We presented the abdominal computed tomography (CT) findings of four cases of Langerhans’ cell histiocytosis (LCH) in children (age ranged from 11 month-old to 2 year-old, two boys and two girls), who clinically had either fever, skin rash, hepatosplenomegaly, persistent jaundice, elevated liver enzymes, or lymphadenopathy. CT showed variable findings, including: hepatomegaly, splenomegaly, liver cirrhosis, hypoattenuating or ring-enhancing, nodular lesions in the liver, hepatic perportal low-density lesions and paraaortic lymph node enlargement.

We assumed that the radiologic appearances of hepatic lesions in LCH might correlate with the histopathological features of hepatic involvement in LCH. Our four cases probably represented the different phases or stages of hepatic involvement in LCH.

Key words: Children; Liver, CT; Histiocytosis

Langerhans’ cell histiocytosis (LCH) refers to proliferation of a unique histiocyte, the Langerhans’ cell [1, 2]. In most cases LCH has systemic involvement in childhood. More than 90% of patients with disseminated disease are less than 21 years old [2]. CT findings in four children with abdominal LCH constitute the subject of this report.

CASE REPORT

Case 1
A 2-year-old girl suffered from head injury and retro-auricular mass for 10 days. The physical examination revealed left temporo-occipital scalp hematoma. Surgical intervention of the head was carried out to remove the left temporo-occipital soft tissue mass, which was proved to be Langerhans’ cell histiocytosis pathologically. On laboratory examination, the liver enzymes were elevated unexpectedly. The serum glutamic-oxaloacetic transaminase (SGOT) was 155 U/L (normal, 0-34 U/L), serum glutamic-pyruvic transaminase (SGPT) 152 U/L (normal, 0-36 U/L), and alkaline phosphatase 112 U/L (normal, 28-94 U/L). The other laboratory tests were all within normal limits. Abdominal ultrasound revealed hepatosplenomegaly. Abdominal CT showed mild hepatomegaly and splenomegaly without focal density changes or abnormal enhancement after use of contrast medium.

Case 2
A 1-year-old boy presented with persistent jaundice for two months. The physical examination revealed jaundiced skin and a hard, distended abdomen. Serum levels of total bilirubin (10.5 mg/dl; normal 0.1-1.2), direct bilirubin (7.0 mg/dl; normal < 0.5), SGOT (313 U/L; normal 0-34), SGPT (462 U/L; normal 0-36), alkaline phosphatase (1771 U/L; normal
28-94), and total cholesterol (868 mg/dl; normal < 200) elevated. He had a history of right temporal skull LCH removed two and a half months ago. His jaundice developed two weeks after the operation. Abdominal sonography showed hepatosplenomegaly. Abdominal CT showed hepatomegaly and low-density lesions in the periportal areas (Fig. 1).

LCH with liver involvement was considered and liver biopsy was planned. However, since then the patient had lost follow-up.

Case 3
A 2-year-old girl experienced low-grade fever for one month without symptoms/signs of acute gastroenteritis or upper respiratory infection. Her pediatrician found hepatosplenomegaly and lymphadenopathy of bilateral neck and inguinal regions on physical examination. Laboratory studies revealed normal liver function tests but leukocytosis. The white blood cell count was 29700/ul (normal, 6700-11800/ul), and the C-reactive protein was 101.6 mg/l. Abdominal sonography showed mild hepatomegaly and splenomegaly. Abdominal CT scan showed low-density lesions in the enlarged spleen and enlarged lymph nodes at the paraaortic region (Fig. 2). Mild hepatomegaly was also observed. Laparoscopic spleen biopsy and neck lymph node biopsy were performed and disclosed Langerhans’ cell histiocytosis pathologically.

Case 4
An 11-month-old boy had skin rash over his trunk and fever for two weeks. Skin rash biopsy disclosed LCH. The physical examination revealed hepatosplenomegaly. Serum levels of SGOT (75 U/L), SGPT (257 U/L), and alkaline phosphatase (668 U/L) were elevated. The abdominal CT scan showed low-density lesions in periportal regions and liver parenchyma. Some of the hepatic lesions showed rim-enhancement after intravenous administration of contrast medium (Fig. 3a).

The boy received chemotherapy with Oncovin and prednisolone due to disseminated LCH and regularly followed up at OPD. Two years later, persistent splenomegaly and coarse liver surface with periportal fibrosis were shown by sonography at OPD. Abdominal CT scan showed lesions of low attenuation in cirrhotic liver parenchyma and periportal area as well as splenomegaly (Fig. 3b). Liver biopsy revealed advanced liver cirrhosis.

