ABSTRACT

Undifferentiated Embryonal Sarcoma of Liver (UESL) is a rare and highly malignant primary hepatic tumor of mesenchymal origin. We present a case of Primary UESL in 14 year old female, presenting with a painful swelling in right hypochondrium and jaundice. CT Scan revealed a 18cm mass involving segment V and VIII of liver. Core needle Biopsy revealed proliferation of pleomorphic spindle cells with few interspersed rhabdoid cells. Immunohistochemical staining revealed tumor cells were immunopositive for Vimentin and Desmin, rendering it to be a UESL. Total resection with preoperative or postoperative radiochemotherapy is currently considered to be the key approach for improving the survival rate. Thus, UESL is a rare hepatic tumor, but an early diagnosis and prompt treatment may be useful in improving patients survival rate.

KEYWORDS

Undifferentiated Embryonal Sarcoma, Liver, Tumor

INTRODUCTION:

Undifferentiated Embryonal Sarcoma of Liver (UESL) is a Rare and Highly Malignant Primary Hepatic Tumor of Mesenchymal Origin that accounts for 09 to 15% of Pediatric Liver Malignancy. Most cases (90%) occur in children aged 6-10 years. Some series report a Female Predominance, while others report an even-gender distribution. Tumor sizes are between 10 cm to 30 cm. UESL arise more commonly from Right lobe of Liver and presents usually with Painful Right Upper Quadrant Mass, Fever, Nausea, Anorexia and rarely Jaundice. Imaging demonstrates Cystic and Solid components. Histopathological Examination of biopsy showed Cellular tumor with Anaplastic Spindle cells having abundant eosinophilic cytoplasm. Immunohistochemistry: Most cases are positive for Vimentin, Desmin, CD68, α1- antitrypsin and CD10. Complete tumor resection followed by adjuvant chemotherapy and/or radiotherapy improves survival and reduces recurrence in patients with UESL. In the present study we present a case of UESL in a 14 year old female.

Case Presentation:

A 14 year Old female Presented to Our hospital with chief complaints of Fever, Jaundice and Pain in Right Hypochondrium since 5 months. Her hemogram revealed haemoglobin- 8.8gm/dl, total count- 14 x 10^3 /cumm and platelets-486 x 10^3 /cumm. Other Laboratory examination results were as follows: α- fetoprotein (AFP) -2.09ng/ml; S.Albumin- 2.73gm/dl ; Lactic dehydrogenase -2273.9U/L ; Alanine transaminase (ALT) -59.3 U/L ; Aspartate transaminase (AST) -86.8U/L ; Alkaline phosphatase (ALP)-230.2IU/L, Total Bilirubin- 1.61mg/dl, Direct Bilirubin-0.99mg/dl. Coagulation profile was normal.

CT Scan showed about 33x18x17 cms sized Heterogeneously enhancing soft tissue density lesion involving Segment V and VIII of liver with peripheral enhancement. Lesion shows exophytic growth and spaying of Right and Left portal vein. Lesion abuts and displaces Right Kidney posteriorly. Lesion Compresses IVC. Ultrasonography guided true-cut biopsy was performed and sent to Histopathological Examination.

On Gross Examination, Specimen showed multiple linear grey white soft to firm tissue pieces measuring about 0.2 to 1.5 cm in length. Hematoxylin & Eosin(H & E) stained section showed Proliferation of Pleomorphic Spindle cells with hyperchromatic nucleus, inconspicuous nucleoli and moderate to abundant Eosinophilic Cytoplasm. Possibility of Poorly Differentiated Malignant Tumor (PDMT) was suggested.

Immunohistochemistry showed Immunoreactivity for Vimentin and Desmin and Immune negativity for Heppar-1, Glypican 3 and Myo-D1. Hence, a diagnosis of Undifferentiated Embryonal Sarcoma of Liver (UESL) was given.

Patient was treated with 5 cycle of Chemotherapy -Cyclophosphamide, Vincristine and Adriamycin only without surgical resection.

