Clinical Brief

Imaging Findings in Hepatic Langerhans' Cell Histiocytosis

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ABSTRACT

We describe ultrasonographic and computed tomographic features of hepatic lesions in two cases of disseminated Langerhans' cell histiocytosis affecting children. In the first case, hyperechoic band like periportal lesions were observed at ultrasonography, which on computed tomography was found to be hypodense admixed with fatty attenuation (HU≅23 to - 57) at places. In addition, the caudate lobe was very prominent. In the second case, the hepatic parenchyma showed predominantly hyperechoic diffusely heterogeneous echogenicity. There were features of cirrhosis of liver with portal hypertension in the form of atrophy of right lobe with hypertrophy of left lobe of liver with lobulated outline, prominent main portal vein and splenoportal axis, splenomegaly and gastroesophageal varices. Both the patients were put on chemotherapy as per schedule (Protocol : DAL HX – 83) and are on follow up. **[Indian J Pediatr 2006; 73 (11) : 1036-1038]** *Email: thulkar@hotmail.com*

***Key words** : Langerhans' cell histiocytosis; Liver; Ultrasound; CT*

The term Langerhans' cell histiocytosis (LCH), earlier known as histiocytosis X, refers to a proliferative disorder characterized by infiltration of one or more organs by mononuclear cells having properties of a unique histiocyte, the Langerhans' cell.[1] The disease usually manifests in infancy and early childhood. Common sites of involvement are bone marrow, lung, skin, liver, spleen and lymph nodes. Clinical manifestations of LCH are varied and depend on the sites and extent of involvement. Hepatic involvement is common in patients with multisystem disease. Hepatic involvement is associated with a high mortality rate in patients with LCH.[1] We describe sonographic and CT scan findings in two paediatric cases of LCH.

CASE REPORTS

Case 1. A three-year-old male child presented with itchy lesions over the scalp, slowly progressing swelling on both sides of the neck, progressively increasing abdominal distension and itchy lesions on the palmar aspect of both hands, for one year, one year, three months and fifteen days duration respectively. He also had discharge from bilateral ear for six months duration. There was no history of bladder, bowel or visual disturbance. On examination, his vital parameters were within normal limit. He had mild pallor and non-tender firm discrete lymphadenopathy involving bilateral jugular groups. There were multiple maculopapular lesions with healing by pigmentations involving the scalp and palmar aspect of both hands. Examination of abdomen revealed firm mildly tender smooth hepatomegaly. He was found to have anemia (Hb≅10.6 gm/dL). Bilirubin, SGPT and albumin levels were normal. However, serum alkaline phosphatase was markedly raised (≅1612 U/L). Serum electrolytes were within normal limits. Ultrasound examination (USG) of abdomen revealed periportal band like hyperechoic lesions along with smooth hepatomegaly (Fig. 1a). No focal hepatic parenchymal lesion was detected. A non-contrast followed by contrast enhanced CT scan of the abdomen revealed periportal hypodense, (HU≅23 to-57), non ehnancing, band like areas (Fig. 1b) corresponding to the hyperechoic lesions seen on USG. The caudate lobe was markedly enlarged. Bone marrow biopsy was normal. Biopsy of skin lesions revealed features of LCH. He has been on chemotherapy as per schedule (Protocol: DAL HX – 83)[2] and is on follow up.

Case 2. A four-year-old male child presented to our oncology department OPD with the history of a soft tissue swelling on the left frontal region, abdominal swelling, polyuria and polydipsia of three years duration. On examination, he was found to be malnourished. He had a firm non-tender fixed subcutaneous swelling in the left

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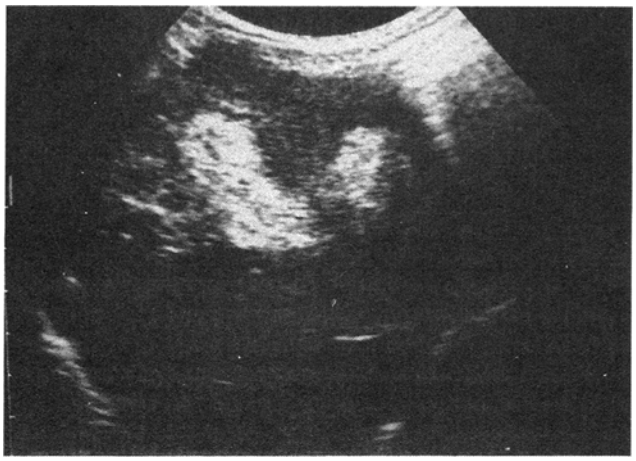

Fig. 1a. Transverse ultrasonogram of liver shows thick hyperechoic bands in the line of portal tracts.

