1130-0108/2016/108/8/494-495
Revista Española de Enfermedades Digestivas

Rev Esp Enferm Dig
2016, Vol. 108, N.º 8, pp. 494-495

PICTURES IN DIGESTIVE PATHOLOGY

Fibrolamellar hepatocellular carcinoma: a rare entity diagnosed by abdominal ultrasound

Esther Estrella-Díez[1], Francisco Javier Álvarez-Higueras[1], Gema Marín-Zafra[3], Águeda Bas[2], María Carmen Garre-Sánchez[1], Juan Egea-Valenzuela[1], Daniel José Nova-López[4], Elena Navarro-Noguera[1], Gonzalo Antón-Ródenas[1] and Luis Fernando Carballo-Álvarez[1]

[1]Digestive Diseases Service, [2]Pathology Service, [3]Medical Oncology Service and [4]Emergency Department. Hospital Clínico Universitario Virgen de la Arrixaca. El Palmar, Murcia. Spain

CASE REPORT

We present the case of a 16-year-old male with no previous medical history. He was referred from another hospital because of 4 kg weight loss, jaundice and upper right abdominal pain. He presented mildly elevated liver enzymes, negative hepatotropic viruses and normal levels of alpha-fetoprotein. A thoraco-abdominal computed tomography had been made in his reference hospital finding multiple nodular lesions in both lungs and a hepatic mass (14 x 10 cm). The patient was sent to us for diagnostic ultrasound-guided puncture.

In our study we found a solid, heterogeneous and encapsulated hepatic mass (13 x 10 cm). The lesion was mainly isoechogenic with hyperechogenic areas, located on left liver lobe and segment 4 (Fig. 1), and there were no signs of chronic hepatopathy or portal or biliary permeability alterations. Using contrast enhanced ultrasonography we found peripheral hyperenhacement during arterial phase with progressive wash-out beginning at 2 minutes and 30 seconds (Fig. 2). The study was therefore suggestive of a solitary liver mass with malignant behavior: fibrolamellar hepatocellular carcinoma (FLHC) *vs* mesenchymal tumor (sarcoma). Fine needle puncture aspiration was made and immunophenotypic (Hep-par-1, CK7, Glypican-3 and CD68) and cytomorphologic findings were compatible with FLHC (Fig. 3). The patient was sent to the oncologist and is under oxaliplatin, folinate calcium and 5-fluorouracil.

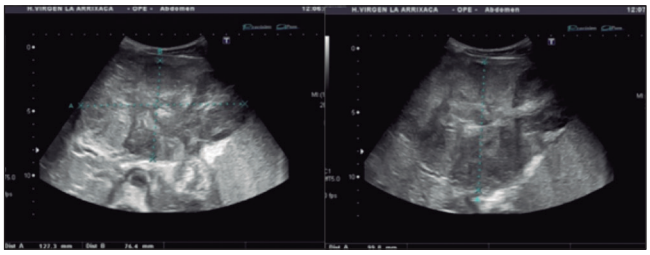

Fig. 1. Homogeneous and regular edges liver with a solid, heterogeneous, nodular and capsulated space occupying lesion (13 x 10 cm) on the left lobe and segment IV.

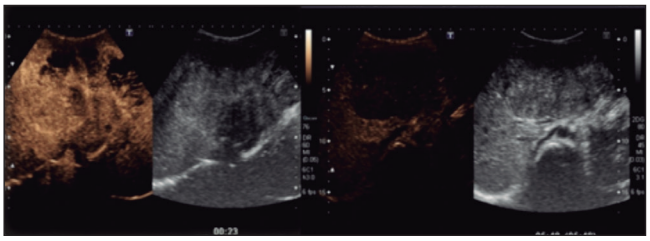

Fig. 2. Using CEUS in the arterial phase we found peripheral hyperenhancement followed by progressive centripetal filling and a central area with hypoenhancement. In the venous phase there was an almost complete washing of the lesion.

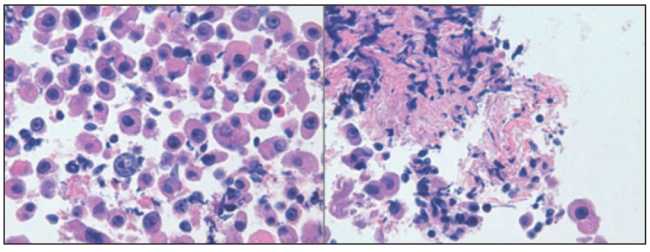

Fig. 3. Pathological specimen showing hepatocytes with big, granular and eosinophilic cytoplasm, resembling oncocytic cells. On the right side fibrotic reaction breaks hepatocytes (nuclei can be identified as chromatin threads).

DISCUSSION

FLHC is a histologic type of hepatocellular carcinoma that represents only 1% of all the primary liver tumors, with an incidence of 0.02 cases/100,000 individuals (1). This tumor is caused by DNAJB1-PRKACA mutation on chromosome number 9, with deletion in 8 genes: the most frequent are MUC4, associated to other gastrointestinal tumors, and GOLCA6L2 (2). The most commonly affected population is individuals under 40 years (65-85%) with no previous history of hepatopathy (3). Up to 70% of the cases are diagnosed in advanced stages (4) as symptoms are unspecific (abdominal pain 21-74%), levels of alpha-fetoprotein are normal and no signs of hepatopathy are present (1). Differential diagnosis must be done with nodular focal hyperplasia, hemangioma and classic hepatocellular carcinoma. First-choice treatment is surgical resection (1,5).

REFERENCES

1. Herman P, Lopes Chagas A, Vinicius M, et al. Surgical treatment of fibrolamellar hepatocellular carcinoma: An underestimated malignant tumor? Hepatobiliariy Pancreat Dis Int 2014;13:618-21. DOI: 10.1016/S1499-3872(14)60294-0
2. Darcy DG, Chiaroni-Clarke R, Murphy JM, et al. The genomic landscape of fibrolamellar hepatocellular carcinoma: Whole genome sequencing of ten patients. Oncotarget 2015;6(2):755-70.
3. Eggert T, McGlynn KA, Duffy A, et al. Fibrolamellar hepatocellular carcinoma in the USA, 2000-2010: A detailed report on frequency, treatment and outcome based on the surveillance, epidemiology, and end results database. United European Gastroenterology Journal 2013;1(5):351-7. DOI: 10.1177/2050640613501507
4. Njei B, Rajesh Konjeti V, Ditah I. Prognosis of patients with fibrolamellar hepatocellular carcinoma versus conventional hepatocellular carcinoma: A systematic review and meta-analysis. Gastrointestinal Cancer Reserch 2014;7:49-54. DOI: 10.1097/MCG.0000000000000070
5. Martínez Isla A, Ferrara A, Badía JM, et al. Hepatocarcinoma fibrolamelar. Resultados de la resección hepática parcial. Rev Esp Enferm Dig 1997;89:699-702.