical CT findings in hepatic LCH include periportal hypodensity, representing edema and infiltration of Langerhans cells around the portal tracts[2]. In advanced disease, nodular lesions may be observed, which can appear hypodense on non-contrast images and show variable enhancement patterns after contrast administration[5].

Magnetic resonance imaging with MRCP represents the gold standard for non-invasive assessment of the biliary system. In LCH-associated sclerosing cholangitis, MRCP demonstrates irregular narrowing and beading of the intrahepatic bile ducts, with areas of stricture alternating with slight dilatation[6]. These findings closely resemble those seen in primary sclerosing cholangitis but must be interpreted in the context of the patient's age and clinical presentation. The extrahepatic biliary system may be relatively spared in early disease, as observed in our patient.

The radiological differential diagnosis for a child presenting with hepatomegaly and biliary abnormalities includes primary sclerosing cholangitis, autoimmune hepatitis with sclerosing cholangitis, congenital biliary disorders, post-infectious cholangiopathy and other infiltrative diseases. LCH should be considered in the differential diagnosis, particularly when there are accompanying systemic symptoms or characteristic lesions elsewhere in the body[5].

The diagnosis of hepatic LCH with sclerosing cholangitis typically requires tissue confirmation through liver biopsy.

However, as noted in the literature, Langerhans cells may selectively involve the major bile ducts, which are not typically sampled in percutaneous liver biopsies. Therefore, a negative liver biopsy does not exclude LCH-associated biliary disease. In our case, a skin biopsy was done which was positive for CD1a-expressing cells, confirming the diagnosis.

The management of LCH with hepatic involvement typically involves systemic chemotherapy according to established protocols. The prognosis for children with LCH-associated sclerosing cholangitis is generally poor, with progression to biliary cirrhosis and liver failure in many cases despite treatment. Liver transplantation may be considered in cases of end-stage liver disease, though the risk of recurrence in the transplanted liver remains a concern[3].

Radiological follow-up plays a crucial role in monitoring disease progression and response to treatment. Serial ultrasound examinations can assess changes in liver size and parenchymal texture, while MRCP is valuable for monitoring the evolution of biliary disease. In cases of disease progression or treatment failure, alternative therapeutic strategies may need to be considered.

Conclusion

LCH with sclerosing cholangitis represents a rare but serious manifestation of the disease in children, with significant morbidity and mortality[3].

Radiologists should maintain a high index of suspicion when evaluating children with diffuse lung lesions with associated hepatomegaly and biliary abnormalities, particularly in the context of systemic symptoms or failure to thrive. It is important to clinically examine the child and look for skin lesions which would further help in reaching a diagnosis as it is relatively easy to perform a skin biopsy.

This case highlights the importance of multimodality imaging in the diagnosis and management of Langerhans cell histiocytosis with sclerosing cholangitis in pediatric patients as this can provide valuable diagnostic information and guide clinical decision-making.

A multidisciplinary approach involving radiologists, pediatricians, pathologists and oncologists is essential for optimal management of these complex cases.

Early recognition of the characteristic imaging features may facilitate prompt diagnosis and treatment, potentially improving outcomes in affected children.

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