UNDIFFERENTIATED (EMBRYONAL) SARCOMA OF THE LIVER-
A CASE REPORT

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Abstract: Primary hepatic tumors are rare, but unfortunately two-thirds of them are malignant, representing the third most common abdominal cancer encountered in pediatric practice, after Wilms tumor and neuroblastoma (1). Undifferentiated (embryonal) sarcoma of liver, which has also been termed mesenchymal sarcoma, malignant mesenchymoma, primary sarcoma of the liver and fibromyxosarcoma is uncommon and typically occurs during the first two decades of life. It is probably the third most common primary malignant hepatic tumour of children, after hepatoblastoma and hepatocellular carcinoma (2). The typical presentation is that of an abdominal mass and pain but fever is a variable feature. The treatment for the embryonal sarcoma of the liver is surgical resection, with adjuvant chemotherapy and possibly radiation therapy. The prognosis is generally poor.

Keyword: Liver- mesenchymal- undifferentiated-embryonal sarcoma.

INTRODUCTION: The incidence of childhood primary malignant tumors of the liver ranges from 0.5-2% of all pediatric malignancies. Embryonal sarcoma represents 9-15% of all hepatic tumors in children, ranking third among malignant tumours of the liver after hepatocellular carcinoma and hepatoblastoma.

CASE SUMMARY: An eight-year-old male child presented with abdominal distension and pain since one month. He also had low-grade fever and loss of appetite. Clinical examination revealed a large mass in the abdomen. Abdominal sonography revealed a large lobulated heteroechogenic mass with cystic areas in the right lobe of liver. Computed tomography of the abdomen revealed a large (13.7x9.4x9.6 cm), multiloculated predominantly cystic lesion with non-enhancing septations and focal peripher al calcifications in the I, V and VI segments of liver extending into Morrison’s pouch with perilesional edema. The features were considered
suggestive of mesenchymal hamartoma (Fig. 1 & 2). Serum Alpha Fetoprotein level was normal.

External surface appeared nodular (Fig. 3). Cut surface showed a well circumscribed lesion, measuring 14x10x9 cm, with a variegated appearance. Solid and cystic areas were seen along with areas of haemorrhage and necrosis (Fig. 4).

**Fig 3: Gross - External surface - nodular**

Fig 1: CT scan - Large (13.7x9.4x9.6 cm), multiloculated, predominantly cystic lesion

**Fig 1:**

Fig 2: CT scan - Large cystic lesion with non-enhancing septations and focal peripheral calcifications

**GROSS:**
Right hepatic lobectomy was done. Grossly the right hepatectomy specimen measured 18x9x8 cm.

**Fig 4:**

**Fig 2:**

Multiple sections studied showed a well circumscribed, unencapsulated neoplasm (Fig. 5), composed of cellular and less cellular areas containing a mixture oval to spindle cells and polyhedral cells dispersed in a loose, richly vascularised, myxoid matrix (Fig. 6). Nuclei were hyperchromatic, round, oval to elongated and showed marked degree of pleomorphism (Fig. 7). Several tumor giant cells were seen (Fig. 8). The mitotic activity was brisk, including few abnormal mitotic figures (Fig. 9). Extracellular hyaline globules (Fig. 10) seen were strongly positive for periodic acid–Schiff (Fig. 12). Areas of haemorrhage and necrosis were present (Fig. 11).
Fig 5: Well circumscribed margin. H&E x 40
Fig 6: Fascicles of spindle shaped cells with anaplastic cells. H&E x 10
Fig 7: Hyperchromatic nuclei with pleomorphism. H&E x 40
Fig 8: Tumor giant cells. H&E x 40
Fig 9: Abnormal mitotic figure. H&E x 40
IMMUNOHISTOCHEMISTRY:
On immunohistochemistry, the tumor cells showed diffuse strong positivity for CK (Fig. 13), focal strong positivity for Vimentin (Fig. 14) and were negative for CD 34 (Fig. 15).

DISCUSSION:
Undifferentiated sarcoma of the liver is very rare and presents in children during the first two decades of life. In the largest published series to date, Stocker and Ishak found that in 16 of their 31 cases the patients were between 6-10 years of age, although the tumor may occur in adults\(^1\). There does not seem to be any sex predominance in contrast to other hepatic cancers of childhood\(^3\). Embryonal sarcoma of the liver is a rapidly growing malignant tumor. It usually presents with abdominal mass and/or abdominal pain. Other complaints include fever, weight loss, anorexia, malaise, lethargy, nausea and vomiting\(^4\). Haemorrhage into or rupture of the tumor occurs occasionally. Tumor may extend into right atrium via inferior vena cava and may cause dyspnoe\(^5\)\textsuperscript{a}. The tumor is usually a single, large, variably haemorrhagic, necrotic or cystic mass in the right lobe of liver. Imaging studies (sonography and computerised tomography scan) demonstrate somewhat heterogenous appearances, ranging from cystic tissue with multiple septa to more solid tissue reflecting the extent of necrosis and cyst formation. Serological investigations and markers are usually negative or unhelpful in identifying the specific nature of the tumor. A non specific defect may be seen on an isotope liver scan and on angiographic examination the tumor may appear hypervascular, hypovascular or avascular\(^3\)\textsuperscript{b}. Periodic acid Schiff positive intracytoplasmic pink globules are present in the Majority of cases\(^6\). Ultrastructurally and immunohistochemically, most of the cells have features of undifferentiated mesenchymal cells, fibroblasts and myofibroblasts, but others may show differentiation toward smooth and skeletal muscle elements on the one hand and toward epithelial cells on the other\(^7\). Thus immunoreactivity has been described for vimentin, alpha 1-antitrypsin, alpha 1-chymotrypsin, lysozyme, CD 10, glypican-3 (a feature it shares with mesenchymal hamartoma), smooth muscle actin, desmin and keratin\(^8\). This peculiar combination of phenotypic features has lead some authors to postulate a histogenetic relationship with
primary hepatic embryonal rhabdomyosarcoma and others to suggest that the tumor represents an anaplastic (sarcomatoid) variant of live cell carcinoma. Both of them seem unlikely prospects, in view of the negativity of these tumors for myogenin and HepPar-1. The tumor rarely metastasizes but spreads by direct extension into adjacent organs and sometimes extends into the right atrium via the inferior vena cava. Rupture of the tumor can occur and massive intraperitoneal spread has sometimes been found. The metastases that have been reported were principally in the lung, bone, pleura and peritoneum. The treatment for embryonal sarcoma is surgical resection. Without the surgical resection the mean survival period is less than one year and mortality is greater than 80%. The lesions seldom are resectable and treatment with radiotherapy and chemotherapy has produced disappointing results. The prognosis is generally poor although there have been some reports of five-year survival and local recurrence is a problem, particularly in cases in which the tumor has been incompletely resected.

CONCLUSION:
The case is presented for its rarity. It is important to distinguish embryonal sarcoma from mesenchymal hamartoma which it can mimic on imaging studies.

REFERENCES:


