HEPATOCELLULAR CARCINOMA REVEALED BY A RIGHT ATRIAL TUMOR

CARCINOME HEPATO-CELLULAIRE REVELE PAR UNE TUMEUR AURICULAIRE DROITE

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Abstract

Hepatocellular metastatic carcinomas of the heart are uncommon malignant tumors that are usually located to the right atrium.
Herein, we present a 50-year-old cirrhotic Tunisian gentleman presented to our department with jaundice, bilateral lower leg edema, continuous cough and dyspnea of one month's duration. His medical history included hepatitis B marker positive. He denied any known cardiac or pulmonary disease.
Echocardiography revealed right atrial obstruction caused by a huge right atrial mass. Abdominal MRI and Computed tomography demonstrated the typical appearance of hepatocellular carcinoma, with associated thrombus, extending from the right lobe of the liver into the hepatic vein and inferior vena cava and into the right atrium. Tumor was surgically excised under cardiopulmonary bypass. One month later a recurrence of the atrial mass was observed. So, when a patient with a history of chronic hepatic disease presents with symptoms of right heart failure one must be cautious and should bear in mind that right heart involvement from a malignant tumor may be present.

Key words: Hepatocellular carcinoma; Cardiac metastatic; Heart failure; Thrombus

Résumé

Les métastases cardiaques des carcinomes hépato-cellulaires sont des tumeurs malignes rares qui sont d'habitude localisées au niveau des oreillettes.
Nous rapportons ainsi le cas d'un homme cirrhotique de 50 ans qui a consulté pour un ictère, des œdèmes des membres inférieurs, une toux continue et une dyspnée évoluant depuis un mois. Il était porteur de l'hépatite B mais il n'avait aucun antécédent cardiaque ou pulmonaire.
L'échocardiographie a révélé une obstruction de l'oreillette droite à cause d'une volumineuse masse auriculaire. L'imagerie abdominale a objectivé l'existence d'un carcinome hépatocellulaire, avec thrombus associé, s'étendant du lobe droit du foie vers la veine hépatique à la veine cave inférieure jusqu'à l'oreillette droite. La tumeur atriale a été chirurgicalement réséquée sous circulation extracorporelle. Un mois plus tard une récidive de la masse atriale a été observée.
Il faut évoquer une métastase cardiaque d'une tumeur maligne hépatique devant toute hépatopathie chronique avec insuffisance cardiaque droite.

Mots clés : Carcinome hépato-cellulaire ; Métastase cardiaque ; Insuffisance cardiaque ; Thrombus

ملخص

الانبثاث من سرطان الخلايا الكبدية على مستوى القلب هي من الأورام الخبيثة النادرة و التي توجد عادة في الأذينين.
قمنا بوصف حالة رجل عمره 50 سنة يعاني من التليف الكبدى و تم فحصه لوجود يرقان و وذمة في الأطراف السفلية و سعال مستمر وضيق تنفس من مدة شهر. كان هذا الرجل حاملاً لالتهاب الكبد (ب) ولكن لم يكن لديه سوابق في أمراض القلب أو الرئتين.كشف تخطيط صدى القلب انسداد الأذين الأيمن بسبب كتلة ضخمة. وقد أثبت التصوير الطبي للبطن وجود سرطان الكبد و حصول جلطة في شكل صمام متخثر في نفس الوقت ويمتد من الفص الكبدي الأيمن إلى الوريد الكبدي و من الوريد الأجوف السفلي إلى الأذين الأيمن. تم استئصال الورم جراحيًا تحت الدورة الدموية خارج الجسم. بعد شهر واحد لوحظ تكرار كتلة الأذين.
فمن الضروري استحضار حصول انبثاث لورم خبيث في القلب صادر عن ورم خبيث في الكبد أمام أي مرض مزمن في الكبد مع قصور القلب الأيمن.

الكلمات المفاتيح: سرطان خلايا الكبد ; انبثاث القلب ; قصور القلب ; خثرة الدم.

BACKGROUND

Hepatocellular metastatic carcinomas to the heart are uncommon malignant tumors that are usually located to the right atrium. Prompt diagnosis of their presence is of major clinical importance because although rare they can cause obstructive phenomena, heart failure and even sudden cardiac death [1-2-3]. Herein, we present a patient with a metastatic hepatocellular carcinoma located in the right atrium and invading the right ventricle, the pre-operative workout and the subsequent management.