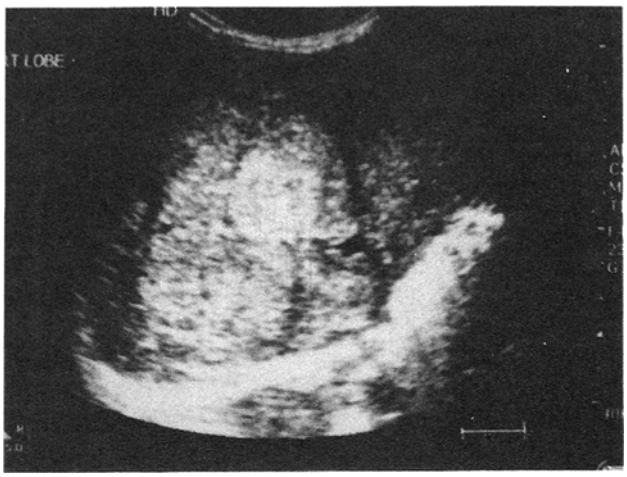

Fig. 2a. Longitudinal ultrasonogram shows predominantly hyperechoic heterogeneous echotexture of liver.

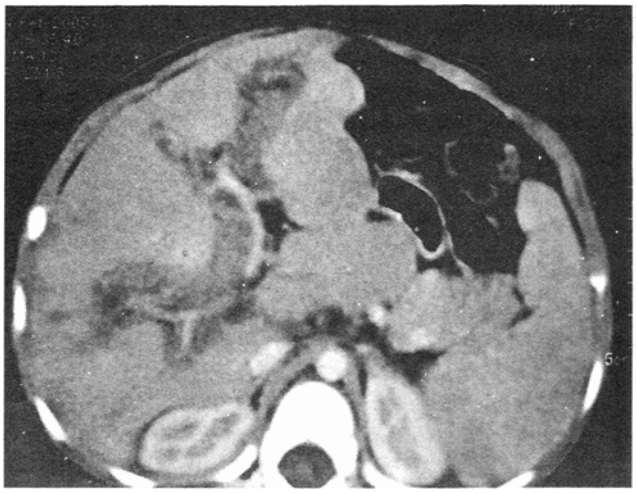

Fig. 1 b. Contrast enhanced axial CT scan shows extensive periportal hypodensities (HU(≅ 23 to -57) that correspond to hyperechoic bands seen on USG. There are occasional areas of fat densities within. Surrounding hypodensities make the portal vein branches standout in contrast. The caudate lobe is significantly enlarged.

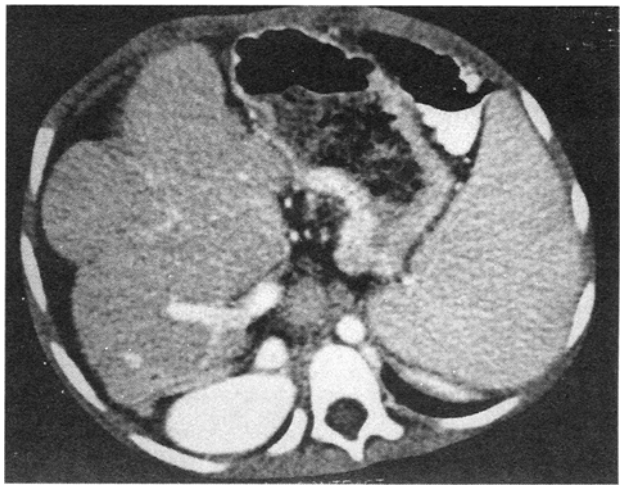

Fig. 2b. Contrast enhanced axial CT scan shows lobulated outline of liver, atrophy of right lobe, hypertrophy of left lobe, prominent portal vein, splenomegaly. These features are suggestive of cirrhosis of liver with portal hypertension.

frontal region. He was also found to have smooth non-tender hepatosplenomegaly. There was no ascites. Pertinent laboratory investigations revealed anemia (HB≅9gm/dL), routine blood biochemistry including serum electrolytes and liver function tests were within normal limit. USG of abdomen revealed lobulated outline of the liver with heterogeneous echogenicity throughout the liver parenchyma (Fig. 2a). The main portal vein and splenoportal axis were prominent with associated splenomegaly. There was evidence of gastroesophageal varices. A contrast enhanced CT scan of the abdomen confirmed the findings of USG. Besides, it revealed atrophy of right lobe of liver with compensatory hypertrophy of left lobe (Fig.2b). There was no focal parenchymal or periportal abnormality detected. USG and CT scan findings were consistent with cirrhosis of liver with portal hypertension. A contrast enhanced CT scan of the head and sella revealed a sharply defined punched out lytic lesion involving both tables of left frontal bone with a hyperdense speck in the center (button sequestrum) (Fig. 2c). The pituitary gland and it's stalk were normal. A biopsy from the left frontal swelling was consistent with a diagnosis of LCH. The bone marrow biopsy was normal. He has been on chemotherapy as per schedule (Protocol: DAL HX - 83) and is on follow up.

