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*Letter to Editor.*

# Primary Hepatic Angiosarcoma in an Adolescent: Letter To Editor

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## Editor

Angiosarcoma (AS) is an uncommon malignant tumor presenting a recognizable vascular differentiation. It accounts for only 2 to 4 % of soft tissue sarcomas[1] and occurs mainly in the adulthood and elderly, but occasional cases in children have been reported.[2] It can develop in any site but the most common locations include the skin, soft tissues, breast, bone, liver, and spleen, while the rare cases seen in children occur especially in mediastinum including the heart and pericardium. Primary hepatic angiosarcomas (PHAs) are rare, and account for <5% of all angiosarcomas[3] and 1.8% of all hepatic malignancies.[4]

16 Year old male was treated for malaria 2 months back, following which fever persisted along with severe right hypochondriac pain. USG-was done which showed, heterogeneous multicystic lesion in the right lobe of liver, with mild subdiaphragmatic collection (Ruptured infected hepatic cyst ?) and multiple portahepatic enlarged lymph nodes. Patient was admitted and USG guided FNAC was planned for the patient, but since pus was thick ,procedure was cancelled and patient was advised to come back again following ingestion of metrogyl for 14 days. Within 1 week of discharge patient developed severe abdominal pain with recurrence of fever, repeat USG-showed ruptured hepatic cyst, CT scan showed ruptured hydatid cyst?. Exploratory laparotomy was planned for the patient which showed hemangioma like tumor that had caused pneumoperitoneum due to its rupture. Peritoneal lavage was done and abgel pack in tumor bed was given. Biopsy was done and sent for HP study .Gross-4 bits of grayish brown tissue received all together measuring 3cm .Histopathology showed small clusters of hepatocytes along with dilated vascular channels with extensive areas of hemorrhage and necrosis, large sheets of pleomorphic looking cells having hyperchromatic nuclei, scanty cytoplasm separated by hemorrhagic areas, mitosis were present Fig 1(a,b,c),Fig 2 &3. Final diagnosis was given as angiosarcoma of liver. IHC was not done as patient was immediately referred to higher centre due to deteriorating health conditions.

The etiology of PHA remains unknown although previous case-control studies have indicated that ~1/3 of all cases of PHA appear to be caused by exposure to environmental carcinogens, including thorium dioxide, arsenical insecticides or polyvinyl chloride ,but exposure to these chemicals is rare.[5]

Epithelioid hemangioendothelioma (EHE), a rare and usually low-grade malignant tumor, should be considered in the differential diagnosis of PHA.[6] The diagnosis of PHA is difficult, particularly if the patient does not present a history of exposure to carcinogens, as in our case. Tumor markers such as CD31, CD34, podoplanin and FVIII-related antigen are often used in combination for the immunohistochemical diagnosis of angiosarcomas. In particular, CD31 and FVIII-related antigen has been suggested to be the most sensitive of all.

PHA is associated with poorer prognosis in comparison to other types of angiosarcomas which is attributed mainly, due to the rapid progression of the disease, its high recurrence rate, and its resistance to traditional chemo and radiotherapies.[7] However, CT or US-guided fine-needle aspiration biopsy are dangerous and non-diagnostic procedures. Thus, the pathological diagnosis of PHA is essential. In our case patient succumbed to death few days of referral to higher centre.

Selective hepatic angiography in combination with dual-phase spiral CT are other diagnostic modalities for diagnosing PHA.[8] Due to the high recurrence rate and poor post-transplant survival rate of patients, liver transplantation is no longer provided.[9] Currently, the best treatment option for PHA is partial surgical resection of the liver to remove the tumor.[10]

Primary angiosarcoma of the liver is an exceptional rare angiosarcoma and very little information on this entity is available in literatures. The diagnosis of Primary Hepatic Angiosarcoma remains challenging and a definitive diagnosis of this uncommon tumor depends solely on histomorphology. An early and correct diagnosis in these cases can lead to proper management and favorable outcome.