DISCUSSION
LCH represents a spectrum of histiocytic infiltration disorder ranging from localized aggregation to dissemination of various tissues. Lipids, mainly cholesterol, often present in the histiocytic deposits within the characteristic foamy histiocytes [3].

The radiological appearance of hepatic lesions in LCH is due to aggregation of foamy histiocytes [2, 4]. Hepatic masses containing fat density can occur in lipomas, angiomyolipomas, liver cell adenomas, myelolipomas, and hepatocellular carcinoma with fatty metamorphosis [14].

In the four previously reported cases of LCH in the literature [4-7], CT commonly showed hypoattenuation.

Figure 1. Case 2. Contrast-enhanced abdominal CT shows hepatomegaly and low-density lesions in the periportal areas (arrows).

Figure 2. Case 3. Contrast-enhanced abdominal CT shows splenomegaly with a low-density lesion (arrow). Paraaortic lymph nodes enlarge on both sides.
ating nodular lesions in the liver and spleen and some had hepatic periportal low density lesions. The ring enhancement on contrast-enhanced CT has also been mentioned [7]. In our four patients with LCH, variable CT findings were shown, which were mostly consistent with those mentioned in the literature. Besides, there were simply hepatomegaly and splenomegaly, paraaortic lymph nodes enlargement and liver cirrhosis. The nodular hypodense lesions in the liver and spleen were consistent with that of fat.

Liver involvement is common in disseminated LCH, representing 40% to 60% of cases, most often affecting children under 2 years of age. Histopathological features of liver involvement have been staged in four phases: (1) a proliferative phase, or infiltration of the portal tract by histiocytes, (2) a granulomatous phase, (3) a xanthomatous phase, and (4) a late fibrous phase leading to cirrhosis [8].

We assumed that the low-attenuation lesion is consistent with the proliferative and xanthomatous histological phases. Our cases would represent the different stages of liver involvement, from simply hepatomegaly, hepatic nodular and periportal low-attenuating lesion, ring-enhanced lesion, to the last stage of liver cirrhosis. Difference in density, delineation, and contrast enhancement of the lesions might be due to different histological stages of LCH lesions. Further investigation will be necessary to answer this question. If CT features of disseminated LCH with liver involvement correlate well with the histopathological features, it could probably be a useful prognostic indicator.

Patients with localized or multifocal disease involving the skin, skull, or long bones but without organ dysfunction may recover without medical intervention [9]. Surgical excision of the localized mass can be both diagnostic and curative. But, those patients with disseminated disease and evidence of organ dysfunction manifested by pneumonia, jaundice, and/or hematological cytopenia are likely to succumb to their disease or its complication despite of aggressive medical therapy [9, 10]. Hepatomegaly alone does not portend a poor prognosis nor does it correlate well with histological findings in liver [11]. Liver dysfunction, characterized by cholestasis, jaundice, pruritus, and/or portal hypertension, usually develops over months or years [10-13]. Reversal of hepatomegaly and improvement of hepatic function after chemotherapy have been reported [2, 5]. Spontaneous resolution of hepatomegaly and of multiple hypodensate hepatic lesions shown by CT also has been reported [2].

We conclude that, although very rare, LCH should be included in the differential diagnosis of widespread space-occupying lesions in liver and spleen, when imaging findings show low-density lesions with features consistent with focal fatty infiltration or fat-containing tumor, especially in the presence of typical cutaneous, osseous, or pulmonary lesions, or associated with lymphadenopathy, or

Figure 3. a. Case 4. Abdominal CT with contrast enhancement shows multiple ring-enhanced low-attenuating lesions in liver parenchyma (arrows) and periportal low-attenuating lesions (arrowheads). b. Case 4. Two years later, abdominal CT shows progressive change of low-attenuating lesion in liver parenchyma and periportal areas, as well as splenomegaly. Liver biopsy revealed advanced liver cirrhosis.
evidence of hepatic dysfunction with or without hepatosplenomegaly.

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小兒腹腔內藍蓋罕式組織細胞症的電腦斷層表現：
四病例報告

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我們提出四位兒童罹患藍蓋罕式組織細胞症之病例報告，年齡從11個月至2歲。臨床上，
他們通常表現出下例症狀，如：發燒、皮疹、黃疸、肝脾腫大、肝功能指數上升和淋巴結腫大
等。腹部電腦斷層的表現包括肝脾腫大，肝內低密度結節病灶合併環狀顯影，門脈旁之低密度
病灶，以及淋巴結腫大等。

我們認為這些影像表現可能與藍蓋罕式組織細胞症之病理變化有相當程度的關聯。這四位
病人或許代表著肝臟之藍蓋罕式組織細胞症之不同面像或階段。

關鍵詞：小兒；肝臟；電腦斷層攝影；組織細胞症