

Hepatocellular Cancer Arising From Ectopic Liver Tissue on Diaphragm in Association with Desmoid Mesenteric Tumor

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Abstract

Ectopic liver tissue is a rare clinical entity; very few cases of extra peritoneal localization have been described in Literature. Desmoid tumors are uncommon benign tumors occurring as a result of excessive proliferation of connective tissue. A 54 years old patient underwent ablation of a thoraco-abdominal mass arising from the left hemidiaphragm; preoperative alpha fetoprotein was high.

Histology: Hepatocellular carcinoma arising from peritoneal ectopic islet of liver tissue.

Preoperative CT and NMR incidentally diagnosed the presence of a mesenteric desmoid tumor that has been removed during operation. The case we have observed showed a coexistence of two very rare tumors; carcinogenesis on ectopic liver should be suspected in this patient with a thoraco-abdominal mass with high alpha fetoprotein levels, in absence of primitive liver chronic diseases and cancer.

Keywords: Ectopic liver; Cancer; Desmoid

Introduction

Ectopic liver tissue is an extremely rare clinical entity; some cases have been reported on peritoneum; extraperitoneal localization is extremely uncommon [1]; these ectopic islets can be affected by all liver diseases, even hepatocellular cancer [2].

Desmoid tumors are uncommon benign tumors occurring as a result of excessive proliferation of connective tissue; 30% of patients with desmoids tumors have the diagnosis of familial adenomatous polyposis, but they can also be seen sporadically [3].

We report the case of a patient who underwent surgical ablation of a thoraco - abdominal hepatocellular carcinoma of ectopic liver tissue involving the left hemidiaphragm; a jejunum mesentery desmoid tumor was also incidentally diagnosed and removed during operation.

Material and Methods

A 54 years old male patient, in good general conditions, with no history of chronic liver disease, was admitted to our Institution because of thoraco - abdominal pain (lower left chest, left hypochondrium), and fever.

No pathological findings have been detected at haematological tests and blood chemistry; hepatitis B and C viruses' antibodies were negative. CEA and CA 19-9 were in range, but alpha fetoprotein levels were high (880 IU / L).

Liver ultrasonography did not show signs of chronic liver disease, nor liver masses. CT scan and NMR showed a thoraco - abdominal mass developing from the left hemidiaphragm, with infiltration of the cranial part of the spleen (Figures 1 and 2); no primitive liver tumors have been detected. A jejunal mesentery mass (6 cm in diameter) was also incidentally diagnosed at abdominal CT scan (Figure 3).

Results

The patient underwent surgical radical ablation of the mass with laparotomic approach; the tumor has been removed "en bloc" with the spleen and the posterior part of the left hemidiaphragm (Figure 4); the diaphragm has been repaired with direct suture.

Histology: hepatocellular carcinoma arising from peritoneal ectopic islet of liver tissue (Figure 5), with infiltration of diaphragmatic muscle and spleen; resection margins were disease free. The mesenteric mass was also radically removed (histology: mesenteric desmoid tumor; resection margin without evidence of tumor involvement) (Figure 6).

Postoperative stay was uneventful and the patient has been discharged ten days after the surgical procedure.

The follow up has been organized with CT scan and alpha fetoprotein dosage every four months; the patient is currently alive and disease free, 3 years after surgical procedure; AFP level is persistently in range.

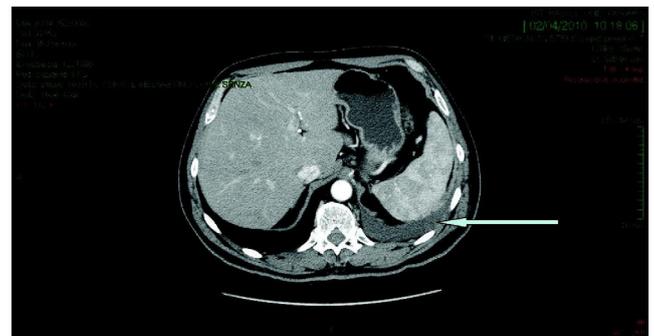


Figure 1: Thoraco - abdominal mass arising from left hemidiaphragm.

