SYNCHRONOUS MALIGNANT HEPATIC AND BENIGN SPLENIC VASCULAR LESIONS - A RARE OCCURRENCE.

MAHALAKSHMI
Department of Pathology,
MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract: Malignant hemangioendothelioma (Angiosarcoma) is a rare high grade neoplasm arising from endothelial cells with predilection for skin and soft tissues. Herewith we report a case of two different lesions occurring in two different visceral organs, simultaneously. A 30 yrs. old female presented with hepatic and splenic space occupying lesions. Microscopy revealed angiosarcoma of liver and haemangioma of spleen. Hepatic angiosarcomas are primary sarcomas of liver constituting 2 percentage of all hepatic malignancies. Splenic haemangioma is the most common benign tumour in that organ. But, synchronous occurrence of these lesions, have not been reported yet, and we report one such case.

Keyword: Synchronous tumors- hepatic angiosarcoma- splenic haemangioma

INTRODUCTION
Vascular lesions of our body vary from a wide spectrum of well differentiated benign lesions like haemangioma to high grade malignant lesions like hemangioendothelioma. Malignant hemangioendothelioma is a rare neoplasm arising from the endothelial cells of blood vessels. Malignant hemangioendothelioma is seen mostly in skin and soft tissues and only 8% occur in liver. However, haemangioma dominates the benign vascular lesions of spleen. Though these lesions may be a part of some syndromes separately or may show sporadic occurrence, their synchronous presentation in two different organs is rarely reported.

2. CASE REPORT
A 30 years old female presented with hepatomegaly and splenomegaly. Ultrasound of liver showed a mass comprising multiple hypoechoic spaces and multiple calcifications in left lobe with infiltration into right lobe. A splenic subcapsular mass was also seen causing enlargement of spleen. CT abdomen showed, in addition, multiple Para aortic lymph nodes. Lobectomy of liver and splenectomy were done.
3. MACROSCOPIC FEATURES
Liver measured 34X28X10 cm. External surface showed several, small nodular masses. Cut surface showed diffuse infiltration by several, grey-white, gelatinous masses along with many, reddish-brown cystic masses (Fig.1). Spleen measured 23X16X10 cm. Cut surface showed a large, ill-defined, reddish-brown mass (Fig.2).

![FIG.1 LIVER-MACROSCOPY](image1)

![FIG.2 SPLEEN-MACROSCOPY](image2)

4. MICROSCOPIC FEATURES
H & E sections from the liver showed an unencapsulated neoplasm comprising several irregularly shaped vascular spaces lined by tall cuboidal cells with micro papillary processes (Fig.3&4). Nuclei were large, vesicular with some of them showing nucleoli (Fig.5). Tumor cells showed positivity for CD 34 (Fig.6). Few vessels showed thrombi in the lumens. Stroma showed focal areas of fibrosis and hemorrhage. Sections from spleen showed an unencapsulated neoplasm containing dilated vascular spaces lined by flattened endothelial cells with no atypia (Fig.7 & 8). Lumens showed eosinophilic material and blood. As the lymph nodes were not biopsied, it could not be ascertained, whether Para aortic lymphadenopathy was due to metastasis or due to reactive changes. Impression of angiosarcoma of liver and haemangioma of spleen was made.

![FIG.3 LIVER WITH TUMOUR X 4](image3)

![FIG.4 MALIGNANT ENDOTHELIAL CELLS X 10](image4)
5. DISCUSSION:
Angiosarcomas are malignant tumors that recapitulate many of the functional and morphologic features of normal endothelium\(^1\). They have predilection for skin and superficial soft tissues, but deeper forms do occur. The presentation and behavior of these tumors differ depending on location. Hepatic angiosarcoma also known as kupffer sarcoma is a primary sarcoma of the liver which accounts for only 2% of all primary hepatic malignancies\(^2\). Angiosarcoma is associated with environmental or occupational exposure to carcinogens (thorium dioxide, vinyl chloride, arsenic and radiation)\(^2\). There is also an association with hemochromatosis and vonRecklinghausen disease\(^2\). In most cases of primary hepatic angiosarcoma, no obvious risk factor can be identified. Mutations of k-ras and p53 are implicated in pathogenesis\(^2\).

The most frequent age at presentation is between the 6th and 7th decade of life with a higher incidence in males (4:1)\(^2\). Clinical presentation includes abdominal pain, hepatosplenomegaly, jaundice, ascites, anorexia, weight loss, intraperitoneal hemorrhage and hepatic failure\(^3\). The utility of imaging tests is
limited. Abdominal CT may reveal heterogeneous hepatosplenomegaly with irregular liver morphology, showing alternate hyperdense and hypodense areas\(^2\).

The definitive diagnosis is always given by histopathology\(^2\). Histology reveals the presence of endothelial neoplastic cells with epithelioid appearance, pleomorphic and hyperchromatic nuclei and prominent nucleoli, lining vascular spaces with cavernous morphology. Focal areas may show formation of intracytoplasmic vacuoles. Immunohistochemistry shows positivity for endothelial markers, especially CD-31 and CD-34, factor VIII, D2-40 and c-kit\(^4\).

Metastases of hepatic angiosarcoma usually affect the lungs and spleen. Angiosarcoma has very limited treatment options. Without treatment, a majority of patients die within 6 months of diagnosis\(^5\). Surgery is the mainstay of treatment\(^3\). Liver transplantation is not useful because of high recurrence rate. There is no data to support, improved survival for patients treated with chemotherapy and radiotherapy.

Prognosis is very poor. Median survival varies between 12-36 months\(^3\). Cause of death in most cases is intraperitoneal haemorrhage or liver failure.

**CONCLUSION:**

Hepatic angiosarcoma is a rare hepatic malignancy, with a very aggressive course and with very limited treatment options. Occurrence along with a benign vascular lesion in spleen is a rare event and to our knowledge no such case has been reported in the literature.

**REFERENCES:**


