Primary yolk sac tumour of the liver in adulthood

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Abstract

Primary yolk sac tumour of the liver is exceedingly rare. A 28 year old woman presented with a cystic liver mass and a markedly raised serum α-fetoprotein concentration. She underwent a partial hepatectomy for a suspected hepatocellular carcinoma but histological examination of the tumour revealed the classical morphological and immunohistochemical features of a yolk sac tumour. There was no evidence of an extrahepatic primary source. Review of this case, together with the six previously reported adult cases of primary yolk sac tumours of the liver, revealed several features of the tumour that may aid differentiation from hepatocellular carcinoma, with potential therapeutic implications.

(*J Clin Pathol* 1998;51:939–940)

Keywords: endodermal sinus tumour; liver neoplasms; α-fetoprotein

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Accepted for publication 23 June 1998

Yolk sac (or endodermal sinus) tumours usually arise from the ovary or testis, though there have been reports of such tumours arising in extragonadal sites such as the mediastinum, bladder, and pineal gland.[1] Primary yolk sac tumour of the liver was first reported by Hart in 1975.[2] Since then a further 12 cases have been reported, of which six presented in adulthood.[1 3–7] Study of the present case and review of these six previous cases has revealed several characteristic features of adult primary yolk sac tumours of the liver which have not previously been commented on. These features may aid in the clinical distinction of the tumour from hepatocellular carcinoma, with potential therapeutic implications.

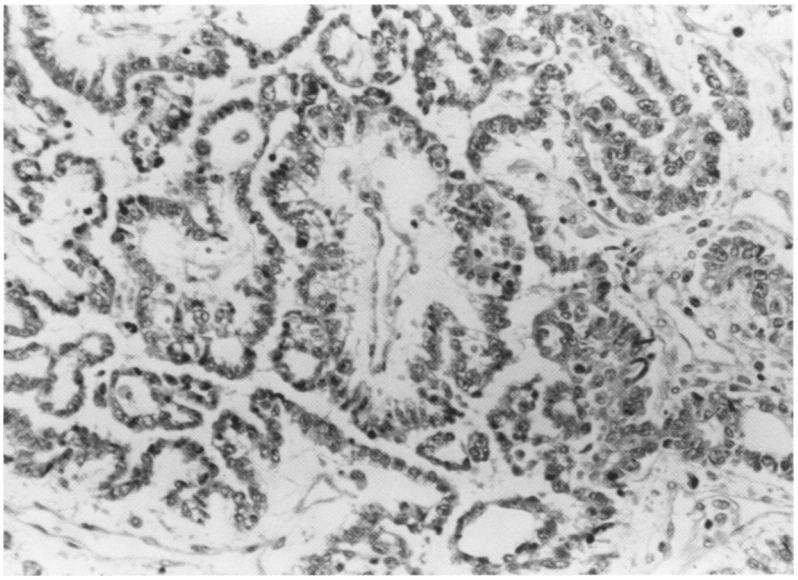

Figure 1 High power histological appearance of the tumour (haematoxylin and eosin, ×400 objective). A Schiller–Duval body is arrowed.

Case report

A 28 year old white female presented to her general practitioner with a month's history of increasing abdominal girth and, more recently, self recognised hepatomegaly. Her only regular medication was the combined oral contraceptive pill, and her weekly alcohol intake was 6 units. Otherwise, she had no risk factors for liver disease. Clinical examination revealed a three fingers breadth, smooth liver edge only. Her serum liver function indices and human chorionic gonadotrophin (hCG) concentration were normal, but her serum α-fetoprotein concentration was grossly raised at 14 614 kU/litre (normal < 10 kU/litre). Computerised tomography showed a large cystic mass in the right lobe of the liver. The cyst wall had a solid nodular component and there was possible invasion into the right hemidiaphragm. No secondary liver or intraperitoneal tumour nodules was seen, nor was any intra-abdominal lymphadenopathy. Hepatocellular carcinoma was considered the most likely diagnosis, and a right hepatectomy performed.