DISCUSSION

Children with LCH usually present with hepatomegaly along with multi organ involvement. Liver involvement is relatively common in disseminated LCH (representing 40% to 60% of the cases), affecting most children under 2

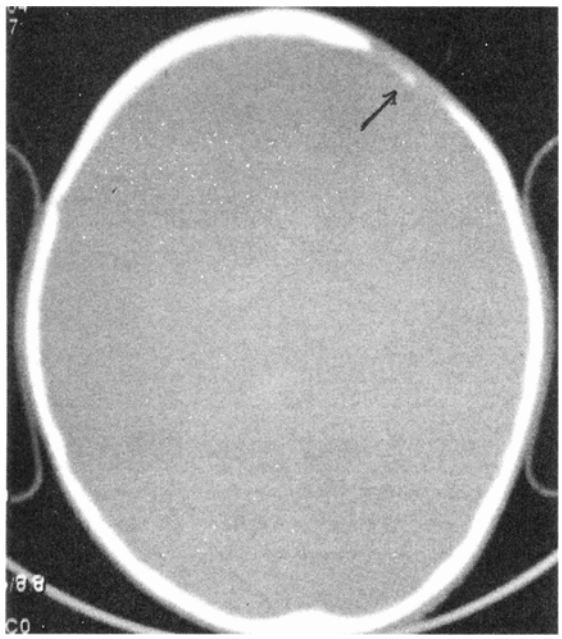

Fig. 2c. Axial CT scan of the head (bone window setting) shows a lytic lesion with sharp margins involving both tables of left frontal bone. A small bony speck (button sequestrum) is seen in the center of the lesion.

years of age.[3] Histopathologically, liver shows periportal infiltration with Langerhans' cells, features resembling sclerosing cholangitis, bile ductular distortion, portal triaditis, variable amount of fibrosis, hepatic nodules or even cirrhosis.[4,5] Liver injury in LCH can be caused by histiocytic infiltration itself (with intrahepatic biliary obstruction) but also extrinsic compression of common bile duct by hilar adenopathy, histiocytic infiltration of extrahepatic bile ducts or rarely by an adverse effect of chemotherapeutic agents used to control the disease.[3] Hepatomegaly is common in LCH. Liver damage in LCH can be independent of local or generalized LCH activity.

Various imaging findings of the liver have been described in children and adults with LCH.[1,4,6,7] Imaging findings depend on the duration of the disease, the main pathological reature and extent of involvement in each case. In children, the liver lesions are characteristically located in the periportal region and have been staged into four different histologic phases: an initial proliferative phase, intermediate granulomatous, xanthomatous phases, and a late fibrous phase.[7]

In the early proliferative phase periportal lesions may appear hypoechoic on USG. On CT, these periportal lesions appear periportal lesions appear hypodense and may show enhancement on post contrast studies suggestive of triaditis. Periportal hypoechogenicity on USG may be band like or nodular, some hypoechoic nodules may look like targets. Periportal hypoechogenicity may be attributed to infiltration by histiocytes and inflammatory cells.[4] Later in the xanthomatous stage, these lesions appear uniformly hyperechoic at sonography, have low attenuation at CT and display characteristics of fat at MR Imaging.[8] This characteristic imaging feature is attributed to the lipid rich nature of the Langerhans' cell in the xanthomatous phase.[7] Kim *et al*[4] described periportal abnormal signal initensity as a common feature on initial MRI in children with disseminated LCH. Besides these typical findings, hepatic parenchymal nodules of varying echogenicity and attenuation have also been described on USG and CT. CT may show hypodense nodular lesions with ring enhancement simulating metastasis or granuloma. The difference in echogenicity, delineation, and contrast enhancement pattern may be due to different histological stage of LCH lesions.[6]

Our first patient demonstrated involvement of periportal region with combined features of granulomatous and xanthomatous phases. Caudate lobe hypertrophy was an associated finding. Imaging features in the second patient were suggestive of sequele of fibrous stage in the form of cirrhosis of liver and portal hypertension. Resolution of hepatic lesions and improvement of hepatic function after chemotherapy have been documented.[1]

Imaging features of hepatic LCH are not widely described. Knowledge of imaging findings in hepatic LCH is useful in the differential diagnosis in a particular case and this also has a definitive role in the follow up of already diagnosed case of LCH.

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