

Angiosarcoma, a Rare Neoplasm of the Liver: a Case Report and Review of Literature

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Citation

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Abstract

We describe the case of a 44-year-old male with a rare malignant neoplasm of the liver: an angiosarcoma. This case is interesting because there was no history of exposure to the known carcinogens for this malignancy, and no predisposing chronic medical condition. We believe that this was a case of primary hereditary angiosarcoma, which has been rarely reported in the literature.

CASE REPORT

A 44-year-old man, a chronic alcoholic with no significant medical history, was admitted with complaints of pain in the right upper quadrant of the abdomen, nausea and loss of appetite and weight for 4 months.

There was no abnormal finding on physical examination. Liver biochemistry and routine blood investigations were normal. Viral markers were normal. USG revealed evidence of chronic liver disease with a mass suggestive of hepatocellular carcinoma in the right lobe of the liver.

CT scan showed a 9 x 5.9 x 8.1cm heterogeneously enhancing mass lesion involving the right lobe of the liver (Fig. 1).

Figure 1

Figure 1: CT scan showing a heterogeneously enhancing mass lesion involving the right lobe of the liver



Tumour markers AFP and CEA were within normal range.

Right hepatectomy was performed and the post-operative period was uneventful (Fig. 2)

Figure 2

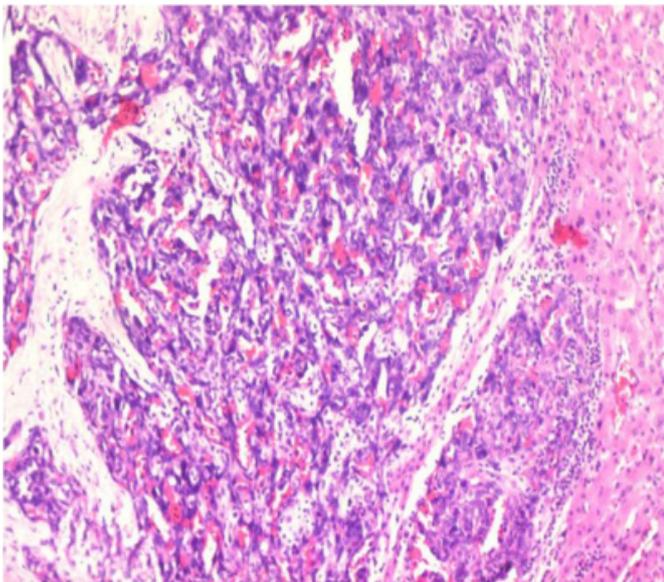
Figure 2: Resected gross specimen of the right lobe of the liver



Histopathology showed a poorly differentiated angiosarcoma with cavernoma formation (Fig. 3).

Figure 3

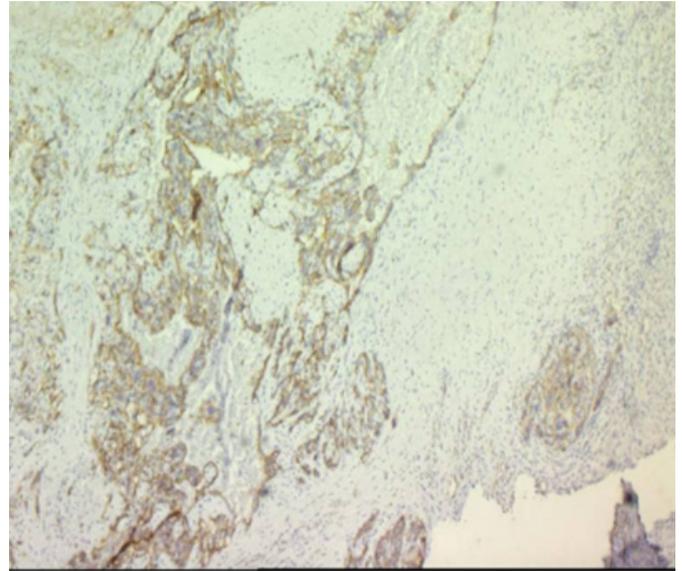
Figure 3: Histomicrograph of poorly differentiated angiosarcoma with cavernoma formation



Immunohistochemistry profile: CD 31, CD 34, factor 8 positive, confirmatory for angiosarcoma (Fig 4).

Figure 4

Figure 4: Immunohistochemistry profile confirmatory for angiosarcoma



Retrospectively there was no history of exposure to any of the known carcinogens for angiosarcoma.

On follow-up at 1 month and 3 months postoperatively the patient was asymptomatic. CT scan showed no evidence of focal lesion in the residual liver.

DISCUSSION

Angiosarcoma is responsible for about 2% of primary liver tumors, and is considered to be the most common of the mesenchymal liver tumors. About 25 cases occur each year in the United States. Males (ratio 3:1) in their fifth or sixth decade are most often affected, but it can also occur in children.³⁴ It often presents with abdominal discomfort and distension, weight loss, and fatigue. On examination, the patient may have jaundice, hepatomegaly, and ascites.⁵ Liver function tests are usually abnormal. Thrombocytopenia, microangiopathic haemolytic anaemia, and disseminated intravascular coagulation may also be present.⁶ There are no tumour markers.

CT images classically show multiple hypodense areas. After administration of contrast medium, the lesions become partly or completely isodense with the normal hepatic tissue.⁷ On T2-weighted MRI imaging, there are areas of high signals with central regions of low signals. Liver biopsy is hazardous as it may cause severe haemorrhage.⁸ Macroscopically, 'blood lakes' may be seen as the tumor is angioinvasive. On histological examination, there are typical spindle-shaped hyperchromatic cells with nucleoli, which,

on immunostaining, are positive for endothelial cells markers.⁹

More than 30% of cases have been linked to exposure to environmental agents including Thorotrast® which was used as a radiological contrast agent in the past. Evidence of previous exposure is seen on a CT scan, and the latency period can be longer than 30 years.¹⁰¹¹

Angiosarcoma is also associated with exposure to vinyl chloride, which is used in rubber and plastic processing¹²¹³. Again the latency period can be very long. The risk ratio is 400:1 compared to the general population,¹⁴ and may be due to increased frequency of p53 mutations in these people.¹⁵ The incidence in this group has decreased since the acceptable exposure level to vinyl chloride has been reduced.

Angiosarcoma has also been reported to be associated with exposure to arsenic¹⁶, anabolic steroids¹⁷, and oral contraceptives.¹⁸ It has been reported in a previous haemangioma,¹⁹ with Von Recklinghausen's neurofibromatosis²⁰, and in patients with certain hereditary conditions such as congenital hereditary lymphoedema (Milroy's disease)²¹ and Von Hippel Lindau disease.²²

Detailed history from our patient failed to reveal previous exposure to any of the known carcinogens or predisposing medical conditions. Our case is one of the few reports of primary angiosarcoma reported in literature.

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