www.ksmrm.org JKSMRM 16(3) : 262-265, 2012
Print ISSN 1226-9751

Case Report

Langerhans Cell Histiocytosis with Pancreatic Involvement: Imaging Findings Including Diffusion-Weighted Imaging

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Langerhans cell histiocytosis (LCH) can affect many different organs. However, LCH with pancreatic involvement is very rare with a few reports about imaging findings. We present a case of multisystemic LCH with pancreatic involvement in a five-week-old infant. Pancreas lesion showed hypoechoic on ultrasonography, low density with poor enhancement on CT, and restricted diffusion on diffusion-weighted imaging. Although LCH with pancreatic involvement is rare, LCH should be considered in the differential diagnosis of pancreatic mass in children.

Index words : Langerhans cell histocytosis · Pancreas · Diffusion-weighted imaging

INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare disorder of children that can affect many organs. It is characterized by abnormal proliferation and infiltration of Langerhans cells and comprises a group of disorders previously known as histiocytosis X, eosinophilic granuloma, Hand-Schuller-Christian disease, and Letterer-Siwe disease (1, 2). Although the exact incidence of LCH is not known, it has been estimated that 1 out of 200,000 children are affected each year in the United States. The clinical spectrum of LCH can be extremely variable, from asymptomatic single system involvement to fulminant multi-systemic disease, often resulting in death. Commonly affected organs include the skin, bone, lungs, liver, spleen, and the pituitary gland. There are several diagnostic tests for LCH including the demonstration of Birbeck granules by electron microscopy or CD1a antigen expression on the cell surface (3).

Because involvement of the pancreas in LCH is rare, only a few cases of sonographically demonstrable lesions of the pancreas have been reported (4–6). This case report illustrates the imaging findings of pancreatic involvement in LCH, including ultrasonography (US), computed tomography (CT), and magnetic resonance (MR) imaging with diffusion-weighted imaging (DWI).

CASE REPORT

A five-week-old male infant presented to the pediatric clinic with granulomatous skin lesions and multifocal ulcerative skin lesions of the fingers, toes, neck, back, and scrotum that were present at birth. The full-term infant was born by Cesarian section after a normal pregnancy. Family history was unremarkable. The skin lesions showed multiple

• Received; August 10, 2012 • Revised; December 19, 2012
• Accepted; January 2, 2013
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hyperpigmented papules and nodules. The infant had intermittent vomiting without palpable mass, abdominal distention, or fever.

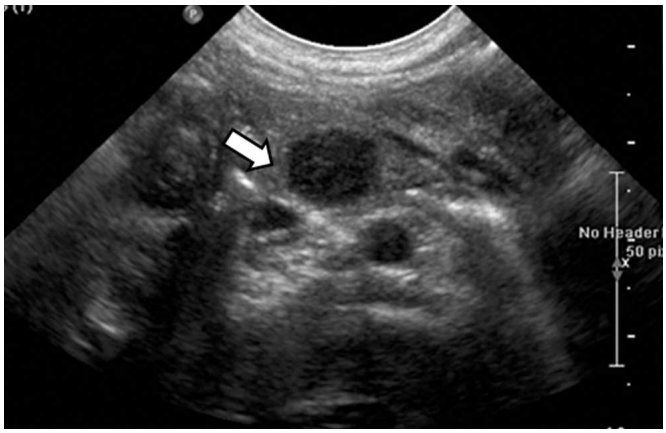
Abdominal US revealed a well-defined hypoechoic mass with internal vascularity in the uncinate process of the pancreas (Fig. 1a and Fig. 1b). A contrast-enhanced abdominal CT scan of portal venous phase was performed to evaluate disease extent in abdomen and it showed a solid lesion with low attenuation and poor enhancement only in pancreas (Fig. 1c). The infant was evaluated by abdominal MR imaging to characterize this lesion using a 1.5 T scanner, including axial T1- and T2-weighted images. Axial DWI was also obtained using b-values of 500 and 1000 sec/mm^2. The pancreatic mass showed low signal intensity on T1-weighted images (Fig. 1d) and intermediate signal intensity on T2-weighted images (Fig. 1e). On DWI, the mass showed diffusion restriction (Fig. 1f). Involvement of other abdominal organs was not noted. Chest CT and a whole body bone scan revealed multiple lesions involving the thymus, lungs, and bone.

Blood and skin lesion cultures showed no infectious organisms. A punch biopsy of a skin lesion demonstrated dermal infiltration of histiocytes (Fig. 1g). Immunohistochemical staining of this infiltrate was positive for CD1a, confirming the diagnosis of LCH.

Chemotherapy was initiated with vinblastine (0.2 mg/kg per week), methotrexate (250 mg/m^2 per two weeks) and prednisolone (1.33 mg/kg per day) for treatment of LCH. After six months of chemotherapy, follow-up abdominal US and CT scans showed a decrease in the size of the pancreatic lesion without any change in the character of the lesion.

