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Primary acinar cell carcinoma of the liver showing good response to chemotherapy

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Abstract

A 48-year old man with no history of liver disease or alcohol addiction developed jaundice, itching and dark urine without any symptom of abdominal pain.

A CT examination revealed the presence of a liver mass extending between segments 4, 5 and 8 that was found to be an acinar cell carcinoma at biopsy. The primary hepatic origin was confirmed mainly excluding a primary pancreatic tumor by diagnostic work-up.

Since the lesion was considered unresectable at the time of the diagnosis, the patient underwent to Xeloda and oxaliplatin neoadjuvant chemotherapy, with progressive reduction in size of the mass as a sign of good response to therapy.

Keywords

acinar cell carcinoma; liver; pancreas; computed tomography; chemotherapy

A 48-year old man with no history of liver disease or alcohol addiction developed jaundice, itching and dark urine without any symptom of abdominal pain.

He presented to the emergency room in another institution, where he was worked up with ultrasound and computed tomography (CT) scan (images not available) revealing a large

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liver mass causing biliary duct dilatation, for which he had two internal external drains inserted. At that time, lab testing documented high bilirubin, alkaline phosphatase, AST and ALT levels.

Once at our institution, a dedicated multiphasic CT scan of the liver was obtained to further characterize the mass. CT images confirmed a 14×15 cm lesion extending between segments 4, 5 and 8 (Figure 1). No other lesions were detected within the liver. At this point, the radiological differential diagnosis included hepatocarcinoma, cholangiocarcinoma, fibrolamellar hepatocellular carcinoma, angiosarcoma, epitheloid hemangioma, hepatoblastoma or metastases [1].

The pathological H&E sections (Figure 2) obtained from the biopsy suggested the diagnosis of acinar cell carcinoma. Since the lesion was considered unresectable at the time of the diagnosis, the patient underwent to Xeloda and oxaliplatin neoadjuvant chemotherapy. Subsequent follow-up CT scans (Figure 3) showed progressive reduction in size of the mass as a sign of good response to therapy.

To note, very few papers described neoplasms with pancreatic acinar cell differentiation located in extra-pancreatic sites and just 6 cases of primary acinar cell carcinomas of the liver have been reported in the literature. Due to its extremely rare incidence, imaging features of hepatic acinar cell carcinoma are unknown and the diagnosis is made through pathology [1–3].

In our case, the primary hepatic origin was confirmed firstly excluding a primary pancreatic tumor by diagnostic workup (no metastatic disease or evidence of a pancreatic tumor at the diagnosis and after 13 months imaging follow-up). Secondly, the hepatic mass was single and not multiple as usually seen in metastatic pancreatic acinar cell carcinomas. Furthermore, metastatic pancreatic acinar cell carcinomas are usually large and about 50% have extra-hepatic metastases at the time of initial diagnosis [1].

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Figure 1. Axial (A) and coronal reformatted (B) CT images show the presence of a very heterogeneous hepatic mass, with cystic components, septa and foci of delayed enhancement. The margins of the lesion are poorly defined and there is no evidence of a perilesional capsule.

Figure 2.

Pathological H&E section showing a highly cellular carcinoma composed of irregular nests of cells separated by capillary sized vessels (A; original magnification, $\times 200$). The individual cells are uniform and polygonal, some with acinar configuration. The immunohistochemical stains (B; original magnification, $\times 200$) revealed positive labeling for trypsin.

Figure 3. Axial CT scans performed during (A) and after chemotherapy (B) show progressive reduction in size of the liver mass (respectively 8.7×8.1 and 7.2×7 cm), as a sign of good response to Xeloda and oxaliplatin neoadjuvant chemotherapy.