ANGIOSARCOMA OF THE LIVER - A CASE REPORT

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ABSTRACT

A 42 year old man presented to our hospital with right hypochondrial pain for 6 months. This had increased in severity and frequency over the last one week and was associated with fever. The patient was febrile and had slightly raised total white cell count. Ultrasonography revealed an enlarged liver with an inhomogeneous mass within the right lobe of the liver. In view of the clinical history, a provisional diagnosis of a liver asbcess was made. A percutaneous needle aspiration was performed under ultrasound guidance. There was no pus but blood stained fluid was aspirated instead. This was sent for cytology. Subsequently a CT scan of the abdomen was performed for further evaluation of the liver mass. This showed an ill-defined, predominantly hypodense mass in the right lobe of the liver and medial segment of the left lobe of the liver suggestive of a malignant liver tumour. A tru-cut biosy of the liver tumour was performed under CT guidance. The histopathological examination revealed that the liver tumour was hepatic angiosarcoma.

The patient went on to develop brain metastasis. He was treated with radiotherapy but succumbed to the disease and died 3 months from the time of diagnosis.

The significant aspects of hepatic angiosarcoma and the imaging modalities used are discussed.

Key Words: Angiosarcoma of Liver, Ultrasonography, C.T. Guidance Biopsy

INTRODUCTION

Angiosarcoma of the liver is a rare tumour with a rapidly fatal course. This tumour constitutes only 2% of all primary tumours of the liver. The following report describes a case of hepatic angiosarcoma. The significant aspects of this malignancy are presented. Diagnostic and therapeutic modalities are discussed.

CASE HISTORY

A 42 year old man presented to our hospital with intermittent pain in the right hypochondrium for 6 months which had increased in frequency and severity over the last week and was associated with fever. He was a business man dealing in household goods and had not been exposed to vinyl chloride, thorotrast, arsenicals and radium. He drank alcohol regularly and was a heavy smoker.

Physical examination revealed tenderness in the right hypochondrium with the liver palpable 5cm below the right costal margin. The liver was smooth and firm. The patient was febrile and had temperature of 37.8 degrees C. He was not jaundiced and the rest of the physical examination was unremarkable.

The initial laboratory test revealed a haemoglobin

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of 14.4g/dl, a slightly raised total white cell count of 11,500/dl, a normal total bilirubin and alkaline phosphatase. Hepatitis B surface antigen was negative. A provisional diagnosis of liver abscess was made.

An ultrasound of the abdomen revealed an enlarged liver with a large mass in the right lobe of the liver. It appeared inhomogeneous but was predominantly hypoechoic. In view of the clinical history this was thought to be an early liver abscess. The spleen, pancreas and both kidneys appeared normal. Subsequently percutaneous needle aspiration of the focal intrahepatic lesion was performed under ultrasound guidance. A small amount of dark blood stained fluid was obtained. This was sent for cytology. No pus was aspirated. To assess this mass further, a CT scan of the abdomen was performed with intravenous and oral contrast. The intravenous contrast was given during the CT scan. Delayed scans were not performed. The CT scan showed a poorly defined, predominantly hypodense mass in mainly the right lobe and the medial segment of the left lobe of the liver suggestive of a malignant liver tumour. The aspirated fluid from the liver send earlier was inadequate for cytology so a percuteneous tru-cut biopsy was performed under CT guidance. Gel foam was injected during the withdrawal of the needle to prevent bleeding.

The histopathological report of the biopsy specimen was that of a primary hepatic angiosarcoma.

10 days later the patient developed a severe headache and became confused. On examination, apart from an up-going plantar reflex on the left there were no localising signs. A CT scan of the brain showed an enhanching focal lesion of about 3cm in diameter within the right cerebellum, associated with moderate hydrocephalus. This was most likely brain metastasis from the hepatic angiosarcoma. A ventriculo-peritoneal shunt was performed and the patient was nursed in the intensive care unit. He improve over the next 10 days and was discharged. He attended the oncology clinic regularly and was given radiotherapy. Two months later the patient died. An autopsy was not performed.

DISCUSSION

A focal, predominantly hypoechoic intrahepatic lesion on ultrasound has a number of differential diagnoses, namely, metastasis, hepatoma, abscess, lymphoma, cavernous hemangioma, hematoma and cyst. In our patient, who had a history of fever, a liver abscess (pyogenic/amoebic) was thought to be the most likely diagnosis. Liver abscesses have a very variable appearance on both ultrasound and CT and may be indistinguishable from hepatic neoplasms (1).