CASE PRESENTATION

A 54year-old cirrhotic Tunisian gentleman presented to the cardiology emergency department with jaundice, bilateral lower leg edema, continuous cough and exertional dyspnea of one month's duration. His medical history included hepatitis B marker positive. He denied any known cardiac or pulmonary disease. He had neither a central venous catheter nor other obvious prothrombotic cause for atrial thrombus formation. His blood pressure was 110/70 mmHg, heart rate 100 beats/ minute, body temperature 38°C. Although pulse oximetry showed 96% oxygen saturation on room air.

On physical examination, the patient had acute ill looking, dyspnea, icterus sclera, ascites and a distended abdomen with engorged superficial veins was noted. A gallop rhythm and grade III/VI holosystolic murmur were detected on auscultation.

Significant biochemistry results were as follows: aspartate aminotransferase (AST) 89 IU/L, alanine transaminase (ALT) 67 IU/L, total bilirubin 29 mg/dL and albumin 24g/L. Coagulation profile was within normal limit. He had a normal serum level of alpha-fetoprotein 5.3 ng/mL (Normal reference upper limit 10 ng/mL).

A 12-lead electrocardiogram and cardiac troponin I test was normal. Urgent cardiac ultrasound revealed a giant mass that partially occupied the right atrium with a diameter of 20 millimeters. (Figure 1). Urgent chest computed tomography (CT) showed a tumor thrombus extending through the right hepatic vein and inferior vena cava (IVC) into the right atrium (Figure 2). There is no mass in liver. The patient had no pulmonary embolization

The patient underwent urgent surgical treatment due to worsening of his clinical condition, hemodynamic instability and risk of sudden death: the tumor was surgically excised under cardiopulmonary bypass. Therefore, initially femoro-femoral cannulation was installed (Figure 3). The specimen was histopathologically investigated and eventually diagnosed as a metastatic___poorly differentiated hepatocellular carcinoma (HCC).

Recovery was uneventful. One month later a recurrence of the atrial mass was observed.

Additionally, abdominal MRI and Computed tomography demonstrated the typical appearance of hepatocellular carcinoma, with associated thrombus, extending from the right lobe of the liver (one tumor, 2.5 cm in diameter, in segments 6), and into the hepatic vein and inferior vena cava and into the right atrium. No further adjuvant therapy was considered necessary in this stage by our consulting oncologists. He was discharged home for palliative care. When last seen at his follow-up consultation 4 months after discharge, the patient was well without any signs of right heart failure. But 12 months later, the patient died with respiratory and circulatory failure induced by hepatic failure.

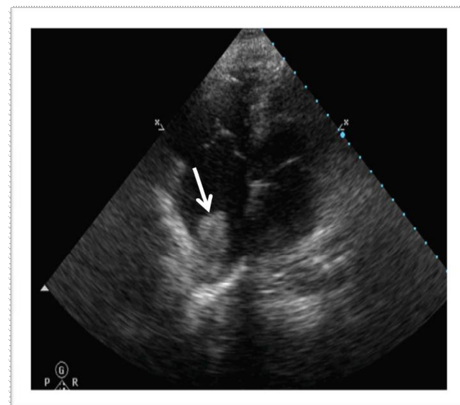

Figure 1: Echocardiography, apical view showing a giant mass in the right atrium (arrow).

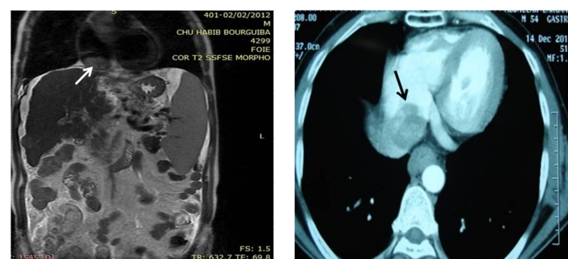

Figure 2 : Chest computed tomography (CT) and abdominal MRI showed a tumor in the right hepatic vein and inferior vena cava into the right atrium.

