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EDUCATION AND IMAGING

Gastrointestinal: Extensive abdominal involvement as an initial presentation of Langerhans' cell histiocytosis

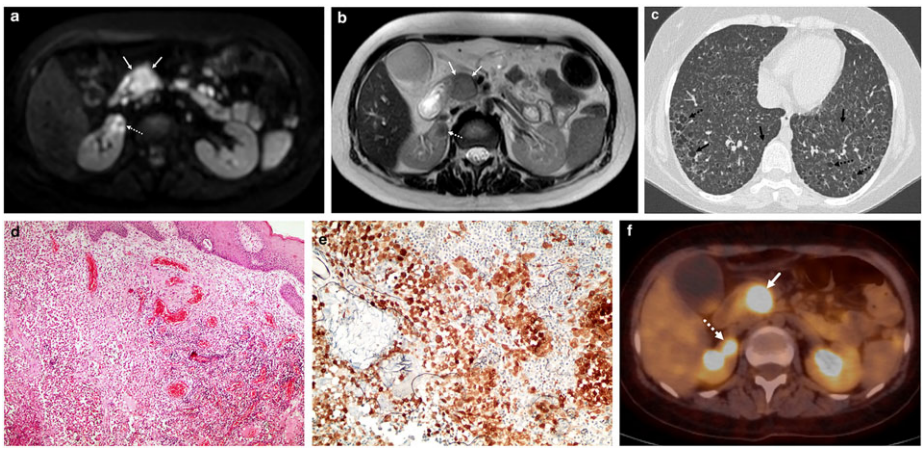

Figure 1 Figure 1 Axial diffusion-weighted (a) and T2-weighted (b) magnetic resonance images of the abdomen from the level of pancreas show the pancreatic mass (arrows) and cortical lesion of the left kidney (dashed arrow). (c) Axial unenhanced chest computed tomography with lung window settings shows multiple lung nodules (dashed black arrows) and bizarre shaped air cysts (black arrows). (d) Axial fused 18 F-FDG PET-computed tomography image shows abnormal accumulation of the tracer in the pancreatic mass (arrow) and kidney lesion (dashed arrow). (e) Malignant histiocytic cell infiltration accompanied by eosinophilic inflammatory cells are seen, which are completely filling the dermis (hematoxylin and eosin × 4). (f) CD1a expression is seen in malignant histiocytic cells (DAB × 20).

An 18-year-old female presented with a 2-week history of increasing fatigue, jaundice, oral mucosa macules, epigastric pain, and vomiting. The patient's medical history revealed a weight loss of 6 kg in 1 month without a history of chronic disease, previous surgery, or smoking. Physical examination revealed multiple macules of oral mucosa and epigastric tenderness. Laboratory tests of liver function showed elevated enzyme levels (aspartate aminotransferase 84 IU/L; normal value < 42 IU/L, lactate dehydrogenase 344 U/L; normal range 135–210 U/L, gamma-glutamyl transferase 182 U/L; normal range 6–42 U/L) and direct bilirubin levels (direct bilirubin: 14.75 mg/dL, normal range < 0.3 mg/dL). Abdominal ultrasonography revealed diffuse heterogeneity within the right lobe of the liver. Abdominal magnetic resonance imaging obtained for further investigation showed pancreatic mass, heterogeneity in liver parenchyma, intrahepatic and extrahepatic biliary dilatations, and multiple hypovascular lesions < 1 cm in both kidneys (Fig. 1). Chest computed tomography (CT) was obtained to explain the reticular and nodular pulmonary opacities seen on chest X-ray. CT showed multiple poorly defined lung nodules and bizarre shaped air cysts (Fig. 1c). Based on the clinical and imaging findings, diagnosis of Langerhans' cell histiocytosis (LCH) was suggested and diagnosis was confirmed after histopathological examination of the tissue obtained through biopsy of oral mucosa macules (Fig. 1 d,e). Severity of the disease was evaluated by 18 (18F) fluorodeoxyglucose (FDG)-positron emission tomography (PET)/CT that showed abnormal FDG uptake in the lung nodules, pancreas, and kidney lesions (Fig. 1f).

Langerhans' cell histiocytosis is a rare disorder with unknown etiology. It is characterized by atypical histiocytic infiltration, as in our case. In cases with LCH, extensive involvement of the abdominal organs is exceedingly rare. To the best of our knowledge, this is the first case who initially presented with extensive abdominal involvement of LCH. In the presence of multifocal abdominal masses and lung cysts, LCH should be considered in the diagnosis.

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