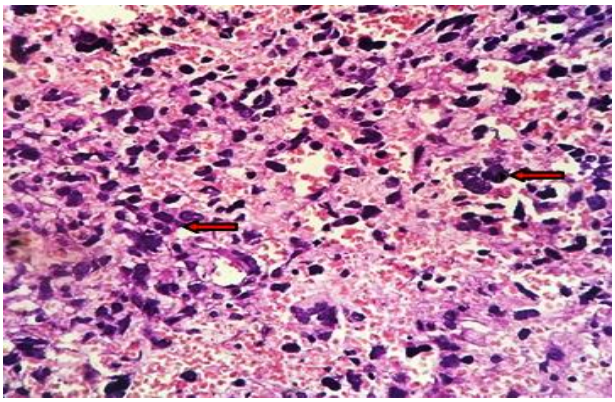

**Fig 1(a): 100x pleomorphic cells (red arrows) with areas of hemorrhage**

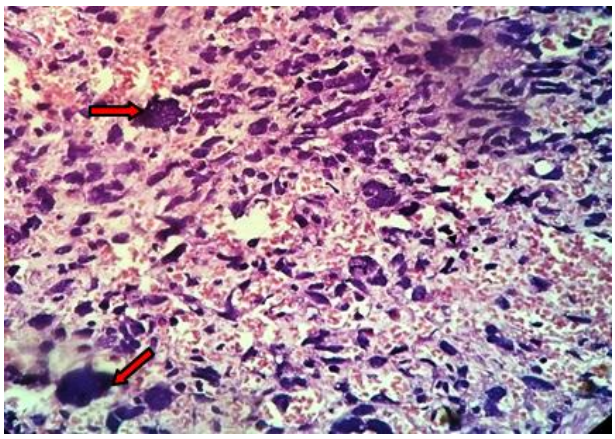

**Fig 1 (b): 100x-pleomorphic cells (red arrows) with areas of hemorrhage.**

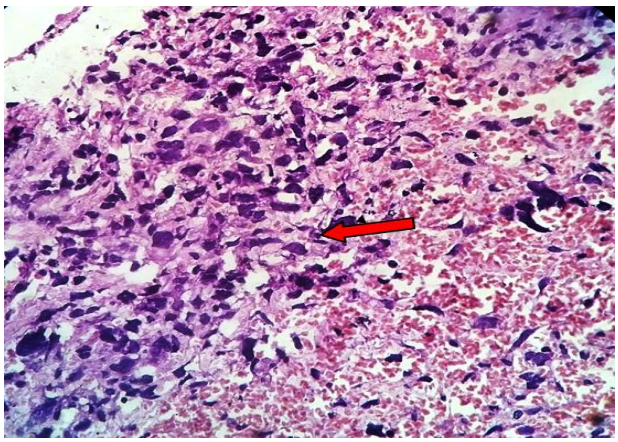

**Fig 1 (c): 100x- Pleomorphic cells in a focus (red arrow)**

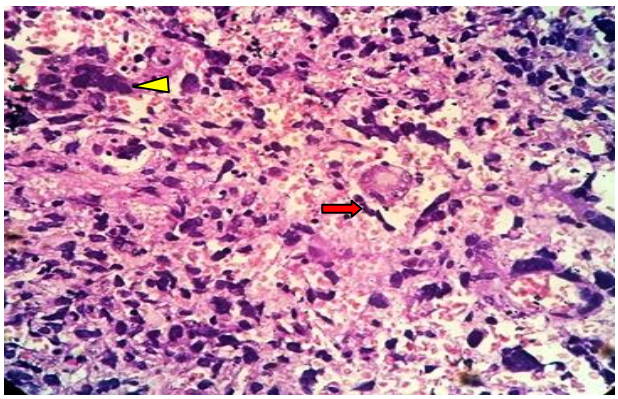

**Fig 2: 100X-Tumour giant cell (red arrow) with area showing pleomorphism (yellow pointer).**

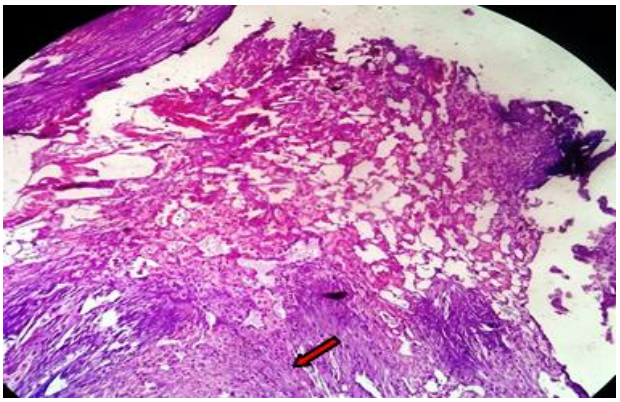

**Fig 3: 100X- area of normal hepatocytes (red arrow)**

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