Metastatic carcinoma is the most common neoplasm of the liver and is responsible for 38% of all tumours in the liver. About 90% of primary carcinomas of the liver are hepatocellular carcinoma and 7% are cholangiocarcinomas. Less common tumours include hepatoblastoma, angiosarcoma and sarcoma (2).

Angiosarcoma, also known as haemangioendothelial sarcoma, constitutes only 2% of all primary tumours of the liver. This neoplasm has been reported in association with exposure to thorotrast, arsenicals, radium and vinyl chloride. This rare neoplasm is also associated with systemic diseases like hemochromatosis and von Recklinghausen's disease. Our patient did not have any history of exposure to the above mentioned chemicals nor did he has any evidence of hemochromatosis or von Recklinghausen's disease. He presented with fever and right hypochondrial pain. Clinically an infective process would be the most likely cause but it is not uncommon for fever to occur in patients with a malignancy.

The ultrasound features of angiosarcoma are nonspecific. It usually appears inhomogeneous and predominantly hypoechoic. The hypoechoic areas probably represent the vascular spaces in this tumour. As already mentioned, these appearances are nonspecific as they are also seen in other benign vascular tumours eg. cavernous hemangioma, primary malignant tumours eg. hepatocellular carcinoma and metastasis.

The computed tomographic appearances of hepatic angiosarcoma have been described in detail in the article written by White P.G. et.al (3). The authors recommended that for accurate assessment of this tumour, non-enhanced scans, dynamic contrast-enhanced scans and delayed post-contrast scans should be obtained.

Prior to intravenous contrast enhancement, angiosarcoma appears hypodense relative to normal liver. The non enhanced scan allows easier recognition of areas of tumour enhancement on subsequent contrast enhanced scans. Dynamic scanning during a bolus injection of contrast may demonstrate areas of intense enhancement which would correspond to the more vascular parts of the tumour. In addition, enhancement of the liver parenpchyma may facilitate detection of other small hypodense foci of tumour which might be overlooked. On delayed post-contrast scans, there is progressive enhancement over a period of minutes. This supports the diagnosis of a highly

vascular tumour. In our patient only dynamic contrast enhanced scans were obtained as there was no suspicion of the tumour being a hepatic angiosarcoma. The CT scan of the patient demonstrated an illdefined hypodense mass in the right lobe of the liver.

The computed tomographic features described are not specific to angiosarcoma. The differential diagnosis include simple hemangioma, giant cavernous hemangioma, hepatocellular carcinoma and vascular metastasis. Hemangiomas may be multiple and the patient is usually asymptomatic and liver function test are usually normal. Hemangiomas have a characteristic CT appearance, such as, appearing hypodense on the pre-contrast scan, early peripheral contrast enhancement, progressive opacification from periphery to the centre and an eventual isodense appearance on the delayed scans. Giant cavernous hemangioma have some similar features of simple hemangioma but are distinguished by the larger size of the former and characteristic non-enhancing central cleft. CT features of angiosarcoma are consistent with a vascular tumour. These features are also seen in other malignant liver neoplasms. In a recent study of patterns of contrast enhancement of liver tumours during dynamic and delayed post-contrast CT, contrast enhancement occurred in about 50% of malignant neoplasms which include liver metastasis and a small percentage of hepatocellular carcinomas. The patterns of contrast enhancement included, central or peripheral enhancement during dynamic scanning and partial or rarely complete isodense enhancement on delayed scans (4).

To obtain a definitive diagnosis in most liver tumours, percutaneous biopsy is usually necessary. Histologic specimens should be obtained from the margins of the tumour. Liver biopsy in hepatic angiosarcoma is a potentially dangerous procedure due to the vascular nature of the tumour. There have been reports of fatal or serious hemorrhage following percutaneous biopsy. In our patient, gel foam was injected during the withdrawal of the biopsy needle to aid hemostasis. Hepatic arteriography is the best method for diagnosing vascular lesions. Typically, there is a "pooling" of contrast into "vascular lakes", central areas of hypovascularity and peripheral contrast staining. The hepatic artery or branches may often be displaced by the tumour. The hepatic arteriogram also

