MR cholangiography in the diagnosis of sclerosing cholangitis in Langerhans’ cell histiocytosis

Abstract Langerhans’ cell histiocytosis (LCH) is a disorder of histiocytic proliferation that primarily affects infants. Imaging findings of a rare case of lung and liver involvement in an adult are presented. High-resolution computed tomography (HRCT) of the lungs showed confluent thin-walled cystic air spaces compatible with advanced LCH. Liver CT and MRI revealed unspecific signs of fatty infiltration. Irregular widening of peripheral bile ducts was displayed in breath-hold MR cholangiography. This pattern is considered characteristic for sclerosing cholangitis and should support the diagnosis of LCH in case of concomitant cystic pulmonary disease, even in adult patients.

Keywords Langerhans’ cell histiocytosis · Sclerosing cholangitis · MR cholangiopancreatography

Introduction Langerhans’ cell histiocytosis (LCH), previously known as histiocytosis X, encompasses clinically heterogeneous but histologically similar disorders including eosinophilic granuloma, Hand-Schüller-Christian lipogranulomatosis and Abt-Letterer-Siwe disease. Systemic disease with a leukemia-like course primarily affects infants and is rarely found in adults [1, 2, 3, 4]. We present the imaging findings of lung and liver involvement in an adult patient with rapidly progressing LCH.
Case report

A 68-year-old female patient presented with fever, night sweat, weight loss, leukocytosis, elevated erythrocyte sedimentation rate (ESR), anemia, and elevated liver enzymes. Diabetes insipidus requiring vasopressin substitution had been diagnosed 3 years previously. A chest plain film showed ill-defined linear opacities in the right upper lobe with cavitated zones, suggesting tuberculosis. A biopsy during thoracoscopy revealed active fibrosing alveolitis with proliferating pneumocytes and focal fibrosis. Microbiological studies were negative; hence, prednisolone was given. An initial endoscopic retrograde cholangiopancreatography (ERCP) and liver sonography were unremarkable except for mild hepatomegaly and gallstones. A bone marrow biopsy demonstrated focal fibrosis and numerous multinucleated plasmocytes.

The liver enzymes further increased over a period of 4 months to a maximum of GOT 97 U/l, GPT 285 U/l, LDH 420 U/l, AP 2050 U/l, gamma-GT 2418 U/l, and bilirubin 4.5 mg/dl. The patient developed mild jaundice and an itching maculopapular rash with ulcerous skin lesions. Laboratory tests ruled out viral or other infection. There was no evidence of concomitant anterior pituitary deficiency, skeletal involvement, systemic amyloidosis, hepatocellular carcinoma, or other malignancies. A mild thrombocytopenia (110,000/μl) developed. The imaging findings are described herein. Percutaneous liver biopsy revealed granulomatous inflammation with histiocytic infiltrates, periportal fibrosis, destructed next to proliferating bile ducts and focal steatosis, compatible with (primary) sclerosing cholangitis (Fig. 1). Lung biopsies were moderately and the liver specimen markedly immunoreactive for S-100 protein, so the diagnosis of Langerhans’ cell histiocytosis was established. Ultrastructural analysis was not performed. Despite steroid therapy, the patient died 6 months after onset of pulmonary and hepatic involvement.

In high-resolution computed tomography (HRCT) of the lung circumscribed areas of low attenuation were found bilaterally with predominance in the apex and the upper segment of the lower lobe, resembling localized peribronchial emphysematous rarefaction of lung parenchyma (Fig. 2). Confluent thin-walled bullous changes were found rather than small cystic air spaces. There was no evidence of micronodules, pneumothorax, lymphadenopathy, or pleural effusion. Ultrasonography of the liver showed mild hepatomegaly with an inhomogeneous, partly hypechoic echotexture and some intrahepatic bile duct dilatation. Liver parenchyma was markedly inhomogeneous on unenhanced CT. Contrast-enhanced CT scans allowed better definition of finger-shaped areas of low attenuation suggesting circumscribed fatty infiltration (Fig. 3). In MRI a focal area of hyperperfusion was depicted on T1-weighted images (Fig. 4b) immediately after administration of Gd-DTPA (Schering, Berlin, Germany). Hypointense infiltration of the liver parenchyma was best shown in the delayed contrast-enhanced T1-weighted series using spectral fat saturation (Fig. 4c), whereas inhomogeneity was only moderate without fat suppression (Fig. 4a, b). Opposed-phase imaging was not performed. T2-weighted images were slightly inhomogeneous due to focal bile duct dilatation.

Endoscopic retrograde cholangiopancreatography (ERCP) was largely unremarkable except for moderate rarefaction of non-dilated intrahepatic bile ducts. Breath-hold MR cholangiopancreato- graphy (MRCP) using the rapid acquisition with relaxation enhancement (RARE) technique [5] revealed peripheral focal cholestasis in each section of the liver (Fig. 5). Multiple slightly dilated bile ducts were displayed showing wall irregularities, filling defects and high-grade obstruction. No extensive ductal dilatation was present. Few segmental branches were displayed with loss of normal tapering. The remaining ducts appeared irregularly defined, partly “ragged,” with ductal stenosis and filling defects next to normal non-dilated ducts. The extrahepatic bile ducts appeared unremarkable with the exception of a short signal void in the CBD next to the hepatic ducts’ confluence. Breath-hold MRCP was performed on a 1.5-T system (Magnetom Vision, Siemens, Erlangen, Germany) equipped with a phased-array surface coil. Half Fourier acquisition single-shot turbo spin-echo (HASTE) multislice sequences (effective TE 95 ms, echo train length (ETL) 128, echo spacing 11.9 ms, 9 slices, section thickness 4 mm, field of view (FOV) 300 mm, acquisition time 18 s) were applied in coronal and semicoronal (LAO 30°) view. Repeated projection acquisitions