A retroperitoneal mass with elevated alpha-1-fetoprotein: not always a testicular carcinoma

F. Toonen¹, T. Smilde²

¹Department of Internal Medicine, Radboud University Nijmegen Medical Centre, Nijmegen, the Netherlands, ²Department of Medical Oncology, Jeroen Bosch Hospital, Den Bosch, the Netherlands, *corresponding author: f.toonen@aig.umcn.nl

ABSTRACT

High levels of alpha-1-fetoprotein are usually associated with nonseminoma carcinoma of the testis or hepatocellular carcinoma of the liver. We describe a male patient with extrahepatic hepatocellular carcinoma who presented with a large retroperitoneal mass and extremely high alpha-1-fetoprotein levels. The importance of taking an adequate biopsy specimen cannot be emphasised enough since both prognosis and treatment are completely different.

KEYWORDS

Alpha-1-fetoprotein, ectopic tissue, hepatocellular carcinoma

INTRODUCTION

Hepatocellular carcinoma (HCC) is known for its high mortality. It also demonstrates a high variability in incidence around the world. Areas such as sub-Saharan Africa and Eastern Asia have an incidence up to ten times higher than the incidence in, for example, Europe.¹ This is probably due to the local prevalence of risk factors as hepatitis B and C virus. A known yet very rare phenomenon is hepatocellular carcinoma in ectopic liver tissue. The incidence of ectopic liver tissue is estimated at 0.1 to 0.5%.² Described locations include the gallbladder, pancreas, diaphragm, thorax and testis. Virtually all cases of ectopic HCC involve the Asian population,³ although three Caucasian patients have been described.⁴ We present a Caucasian male with no relevant medical history and no risk factors for HCC with a retroperitoneal HCC without lesions in his liver.

CASE

A 46-year-old Dutch male visited our emergency department after referral by his general practitioner with symptoms of stomach ache in his umbilical region that had been progressive over the last three months. Over that period his bodyweight had dropped about 5 kg. He complained of some diarrhoea without blood loss. His medical history comprised a surgical correction of a pyloric stenosis in his first year. He was not on any medication, did not smoke and used alcohol in moderate quantities.

On physical examination, a pale man was seen, in no acute distress with normal vital signs. There were no further abnormalities, aside from some peri-umbilical tenderness on palpation. There was no palpable mass in his testicles. Biochemical investigation showed a microcytic anaemia (Hb 4.2 mmol/l; MCV 65 fl) and moderately elevated aspartate aminotransferase and alanine aminotransferase (76 U/l and 147 U/l, respectively). Ultrasonography of the abdomen revealed a soft-tissue mass in the upper abdomen. A computed tomography (CT) scan of the abdomen demonstrated a normal liver, but a large retroperitoneal mass in the upper abdomen (figure 1) with multiple enlarged lymph nodes in the mesentery. Magnetic resonance imaging did not show any hepatic lesions either. Because a testicular carcinoma was considered a possible diagnosis, an alpha-1-fetoprotein (α¹-FP) level was determined. This was extremely elevated at 24,000 kU/l (reference value <7 kU/l). The ultrasound investigation of his testes, however, was normal.

For further investigation the patient underwent an ultrasound-guided biopsy of the tumour. Microscopic examination revealed a tumour growing in trabeculae and some tubular structures with an aspect of hepatocytes. Some cells showed a positive alpha-1-fetoprotein (α¹-FP) immunostain. No bile production was seen. Further staining showed pankeratin
in the tissue sample. The phenomenon of ectopic liver tissue developing into carcinoma remains a diagnostic challenge. Learning more about the mechanisms leading to this higher carcinogenicity might lead to a better understanding of the principles behind the origin of hepatocellular carcinoma. Until recently no proven or standard systemic treatment for advanced HCC was available. The SHARP trial, a randomised phase III trial in HCC, performed in the Western population, compared sorafenib 400 mg twice daily with placebo. Sorafenib is an oral multikinase inhibitor with antiproliferative and antiangiogenic effects. Treatment with sorafenib showed a progression-free survival (PFS) benefit of 2.4 months compared with placebo (PFS 5.2 vs 2.8 months in sorafenib and placebo respectively). The overall survival (OS) was 10.7 months vs 7.9 months in sorafenib and placebo, respectively. Cheng et al. also showed a survival and PFS benefit in sorafenib vs placebo in 271 patients from 23 centres in China, South Korea and Taiwan (PFS 2.8 vs 1.4 and OS 6.5 vs 4.2 months in sorafenib and placebo, respectively). The effect of systemic treatment of extrahepatic HCC is unknown. Sorafenib is generally well tolerated although side effects are sometimes severe and dose limiting. The most common drug-related adverse events include hand-foot syndrome, diarrhoea, alopecia, fatigue, rash or desquamation and hypertension.

CONCLUSION

A retroperitoneal mass with elevated levels of α-1-FP and no liver lesions does not always mean a testicular carcinoma. Even in otherwise healthy Caucasian patients, ectopic hepatocellular carcinoma is a possibility, although a rare one. The importance of having a good enough biopsy specimen of the tumour for determining its origin cannot be emphasised enough.

REFERENCES