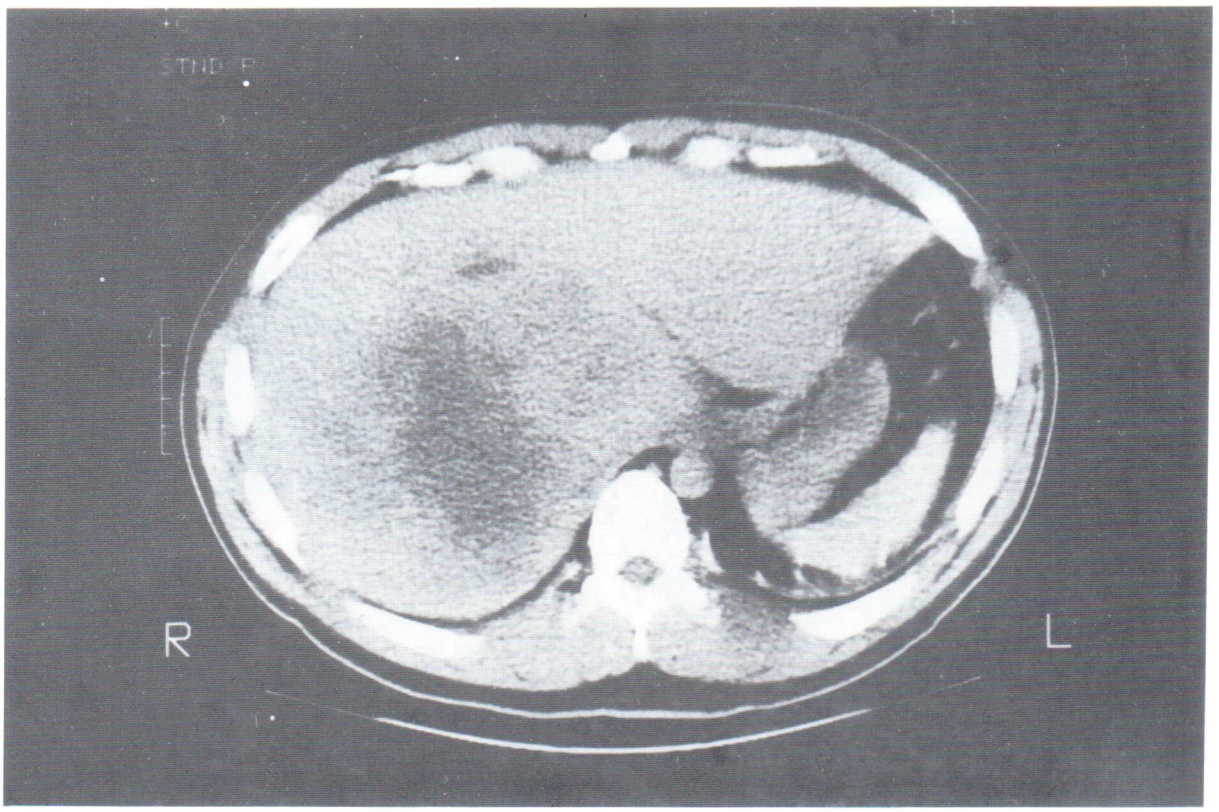

Fig. 1 Non-contrast CT scan of the liver showing a poorly defined, predominantly hypodense mass in mainly the right lobe and medial segment of the left lobe.

serves to outline the vascular supply of the tumour and whether the tumour is localised to one lobe and the remainder of the liver is not compromised. Surgery may then be an option. In our patient surgery was not considered as the tumour involved both the right and left lobes of the liver. Neoadjuvant chemotherapy via a hepatic artery catheter may in the future prove to be useful in following resection in more cases.

Brain metastasis from angiosarcoma is rarely seen because of the rarity of the primary tumour. Sites affected by metastasis from angiosarcoma in decreasing order of frequency are the cervical lymph nodes, lung and spleen. Our patient developed a focal enhancing mass in the right cerebellum. This cerebellar mass was most likely a metastasis from the primary angiosarcoma of the liver.

In summary, inhomogeneous focal intrahepatic lesions are commonly encountered in ultrasonography and CT of the liver. Angiosarcoma appears inhomogeneous and predominantly hypoechoic on ultrasonography. Computed tomographic features are that of a highly vascular tumour. On pre-contrast scans, angiosarcoma appears hypodense relative to normal liver. Contrast enhanced scans demonstrate areas of intense enhancement which corresponds to the more vascular parts of the tumour. Progressive enhancement of the tumour is seen on the delayed post-contrast scans. Although these features are helpful in the diagnosis of angiosarcoma, they are not specific to angiosarcoma. To achieve a definitive diagnosis in most cases of a liver mass, a biopsy is usually necessary, although certain precautions must be taken when there is a suspicion of angiosarcoma.

