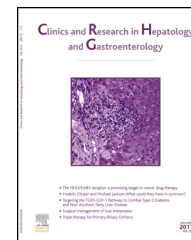




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CLINICAL CHALLENGE

Primary hepatic angiosarcoma and liver transplantation: Radiological, surgical, histological findings and clinical outcome

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KEYWORDS

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Summary Angiosarcoma is a rare type of soft tissue sarcoma that accounts for less than 1% of all sarcomas and only 2% of all primary hepatic tumours. Thorotrast, arsenic, and vinyl chloride monomer are frequently listed as occupational exposure risks. The estimated latency is long (10–40 years) in occupational cases and very long (60 years or more) in non-occupational cases. The symptoms and CT-scan appearance of hepatic angiosarcoma (HAS) are non-specific. We present a case of a 65-year-old Caucasian male with history of cryptogenic cirrhosis, low alpha-foetoprotein levels and a single, 4-cm nodule of potential atypical hepatocellular carcinoma (no washout at MRI and CT-scan) in segment VIII. Laparoscopic radiofrequency ablation (a biopsy of the neoplastic lesion was technically impossible) was performed, followed by liver transplantation (LT) 6 months later. High-grade multifocal HAS was found in the explanted liver, with extensive involvement of the venous portal structures. No complications were observed during the postoperative course, and initial immunosuppression included tacrolimus, mycophenolate mofetil and corticosteroids. Because of the histological findings, tacrolimus was switched to everolimus as the main immunosuppressive drug one month after LT. Despite this conversion, the patient developed bone metastases 3 months after LT and peritoneal carcinosis one month later. This case report suggests that everolimus conversion does not inhibit the development of

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tumour metastases. Consequently, HAS remains an absolute contraindication to LT because of the poor outcome. If LT has been performed for incidental HAS, new molecular therapies (e.g. vascular endothelial growth factor antagonists) should be considered immediately after LT to improve the outcome.

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Introduction

Angiosarcoma is a rare tumour that account for less than 1% of all sarcomas and only 2% of all primary liver tumours [1,2]. Angiosarcoma may derive from the endothelia of lymphatic or blood vessels and is a rarely encountered type of soft tissue sarcoma. Thorotrast, arsenic and vinyl chloride monomer, the use of thorotrast in angiography and androgenic anabolic steroids are frequently listed as risks [2,3]. Exposure to vinyl chloride is linked to a series of mechanistic events that lead to a carcinogenic outcome; mutations of K-ras and p53 are also detected, but their aetiology remains unknown [2,4] Hepatic angiosarcoma (HAS) is three times more common in males than in females [2]. The estimated latency of this rare cancer is long, at around 10–40 years, in occupational cases and longer, at 60 years or more, in non-occupational cases [5,6]. An accurate diagnosis of this tumour is difficult, particularly if the patient has no history of exposure to specific carcinogens. At presentation, the symptoms, signs and liver function test findings are not specific [7]. Targeted therapies such as bevacizumab and sorafenib have been shown to have limited efficacy [5]. The prognosis is poor and liver transplantation (LT) is not recommended because of universal tumour recurrence [2]. The aims of this publication were to describe the clinical outcome of transplantation for HAS and to also to make a brief review of the current literature.

Case report

A 65-year-old man without any risk factors for liver disease was referred to our centre in August 2013 for asthenia without weight loss. No chronic drug use or excessive alcohol consumption were reported (occasionally 20 g/day, stopped in July 2013). He worked as an engineer on an oil tanker.

A histological diagnosis was made of cryptogenetic cirrhosis (Child–Pugh B7 and Model for End-stage Liver Disease [MELD] 10) with grade II oesophageal varices, and primary prophylaxis with beta-blockers was initiated. No significant co-morbidities were reported. Clinical examination revealed normal vital signs, a weight of 79 kg with a BMI of 24.9 kg/m², hepatomegaly, moderate ascites and no peripheral oedema. Laboratory tests showed 7500 WBC/mm³, Hb 15.2 g/dL, platelets at 100,000/mm³, AST 65 U/L (17–27), ALT 52 U/L (11–26), γ GT 46 U/L (8–36), total bilirubin 19 mmol/L (2–17), creatinine 81 mmol/L (44–80), albumin 35 g/L, TP 43% and Alpha-fetoprotein (AFP) 4.01 ng/mL, with serological markers of HBV, HCV

and HIV and autoimmunity tests all negative. A Doppler ultrasound of the liver showed signs of portal hypertension without hepatocellular carcinoma (HCC).

In October 2014, the patient was diagnosed with a single nodule of atypical HCC (no wash out on magnetic resonance imaging [MRI] and computed tomography [CT] scans of the liver), measuring 4 cm in segment VIII, and low AFP levels (Fig. 1). Laparoscopic radiofrequency ablation was performed, but the biopsy initially planned of the neoplastic lesion was technically impossible (Fig. 2). Six months later, an alteration of liver function was reported (CPT 12, MELD score 19) and LT was performed. No surgical complications, but early acute renal failure post-LT, were reported. The patient received prednisone (20 mg/day), mycophenolate mofetil (1 g bid) and tacrolimus (2.5 mg bid, with residual concentrations ranging from 8–10 μ g/L) as immunosuppressive therapy. Histological analysis of the explanted liver revealed multifocal angiosarcoma (larger than 6 cm), with microvascular invasion (Fig. 3) and positivity for the immunohistochemical markers CD31, CD34 and ERG (Fig. 4). One month after LT, a conversion was made to an mTOR inhibitor: tacrolimus was gradually withdrawn and everolimus (3 mg bid, with residual concentrations ranging from 8–10 μ g/L) was initiated. A meeting of the multidisciplinary team recommended follow-up by means of a thoracic CT and abdominal MRI every 3 months. Three months after LT, the patient reported spinal pain, anaemia and ascites, and was re-admitted in our Transplant Unit for deterioration in his overall condition and ascites. A PET-scan (positron emission tomography) and spinal MRI revealed bone lesions compatible with bone metastases. The diagnosis of peritoneal carcinomatosis was made from the findings of an abdominal CT-scan (no neoplastic cells were found in ascites fluid). Chemotherapy was initiated with Paclitaxel (80 mg/day for 7 days) but two weeks later the patient died from a lung infection.

Discussion

We report here on an unusual clinical case that illustrates the difficulty of diagnosing an atypical HCC versus HAS. The clinical features of HAS are non-specific and may mimic chronic or acute liver disease. Symptoms include fatigue, anorexia, weight loss, fever and abdominal pain [2,8,9]. Physical findings include ascites, hepatomegaly, jaundice and acute abdominal bleeding [5,8]. Abnormal liver function markers (bilirubin levels, altered protein-albumin/globulin ratio, liver enzymes) may appear in large tumours or

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