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An unexpected cause of heterogeneous liver

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A 43-year-old man, with a history of smoking abuse and traumatic brain injury secondary to a car crash, was referred to our department for suspicion of metastatic liver tumor. He presented right hypochondrial pain since a few weeks, associated to isolated elevation of gamma-GT. Liver MRI showed a multinodular liver, with all nodules inferior to 1 centimeter, with few excavations (fig1). Liver biopsy revealed an inflammatory nodule infiltrated by eosinophils and histiocytes, which were CD68+, PS100+ and CD1a+, without tumoral tissue neither cholangitis, thus affirming the diagnosis of hepatic Langerhans cell histiocytosis (LCH) (fig2). Body-scan and PET-scan showed a right mastoid cavity infiltration, suggesting a multifocal LCH.

LCH is a rare disease that mainly affects children. In adults, the prevalence is about 1 to 2/100 000. When liver involvement occurs, LCH is always multisystemic. Two types of liver injury exist. At early stage of the disease, hepatic involvement presents with hepatomegaly due to the direct infiltration by Langerhans cells, with good response to chemotherapy. At a later stage, infiltrations are around the bile ducts, resulting in primary sclerosing cholangitis-like,

with potential progression to biliary cirrhosis and finally liver failure. This stage is less sensitive to chemotherapy. Since 2010, BRAF mutation (V600E) was identified in more than half of LCH patients, who be responsive to treatment with vemurafenib, a BRAF inhibitor (2 - year progression - free survival rate 86%, overall survival rate was 96%). Acid ursodesoxycholic could be proposed to prevent progressive irreversible damage of cholestasis. In rare cases, liver transplantation is performed.

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Fig 1 : Contrast-enhanced MRI obtained at the portal venous phase and showed multifocal nodular liver, with some included peripheral ring-like enhancement with a central low signal intensity

Fig 2: Histopathology examination of liver biopsy revealed an inflammatory nodule infiltrated by eosinophils and histiocytes, which destroy liver parenchyma (big square, HES coloration x 400). immunohistochemistry examination revealed histiocytes marking by CD1a- antibodies (little square, x200)