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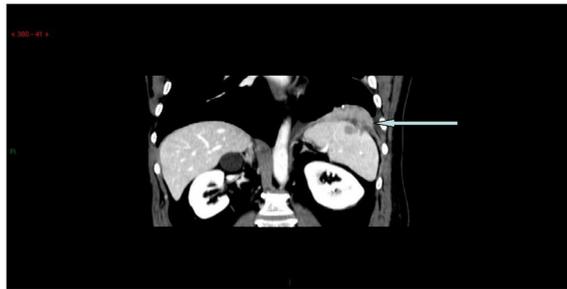


Figure 2: CT scan showing a thoraco - abdominal mass arising from left hemidiaphragm with infiltration of spleen.



Figure 3: CT scan showing a mesenteric mass



Figure 4: The tumor has been removed "en bloc" with the spleen and the posterior part of the left hemidiaphragm.

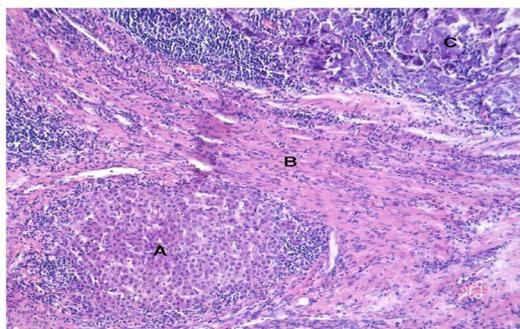


Figure 5: Histology of thoraco – abdominal mass
 A. Islet of ectopic liver tissue
 B. Muscular tissue (left hemidiaphragm)
 C. Hepatocellular Cancer

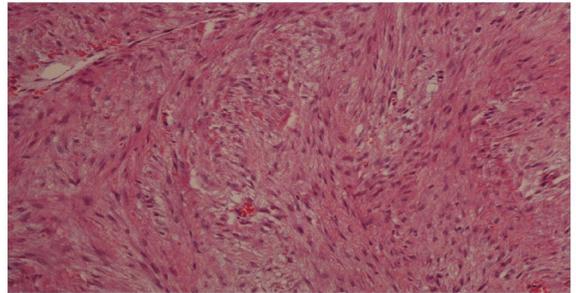


Figure 6: Mesenteric mass histology: desmoid tumor.

Discussion

Ectopic islets of liver tissue represent an unusual cause of abdominal mass; the most common localisation is the gallbladder wall, but Literature reports some cases located in adrenal glands, pancreas, spleen, liver ligaments and peritoneum [4].

Several embryological theories have been proposed in order to explain the genesis of ectopic liver tissue (accessory hepatic lobe with regression of the connection to the liver, migration of part of primitive liver to abnormal sites, budding of cranial parts of primitive liver before the closure of pleura-peritoneal channels) [5].

Ectopic liver tissue can present all liver diseases, such as steatosis, chronic hepatitis, cirrhosis and cancer [6].

The incomplete development of bile ducts leads to exposition to potentially carcinogenic agents [7].

Desmoid tumors, that are also called abdominal fibromatosis, are benign tumors which are not seen very often [8], with an estimated incidence of 2-4 per million people per year [9]; the etiology of desmoids tumors is genetic predisposition (in patients with FAP or Gardner syndrome), trauma, prior abdominal surgery and hormonal factors (endogenous levels of estrogens, pregnancy) [10].

Fibromatosis can be classified, on the basis of localization, as superficial (palmar, plantar, penile, juvenile aponeurotic fibroma, infantile digital fibromatosis) and deep (intra-abdominal, mesenteric, pelvic, abdominal, extra- abdominal, aggressive infantile, fibromatosis colli) [11].

Mesenteric desmoid tumors are often asymptomatic, but they can result in abdominal pain, palpable abdominal mass, fever, intestinal obstruction, perforation or ischemia [12]; although histologically benign, desmoid tumors are often locally invasive and associated with a high local recurrence rate after resection; many issues regarding optimal treatment of this disease remain controversial; surgical excision remains the treatment of choice; anyway, involvement of margins leads to an high risk of recurrence; for this reason some studies points out the need of considering other approaches, such as watchful waiting or surgery in association with other treatments (in particular radiotherapy) [13].

The case we have observed and treated shows an association between two very rare tumors in a patient without diagnosis of chronic liver disease, familial adenomatous polyposis or Gardner Syndrome; in this case, ectopic hepatocellular cancer is a very uncommon clinical condition that anyway should have been suspected in presence of a thoraco-abdominal mass with high levels of alpha fetoprotein.

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