DISCUSSION:

Undifferentiated Embryonal Sarcoma of Liver is a rare aggressive neoplasm with Poor Prognosis, which predominantly affects Children aged 6-10 years without gender predilection 11. The Tumor is mainly localized or found in the Right hepatic lobe (50%), rarely develops in Left hepatic lobe (22%) or the Bilateral lobe (20%). Laboratory
Studies are non-specific and have shown that patients with UESL exhibit low Albumin, elevated LDH, Anemia and Abnormal liver function [2-5]. Serum assays for tumor markers including AFP, Cancer antigen 19-9 and Carcinoembryonic antigen yield normal results. UESL typically has a diameter of 10-30 cm with a solitarily clear boundary. Haemorrhage, Necrosis and Cystic degeneration are frequently observed while clinical manifestations include abdominal mass, pain, fever and rarely jaundice.

Abdominal CT images show large cystic, solitary and well-circumscribed mass with areas of necrosis and haemorrhage. Abdominal USG reveals large multicellular or unicellular cystic and solid liver mass. The cystic region exhibits a large, mixed and disorderly low level echo and the solid areas of the mass demonstrate a mixture of high and low level echos. Positron emission tomography (PET)-CT Scans are also used in the diagnosis of UESL. They have a critical diagnosis value for UESL patients, particularly those with metastasis of an extra-hepatic organ.

UESL usually occurs as a single and well-circumscribed lesion grossly. The Well-demarcated appearance is created by a fibrous pseudocapsule, which is formed by compressed liver parenchyma. Cut Surface reveals a heterogeneous appearance of grey-white, glistening solid tumor alternating with cystic, gelatinous areas. In addition, dark-brown areas of haemorrhage and yellow, softer areas of necrosis are often seen grossly. Microscopically, the solid component of UESL appears Sarcomatoid, with a myxoid background. The cells are spindle or stellate shaped, with inconspicuous nucleoli and ill-defined cell borders. Multinucleated cells and bizarre cells with hyperchromatic nuclei are often seen between the sarcomatoid cells. Numerous mitotic figures are easily identified throughout the tumor. Characteristically, numerous eosinophilic globules can be seen in the tumor cell cytoplasm and extracellular matrix. These globules are PAS positive and Diastase resistant. UESL is often considered a Malignant evolution of Mesenchymal Hamartoma (MH). Many Studies have shown that UESL does not have a Specific Immunophenotype. Most cases are positive for Vimentin, Desmin, CD-68, e-1 antitrypsin and CD-10. The main differential diagnosis for UESL in the Pediatric population are Hepatocellular carcinoma (HCC), Rhabdomyosarcoma (RMS), Hepatoblastoma and Mesenchymal Hamartoma(MH). Individual markers are often not helpful in differentiating UESL from other liver tumors. Therefore, IHC Pannel is usually performed to help with the diagnosis. In Practice, Negative markers are valuable to rule out the differential diagnosis. Heppar-1 and Glypican 3(GPC3) known to be a diagnostic markers for Hepatocellular carcinoma and Hepatoblastoma. Myogenin and MyoD-1 positivity seen in Embryonal rhabdomyosarcoma. CD-34 is positive in Solitary fibrous tumor and Vascular neoplasms. Gastrointestinal stromal tumor(GIST) is positive for both C-kit and CD-34. Negative staining for S100 and Melanin markers is helpful to exclude Melanoma and Neural tumors.

Previously, Prognosis of UESL had been poor. In 1990 leuschner et al. reported a low survival rate (37%) of patients with UESL [6]. At that time management of UESL relied primarily on surgical resection. However, the Prognosis has slowly improved as these Patients are managed with Multimodal treatment including Radiation therapy and Chemotherapy. Neoadjuvant Chemotherapy is often helpful in Unresectable cases. Postoperative Chemotherapy and Radiotherapy are reasonable option particularly in surgical cases with positive margins. Studies have shown Improved Survival rates ranging from 70% to 100% in patients who were treated with multimodal therapy. The recurrence rate in UESL is higher during the first 2 years after surgery and the risk is higher with Positive resection margins and Cases with Spontaneous or Iatrogenic rupture of the hepatic lesion.

CONCLUSION:
Although Primary Undifferentiated Embryonal Sarcoma of Liver is predominantly seen in children, it can occur at any age. Because its Clinical and Radiological findings are often not specific, the diagnosis of UESL, relies on Histopathological diagnosis and Immunohistochemical stain for confirmation. Studies of more number of cases required to identify more effective strategies for treating and to improve the survival of patients with UESL.

REFERENCES