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Fig. 2 Contrast enhanced CT of the liver demonstrate areas of intense enhancement, which corresponds to the vascular parts of tumour.

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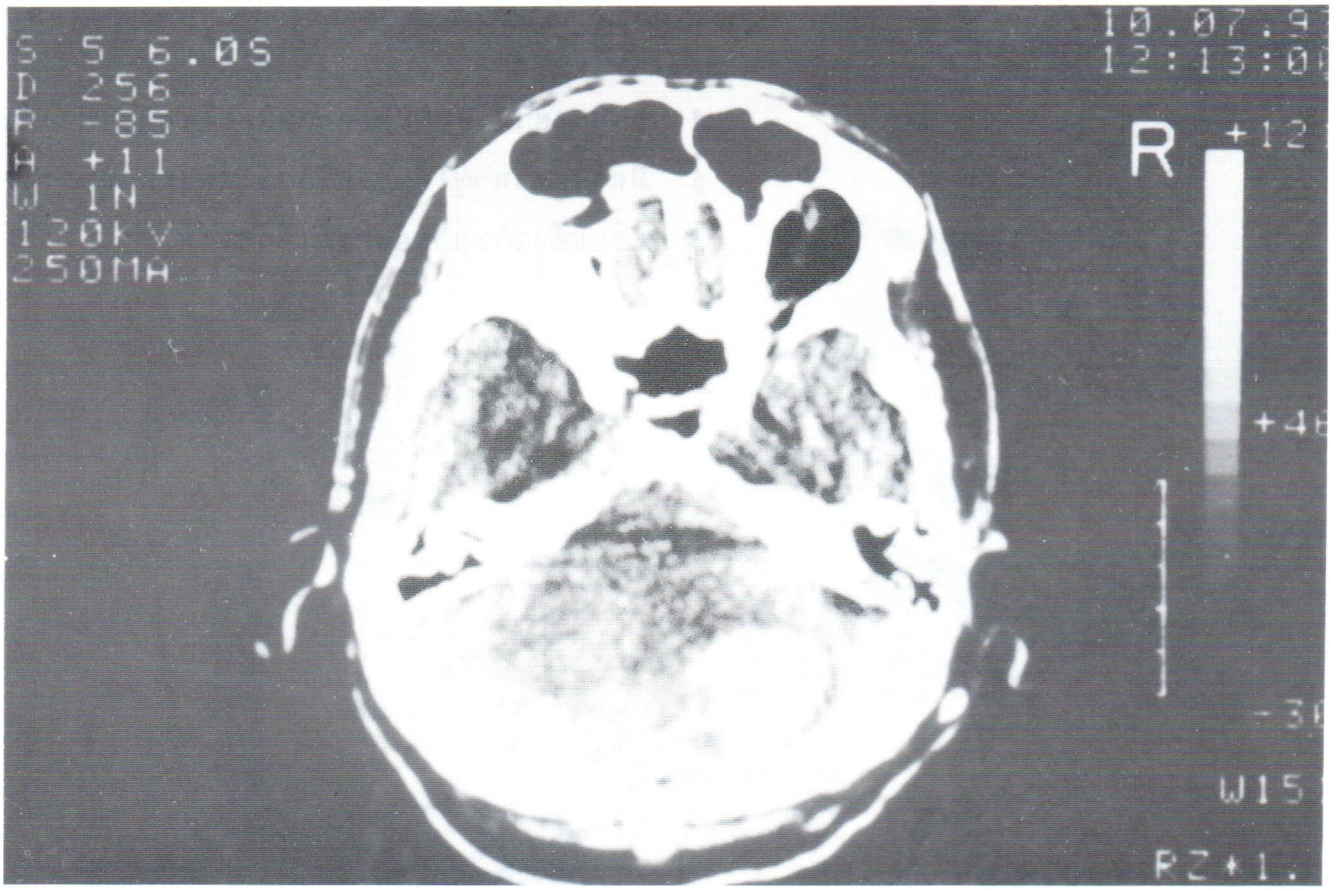

Fig. 3 Contrast enhanced CT of the brain shows a focal enhancing nodule in the right cerebellar hemisphere representing metastatic angiosarcoma.