The right lobe of liver had a smooth intact capsule with an attached disc of diaphragm. Sectioning of the lobe revealed a solitary, cystic mass (15 × 15 × 6 cm) with areas of haemorrhage and necrosis set among solid peripheral nodules of friable white tissue. The tumour cells showed pleomorphic nuclei with numerous mitoses, and a mixed reticular and solid architecture with several Schiller–Duval bodies (fig 1). Periodic acid Schiff positive globules were present within and around the tumour cells, which also showed focal immunoreactivity for α-1-antitrypsin, α-fetoprotein, and placental alkaline phosphatase. Neither immunoreactivity for hCG, a germ cell component, nor features of HCC or hepatoblastoma were seen. Despite the presence of infiltration into the attached diaphragm, excision of the tumour was complete. The background liver was normal. Postoperative clinical and radiological assessment, including transvaginal ultrasonography, showed no evidence of an ovarian or other extrahepatic tumour. At the time of writing (one year postoperatively), the patient remains well, with a normal serum α-fetoprotein concentration and no sign of recurrence.

Discussion

The presentation of a young adult with a malignant, cystic tumour of the liver is unusual. The raised serum α-fetoprotein concentration, in the absence of parenchymal hepatic disease, suggested a preoperative differential diagnosis

Table 1 Reports of adult cases of primary yolk sac tumour of the liver

Author (year)	*Sex*	*Age (years)*	*Ethnic group*	*α-FP (kU/l)*	*Size (cm)*	*Location in liver*	*Central necrosis/ cystic change¶*
Yan (1982)[3]	M	58	Oriental	330 000	Multiple nodules†	Disseminated	‡
Narita (1982)[5]	F	27	Oriental	23 489	11×11×9.5	Right lobe	Yes
Natori (1983)[4]	F	29	Oriental	3523	(1) 17×15×10 (2) 20×25	Right lobe	Yes
Villaschi (1991)[6]	F	28	European	413	15D	Left lobe	Yes
Whelan (1991)[1]	F	27	Afro-Caribbean	89 000	15D	Right lobe	Yes
Higuchi (1993)[7]	F	24	Oriental	115 500	"Large tumour"	Right lobe	Yes
Wong (1998)*	F	28	European	14 614	15×15×6	Right lobe	Yes

*This study.
†Size not specified.
‡Not commented on.
¶Central necrotic/cystic change detected by preoperative computed tomography.
α-FP, serum α-fetoprotein concentration on presentation.

of hepatocellular carcinoma, hepatoblastoma, or metastatic yolk sac malignancy. The latter was unlikely in view of the solitary nature of the liver tumour and the absence of any extrahepatic tumour. Regarding hepatoblastoma, this primitive tumour only rarely occurs in adults and usually presents as a solid mass.[8]

While this is only the seventh reported adult case of pure primary yolk sac tumour of the liver, there are several emerging correlates that may aid clinical distinction from hepatocellular carcinoma (table 1). Of the seven cases so far reported, six involved women aged between 24 and 30 years,[1 3-7] whereas hepatocellular carcinomas show a male preponderance and usually present after the age of 50 years in Western countries.[8 9] It is uncommon for a hepatocellular carcinoma to present as a predominantly cystic tumour within a non-cirrhotic liver, as was seen in six of the seven cases. Finally, six of the seven cases showed a serum α-fetoprotein concentration of more than 3000 kU/litre, compared with only a minority of hepatocellular carcinomas.[9]

Distinguishing preoperatively between primary yolk sac tumours of the liver and hepatocellular carcinoma has important therapeutic implications. Hepatocellular carcinomas respond poorly to chemotherapy.[9] While earlier reports of primary yolk sac tumours of the liver suggested a uniformly poor prognosis,[5] Whelan and colleagues have more recently reported complete cure through chemotherapy followed by surgical resection.[1] The diagnosis of primary yolk sac tumours of the liver was made preoperatively by computed tomography guided percutaneous liver biopsy, and following treatment with a combination of cisplatin, etoposide, and bleomycin, the resected lobe of liver showed no evidence of viable tumour and the patient remained disease-free five years after the operation.[1] Our patient remains well with no evidence of tumour recurrence one year after surgery. However, as the long term outcome of surgically treated primary yolk sac tumours of the liver has yet to be established, our patient will continue to receive close clinical follow up.

In conclusion, primary yolk sac tumour of the liver, albeit rare, should always be considered as an alternative diagnosis to hepatocellular carcinoma in a young patient with grossly elevated serum α-fetoprotein concentrations and a cystic tumour within a non-cirrhotic liver.

We thank Drs R L Jones and C P Case and Mr C P Barham for their contributions towards the assessment of this case.

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