DISCUSSION

LCH with pancreatic involvement is very rare. In a



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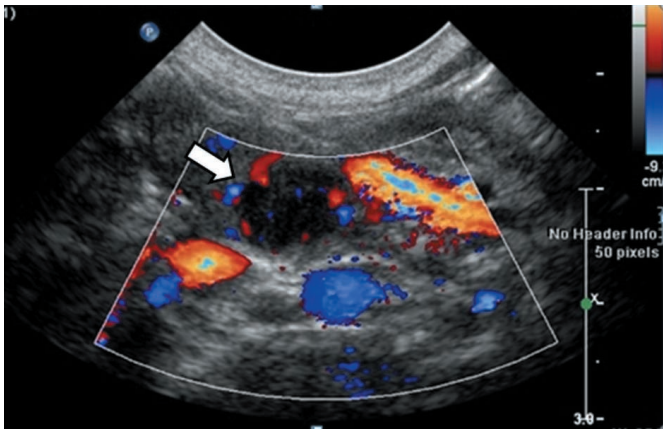

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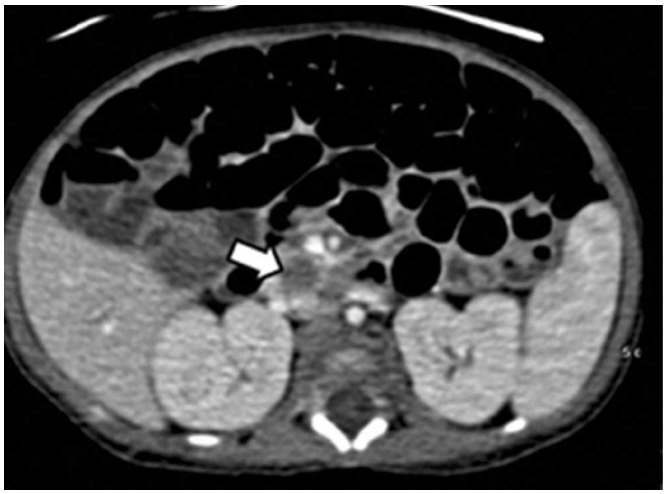

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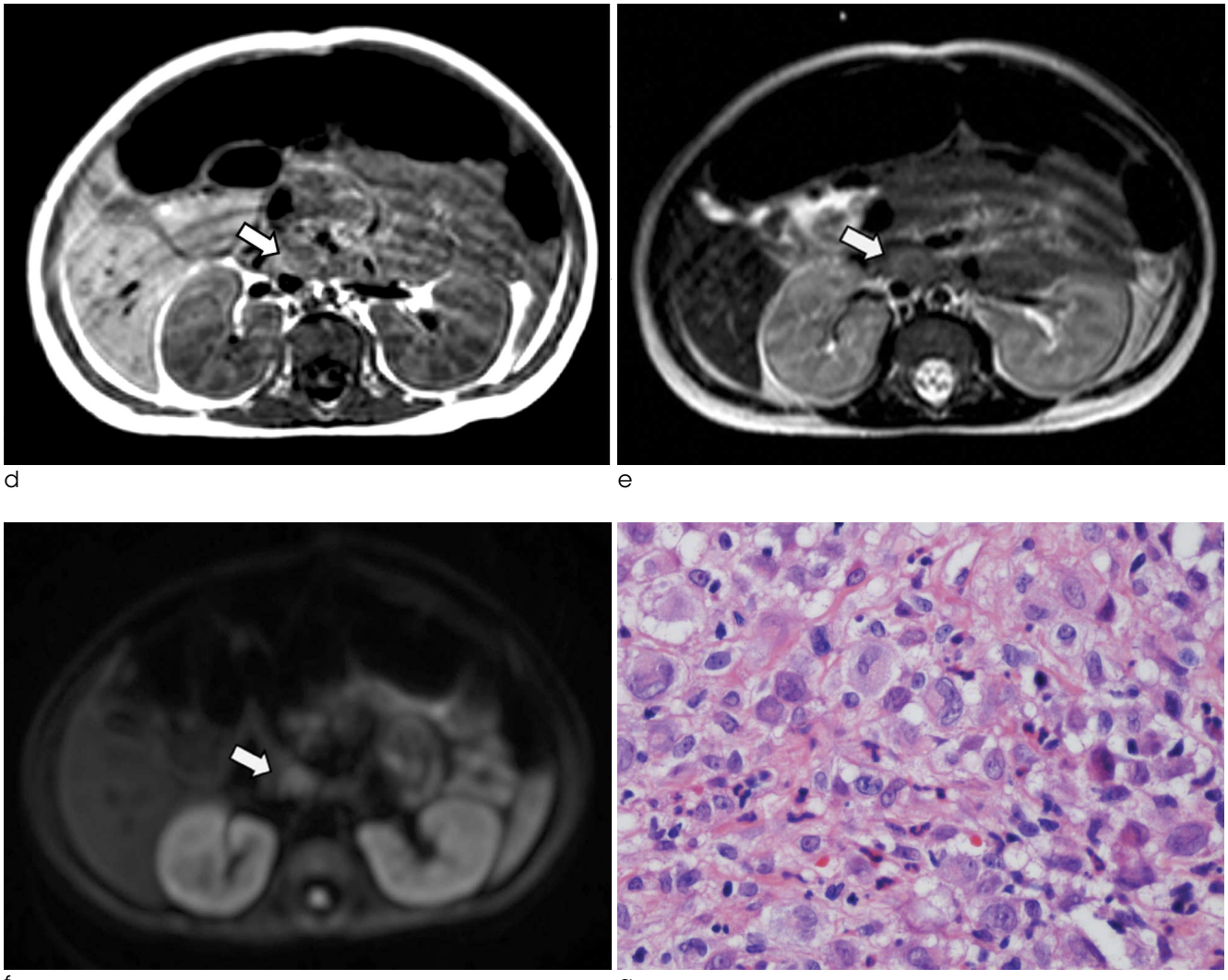
Fig. 1. 5-week-old male infant with granulomatous skin lesions.
a. Abdominal sonograph showing a round hypoechoic mass (arrow) in the uncinate process of the pancreas.
b. Doppler sonograph showing internal vascularity in the pancreatic lesion (arrow).
c. Axial image of the abdominal CT scan showing a round lesion with low attenuation and poor enhancement at the uncinate process (arrow) of the pancreas.



