CASE REPORT

Angiosarcoma of the Liver

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Summary

A 47-year-old Malay man presented with unremitting epigastric pain and loss of weight for 3 months. At laparotomy, a large tumour mass was found in the liver and a few small nodules in the spleen.

Histopathological examination revealed a hepatic angiosarcoma with metastasis to the spleen. He had exposure to formic acid fumes for more than 23 years in his work as a rubber tapper. The various chemical carcinogens that are implicated in this type of tumour are discussed. The possiblity of an association between formic acid fumes and development of angiosarcoma in the liver is highlighted.

Key Words: Angiosarcoma, Chemical exposure, Probable association, Hepatic, Splenic, Formic acid

Introduction

Angiosarcoma of the liver is a primary mesenchymal tumour of the liver. It is an exceedingly rare tumour. It represents only 1 % of all primary liver neoplasms. Therapeutic irradiation, thorium dioxide administration, pyothorax and polyvinyl chloride exposures have all been shown to be predisposing factors to developing angiosarcoma.

This paper highlights the possibility of an association between formic acid and development of angiosarcoma in the liver.

Case Report

A 47-year-old Malay man was admitted with complaints of unremitting epigastric pain and loss of weight for 3 months. He was a rubber tapper for 23 years with a history of exposure to chemical fumes used in his daily work. He used formic acid to coagulate the latex. There was no exposure to other chemicals, radiation or any radiological examination prior to this admission.

On examination, the epigastrium was tender but no mass was palpable. Investigations for his epigastric pain showed normal levels of serum amylase, normal electrocardiograph, normal erect abdominal and chest Xray. Further tests done included abdominal ultrasound that showed hyperechoic lesion in the gallbladder, suggestive of a polyp. The liver, spleen and pancreas were reported as normal. OGDS and ERCP were also normal.

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The patient subsequently underwent an open cholecystectomy. At laparotomy, a hard large nodular mass was seen in the left lobe of the liver and small nodules over the surface of the spleen. A left lateral segmentectomy of the liver, splenectomy and cholecystectomy were done.

Pathological Findings:

Macroscopic examination:

The segment of the liver received measured 16x7x6cm. Cut sections of the specimen showed an irregular solid lesion that measured 8x4x4 cm (Figure 1). The spleen measured 13x8x5cm and had multiple small nodules on its surface. Cut sections of the spleen also showed several large dilated vascular spaces within the parenchyma.

The gallbladder contained a few cholesterol stones. No nodules or polyps were present.

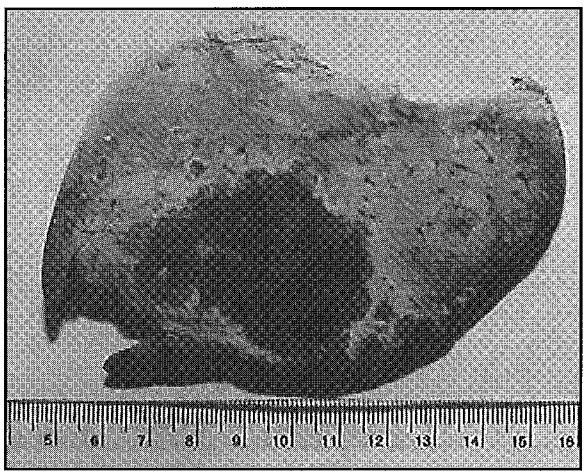

Fig. 1: Shows sliced section of liver with angiosarcoma. The tumour has irregular borders

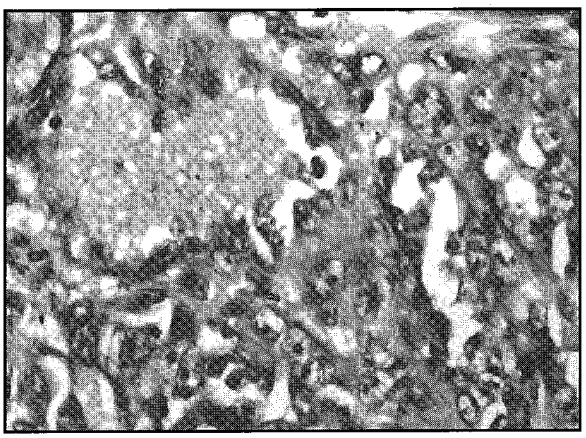

Fig. 2: Shows angiosarcoma with numerous vascular spaces lined by plump malignant endothelial cells. (H&E, original magnification X 400)