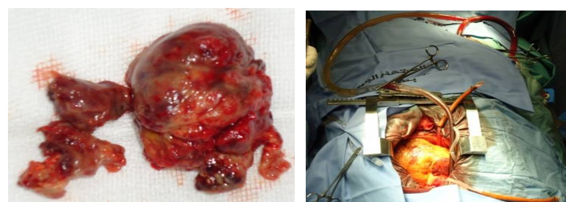

Figure 3: Surgical treatment: the tumor was surgically excised under cardiopulmonary bypass.

DISCUSSION

Primary liver cancer is the fifth most common neoplasm. Predisposing factors for orthotopic primary Hepatocellular Carcinoma (HCC) generations are chronic hepatitis B or C, infection and cirrhosis secondary to other chronic liver disease. Worldwide, most patients with HCC have underlying cirrhosis, and it is uncommon to find HCC in patients without cirrhosis [1]. Although HCC has a very aggressive metastatic profile, its tendency to spread towards the heart is unusual but well documented through several published case reports which define an incidence of cardiac metastasis at 0.67-3% [2, 3].

However very few cases of giant metastatic HCC within the right cardiac cavities that cause significant occlusion of the tricuspid valve are described in the current literature.

Therefore, an interesting feature of HCC can be its varied and sometimes bizarre presentation [4]. This report describes an unusual presentation of HCC. The patient appeared with symptoms of acute heart failure caused by a giant right Metastatic disease as the initial presentation of HCC appears in less than 5% of cases [5]. In addition, histological investigation defines whether the mass derives from an occult HCC or is presented as an ectopic one with no liver involvement.

Regarding the symptoms, there is a variety of clinical manifestations caused by the atrial neoplasm and those are mainly tumor-size dependent. Patients may have no symptoms, dyspnea due to pulmonary embolism, syncope, or heart failure. Physical findings include edema, pan systolic murmur with diastolic rumble over the tricuspid valve, and improvement of symptoms with left lateral decubitus position [6].

Extracardiac tumors involving inferior vena cava and right atrium include renal cell tumor (4-10%) [7], thyroid carcinoma, testicular tumors and HCC. In most cases of advanced HCC, the extent of the disease is verified with presence of metastasis at the lungs, peritoneum, adrenal glands and bones. Generally, HCC appears to tend to invade vascular structures [8]. A right atrial intracavitary mass is then formatted which causes significant hemodynamic instability. In addition, left atrium, right ventricle, and intramyocardial involvement of the left ventricle have also been reported as rare sites of HCC metastasis as well as spreading of the cancer to the left chambers through pulmonary metastasis or patent foramen oval [9]. Regarding the case described here, the appearance of a metastatic HCC tumor inside the right atrium as the only manifestation and without apparent primary focus is unique. It is important to know the tumor location before bicaval cannulation to prevent fragmentation and embolization of the tumor. The major causes of death are either sudden pulmonary embolism of the thrombus or acute obstruction of the tricuspid valve or both. Resection can provide relatively good mid-term survival regarding this clinical situation but not more than 2 years [10]. A few reports describe the successful removal of HCC from the right atrium without extracorporeal circulation as an alternative [11]. However, both curative resection treatments have a dismal prognosis, with 5 years reported survival around 12-39% [3].

After resection of an HCC, tumor recurrence exceeds up to 70% at 5 years, including recurrence due to dissemination and de novo tumors of the liver. The most important statistically predictor of recurrence seems to be the presence of micro vascular invasion and/or additional tumor sites besides the primary lesion [11]. There is no effective adjuvant therapy that can reduce the recurrence rates. (Recommendation level II) [11, 12] Internal radiation and adoptive immunotherapy by activated lymphocytes may have some anti-tumor efficacy but the early results have not been statistically powered yet [13]. There are no adequate published data to indicate proper treatment of recurrences. Solitary recurrent masses might benefit from repeat resection but in most of the cases recurrence appears to be multifocal and so further treatment is impossible [13].

CONCLUSION

In conclusion, when a patient with a history of chronic hepatic disease presents with symptoms of right heart failure one must be cautious and should bear in mind that right heart involvement from a malignant tumor may be present. Echocardiography computed tomography and magnetic resonance imaging are the standard

imaging modalities to determine the nature of tumors presented as secondary cardiac neoplasms. Urgent Computed tomography can easily and quickly be performed prior to surgical treatment of emergency cases.

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