d-f. Abdominal MRI showing a solid mass (arrow) with low signal intensity on the T1-weighted image (**d**) and intermediate signal intensity on the T2-weighted image (**e**). DWI showing the lesion as high signal intensity on the b-value 1000 sec/mm^2 image (**f**), suggestive of diffusion restriction.
g. Skin biopsy showing dermal infiltration of histiocytes suggestive of Langerhans cell histiocytosis.

multicenter survey of LCH, there were no cases with pancreatic infiltration among the eight cases of LCH with gastrointestinal tract involvement (7). Keeling et al. presented autopsy findings for 12 cases of LCH with gastrointestinal involvement, but none of those 12 cases showed pancreatic abnormalities (8). There are only three reported cases presenting pancreatic invasion of LCH. Pancreatic infiltration of Langerhans cells was confirmed by autopsy for two of these cases (4, 5). Muwakkit et al. reported the sonographic appearance of pancreatic lesions in a known case of LCH and described the case as a cystic lesion (6). The cystic lesion decreased in size after chemotherapy. However, whether the lesion represented a primary LCH lesion or a pseudocyst secondarily caused by Langerhans cell infiltration was not established.

In our patient, abdominal US demonstrated a hypoechoic mass with internal vascularity in the uncinate process of the pancreas. An abdominal CT scan also revealed a solid mass with poor enhancement. MR imaging findings of LCH involvement in the pancreas have not yet been reported. We evaluated the pancreatic mass by MR imaging, and it showed low signal intensity on T1-weighted images, and intermediate signal intensity T2-weighted images, respectively. The mass also showed diffusion restriction on DWI.

These results implied that the pancreatic lesion in our patient was a solid mass and not a pseudocyst. Although, lack of direct pathological proof is a limitation of our case report. Solid pancreatic tumor is very rare in infants (9). And pancreatoblastoma or lymphoma involvement in pancreas can be included in the differential diagnosis. But pancreas biopsy is invasive and risky procedure in infant, and the skin biopsy proved the typical diagnosis of LCH. Furthermore, the lesion improved with systemic chemotherapy; therefore, we concluded that the lesion likely represented LCH infiltration.

Prognosis of LCH depends on the number of affected organs and the degree of dysfunction of involved organs (10). However, pancreatic involvement is very rare, and the symptom or prognosis of pancreatic involvement is not yet precisely reported. Therefore, the clinical relevance of our imaging findings in the pancreas is not yet clear, more cases will need to be studied in order to determine the importance of our findings.

This case report demonstrates the imaging findings of LCH with pancreatic involvement, including US, CT, and MR imaging with DWI. Although rare, LCH infiltration of the pancreas should be considered in the diagnosis of solid pancreatic masses in children.

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대한자기공명의과학회지 16:262-265(2012)

소아에서의 랑게르한스세포 조직구증식증의 췌장 침범 : 확산강조 자기공명영상을 포함한 영상소견에 관한 증례보고

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랑게르한스세포 조직구증식증 (Langerhans cell histiocytosis, LCH)은 다양한 장기를 침범할 수 있다. 그러나 췌장의 침범은 매우 드물어 영상 소견에 대한 보고도 적다. 이에 본 저자들은 생후 5주 유아에서 췌장을 포함한 다장기 침범 LCH 증례를 보고하고자 한다. 췌장 병변은 초음파상 저에코성 병변으로 보였고, CT상 저음영으로 조영증강이 잘 되지 않았으며, 확산강조영상에서 확산 제한을 보였다. 비록 LCH의 췌장 침범은 매우 드무나, 소아에서 췌장 종괴가 있을 때 감별진단으로 고려해야 한다.

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