Microscopic examination:

Sections of the liver showed irregular vascular spaces that replaced a large area of the liver parenchyma diffusely. Large malignant endothelial cells lined the numerous anastomosing vascular channels. These tumour cells had increased nuclear-cytoplasmic ratio and marked nuclear pleomorphism. A few of the large dilated cavernous spaces were filled with red blood cells

Sections of the spleen showed multiple foci of similar tumour. In other areas, there were dilated cavernous channels lined by flattened endothelial cells.

Microscopic examination of the gallbladder showed no tumour.

Discussion

Angiosarcoma is a rare malignant tumour arising from the endothelial cells of the blood vessels or lymphatic channels. The head and neck are the most common primary sites followed by the liver, trunk, pleural cavity, chest wall, abdominal wall and buttock [1].

Hepatic angiosarcoma is an infrequent tumour. In a USA study, the annual incidence is reported as 0.14 to 0.25 per million population[2]. Recently, its incidence worldwide is dramatically increased because of its epidemiological association with exposure to certain chemicals. Chemical carcinogens are implicated in 1/4 of the cases of angiosarcoma[3]. The most frequently associated chemicals are thorium dioxide (thorotrast), vinyl chloride and arsenic. Diethylstilbestrol, phenelzine and Fowler's solution (potassium arsenite) have also been reported to be associated. Other rare aetiological agents include oral contraceptives, androgen/anabolic steroids and copper sulfate spray. However, in the majority the cause remains unknown.

This patient worked as a rubber tapper for 23 years. His daily activity included adding formic acid to latex in the cottage industry. He did not wear any protective mask during his work. He admitted that he had inhaled chemical fumes emitted when formic acid was used to coagulate the latex. Thus, there was a daily exposure over a period of 23 years to these chemical fumes. It had been reported that 1 in 4 cases of angiosarcoma were associated with exposure to chemical carcinogens[3]. Thus, there could be an association between exposure to chemical fumes when formic acid was added to latex and the development of angiosarcoma in the liver and spleen in this patient.

In my literature search there was no case of angiosarcoma reported with such association. However, awareness and case reports of this rare tumour in patients who may be exposed to chemicals in their occupations would help provide a probable association between possible chemical carcinogens and angiosarcoma.

Angiosarcoma is a high-grade sarcoma with an aggressive course and has a high metastatic rate. Majority of the cases had metastases to other organs namely lung, liver and lymph nodes. This patient had multiple metastatic lesions on the surface and within the parenchyma of the spleen. He also developed lung metastasis 3 weeks after the laparotomy.

Hepatic angiosarcoma runs a fatal course with abysmal prognosis. The majority of patients survive 6 to 8 months after diagnosis. Various treatments have been tried with dismal results. Patients who were treated conservatively with chemotherapy succumbed to the disease in less than 8 months. Radical resection of the liver with transplantation had also been tried. However, survival was not prolonged compared with those on conservative treatment. These disappointing results confirm the aggressive nature of the tumour.

This patient had surgical intervention. A segmental resection of the liver with splenectomy was done followed with chemotherapy. However, he succumbed to his illness 6 months after diagnosis.

Conclusion

Angiosarcoma of the liver is exceedingly rare. It has an aggressive nature and treatment appears futile against this tumour.

The association of several chemical carcinogens with potential hepatotoxicity to cause angiosarcoma has been documented. Cases of angiosarcoma in patients with occupation exposed to possible carcinogenic chemicals need to be reported to highlight any possible link between other toxic substances and this rare tumour.

Subsequently, regulatory actions and measures may be recommended for a safe working environment to reduce morbidity and mortality.

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