Hepatocellular cancer arising from ectopic liver tissue on diaphragm

Case report

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Hepatocellular cancer arising from ectopic liver tissue on diaphragm. A Case report

AIM: Ectopic liver tissue is a rare clinical entity; very few cases of extra peritoneal localization have been described in Literature.

MATERIAL OF STUDY: A 54 years old male patient was admitted to our Institution because of thoraco - abdominal pain (lower left chest, left hypochondrium), and fever; CEA and CA 19-9 were in range, but alpha fetoprotein levels were high (880 IU / L); CT scan and NMR showed a thoraco – abdominal mass developing from the left hemidiaphragm, with infiltration of the cranial part of the spleen; no primitive liver tumors have been detected.

A jejunal mesentery mass (6 cm in diameter) was also incidentally diagnosed.

RESULTS: The patient underwent surgical resection of the thoraco – abdominal tumor and of the mesenteric mass.

Histology: hepatocellular carcinoma arising from ectopic islet of liver tissue on left hemidiaphragm; mesenteric desmoid tumor.

CONCLUSIONS: 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.

KEY WORDS: Desmoid, Hepatocellular cancer, Liver

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 jejunal mesentery desmoid tumor was also incidentally diagnosed and removed during operation.

Case Report

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 hematological 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 portion of the spleen; no primitive liver tumors have been detected. A jejunal mesentery mass (6 cm in diameter) was also incidentally diagnosed at abdominal CT scan (Fig. 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 (Fig. 4); the diaphragm has been repaired with direct suture. Histology: hepatocellular carcinoma arising from ectopic islet of liver tissue on left hemidiaphragm (Fig. 5), with infiltration of cranial portion of the spleen; resection margins were disease free. The mesenteric mass was also radically removed (histology: mesenteric desmoid tumor; resection margin without evidence of tumor involvement) (Fig. 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.

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

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

Fig. 3: CT scan showing a mesenteric mass.

Fig. 4: Surgical specimen thoraco-abdominal mass arising from left hemidiaphragm with spleen involvement.

Fig. 5: Mesenteric desmoid tumor (histology).

Fig. 6: CT scan showing a mesenteric mass.
Discussion and Comments

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. 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 pleuro – 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 desmoid 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 coli) 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.

Conclusions

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.

Riassunto

INTRODUZIONE: Le ectopie peritoneali di tessuto epatico sono entità cliniche di raro riscontro; in Letteratura sono stati riportati eccezionalmente casi di localizzazione extra – epatica.

CASE REPORT: Un paziente di 54 anni di età è giunto alla nostra osservazione a seguito dell’insorgenza di dolore a livello dell’emitorace e dell’ipocondrio sinistro; agli esami ematoclitici è stato riscontrato un innalzamento dell’alfa fetoproteina (880 UI/L).

Il paziente è stato sottoposto a TC e RMN, con evidenza di una massa toraco – addominale di pertinenza dell’emidiaframma sinistro; agli esami ematochimici è stato riscontrato un innalzamento dell’alfa fetoproteina (880 UI/L).

Le indagini diagnostiche hanno evidenziato incidentalmente una neoformazione solida di pertinenza del meso di un’ansa ileale di diametro di 6 cm.
RISULTATI: Il paziente è stato sottoposto ad intervento chirurgico di asportazione radicale della neoformazione toraco-addominale e della massa mesenterica.

Esame istologico: epatocarcinoma ad insorgenza da ectopia di tessuto epatico localizzato all’emidiaframma sinistro; tumore desmoide del mesentero.

CONCLUSIONI: Il caso che abbiamo osservato mostra la coesistenza di due neoplasie molto rare.

La degenerazione neoplastica di ectopie di tessuto epatico è di eccezionale riscontro, ma dovrebbe essere sospettata in questo paziente portatore di neoformazione toraco-addominale ed elevati livelli di alfafetoproteina.

References


