A rare cause of high liver stiffness

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Case presentation

A 71-year-old man with a past medical history of arterial hypertension was referred to our outpatient clinic for a suspected liver disease, based on biochemical and radiological findings. He was poorly symptomatic with discrete weight loss due to reduced appetite. Physical examination revealed a good general condition, hepatosplenomegaly, overweight (BMI 27) and enhanced waist circumference. Biochemical examination showed pancytopenia (hemoglobin 11,4 g/dL [13.3-16.7 g/dL], white cells count 3870/µL [4-10x10^3/µL], platelets count 38000/µL [150-350x10^3/µL]) and cholestasis (gGT 147 U/L [<60U/L], alkaline phosphatase 152 U/L [40-130 U/L], total bilirubin 2.1 mg/dL [<1.2mg/dL]). ALT and AST levels were normal. The usual liver biochemical etiology tests were negative unless elevated type M immunoglobulins and an antinuclear antibody titer of 1/160. Doppler-abdominal ultrasonography showed a heterogeneous liver (Figure 1A), large splenomegaly (Figure 1B) and several enlarged lymph nodes. Liver vascularization was normal. Differential diagnosis included a chronic liver disease (seronegative primary biliary cholangitis, fatty liver disease or cardiac liver disease) and a hematological disorder infiltrating the liver. Median liver elasticity (E), evaluated by transient elastography (TE), was compatible with cirrhosis (22.8 kPa) while controlled attenuation parameter (CAP) evaluation suggested slight steatosis (Figure 1C). What do you recommend?

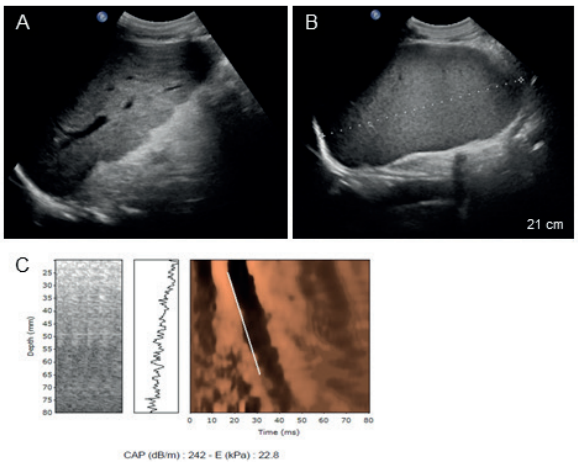

Figure 1. — (A) Liver and (B) spleen ultrasonography; (C) controlled attenuation parameter (CAP) and elasticity (E) by transient elastography.

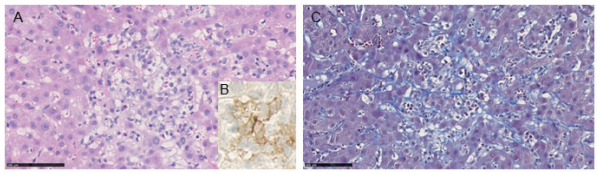

Figure 2. — (A) Liver section with hematoxylin and eosin staining and (B) CD1a immunohistochemistry; (C) Masson trichrome staining.

Diagnosis

Liver histology showed histiocytic infiltration (Figure 2A) positive for CD1a and protein S100 in portal tracts and liver parenchyma (Insert figure 2B), compatible with Langerhans cell histiocytosis (LCH). Surprisingly, only slight pericellular fibrosis (Figure 2C) was noticed. *NRas* but no *BRaf* mutation was detected in liver histiocytic cells. Systemic staging by F-18 FDG PET/CT did not evidence any hypermetabolic activity. The patient was transferred to the hematology department for a treatment with vinblastine chemotherapy and steroids.

We report thus a case of hepatic LCH demonstrated by liver biopsy associated with spleen and lymph nodes involvement. While suspected by TE results, advanced liver fibrosis was not confirmed by liver histology. LCH is a rare entity with a poor prognosis characterized by the clonal expansion of dendritic cells infiltrating any organ of the body. Liver involvement is well described in children but is poorly recognized in adults (1). This new false positive results of liver fibrosis evaluation by TE due to histiocytic cell infiltration could be added to the other well described causes such as transaminases flares, liver steatosis or necroinflammation, recent food intake, hepatic congestion, high body mass index (2). F-18 FDG PET/CT is used for the staging and therapy response assessment in histiocytosis with a high sensitivity, especially for the bone lesions and lymph node

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invasion, but is less established for hepatic LCH (3). Hematological disease affecting the liver should be considered in case of cholestasis especially when associated with pancytopenia, lymph nodes enlargement and splenomegaly.

Abbreviations

Transient elastography (TE), elasticity (E), Langerhans cell histiocytosis (LCH).

Disclosure

The authors do not have any conflict of interest regarding this manuscript.

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