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UNDIFFERENTIATED EMBRYONAL SARCOMA OF THE LIVER – A RARE CASE REPORT

Gastroenterology

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ABSTRACT

Undifferentiated Embryonal sarcoma of liver (UESL) is a rare and unique entity of liver which primarily affects children and adolescents. Delayed diagnosis is very common due to lack of clinical characteristics, laboratory markers and radiological findings. We report a case of an 11-year-old boy presented with right upper quadrant abdominal pain, mass in upper abdomen, non-projectile vomiting and 2 kg weight loss in three weeks, diagnosed to have UESL. Extended right hepatectomy was performed followed by adjuvant chemotherapy. The prognosis of UESL was not good in past but recent evidences revealed that long term survival is possible if complete resection is done, followed by postoperative chemotherapy.

KEYWORDS

INTRODUCTION

Undifferentiated Embryonal Sarcoma of liver (UESL), also known as malignant mesenchymoma or embryonic sarcoma is a highly malignant mesenchymal neoplasm of liver. UESL is the third most common primary liver malignancy that affects children of 6 – 10 years age, without racial, ethnic or gender predominance. (Putra; & Ornvold, 2015) About 90% of UESL cases reported in children, which represents 5 – 13% of all paediatric hepatic tumours. Herein, we present a case of UESL. (Gao et al., 2013) The case was managed successfully with surgery and post-operative adjuvant chemotherapy.

CASE REPORT

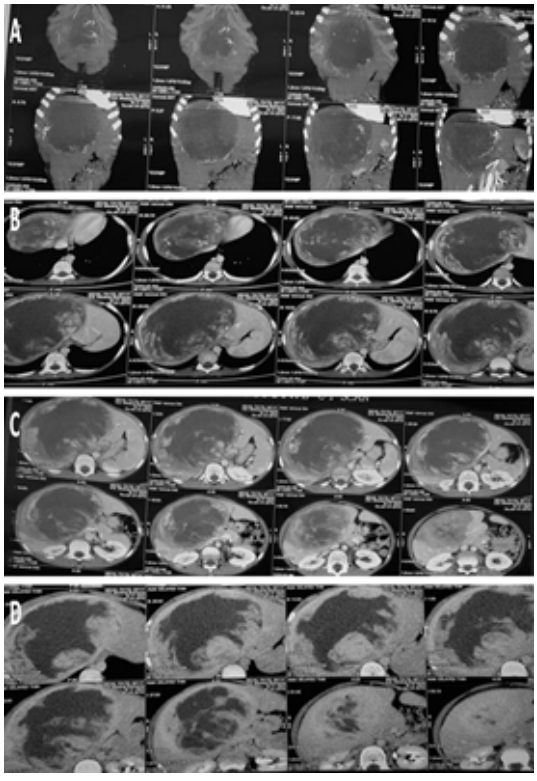

Figure 1: Contrast enhanced computed tomographcal examination of abdomen (A) Arterial scan; (B) Portal scan; (C) Venous Scan; (D) Delayed Scan

An 11 year old boy admitted to our tertiary referral centre with twenty days history of nonspecific right upper quadrant abdominal pain, a visible mass in upper abdomen and had non-projectile vomiting. Moreover, he had history of 2 kg weight loss in last 3 weeks' time. His vitals were normal. His total blood counts, liver function tests and α–fetoprotein were normal. A mass was palpable in right upper quadrant mobile (on respiration); 10cm ×15cm in size. Ultrasonography revealed hepatomegaly with large mass of 14cm ×15cm ×14cm in segment 4, 5, 7 & 8 of liver. Contrast enhanced computed tomography (CECT) scan showed a large liver lesion with delayed fluffy enhancement without venous washout in central and right liver (Fig1 : A, B, C, D) which suggested findings in favour of atypical giant haemangioma versus sarcoma. Patient was admitted for surgery 2 days prior, he developed high grade fever (101-105°F) a night prior to surgery. He was treated with Linazolid and Meropenem for 7 days but fever did not subside. On 7th day his Procalcitonin level was 0.15 ng/ml and total leukocytes counts were also normal hence diagnosis of tumour related fever was made. Patient was posted for surgery on next day. Modified extended right hepatectomy was performed (4a, 5, 6, 7 & 8) (Fig.2 A,B, C,D).

The mass was 14cm ×13 cm x 13 cm in size. Post operatively fever disappeared and patient recovered uneventfully. Pathologic examination of resected specimen revealed 2650 gm right hepatic mass.

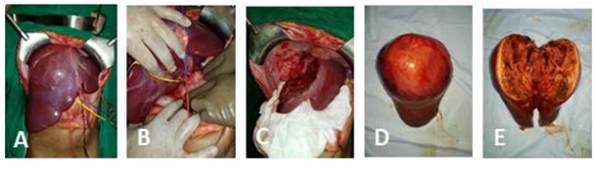

Figure 2 Modified Extended right hepatectomy (A) Exposure of liver; (B) Identification of pedicle; (C) resection of specimen; (D, E) Resected specimen with solid tumour

The histopathology showed malignant neoplasm with predominantly spindle cells suggestive of undifferentiated liver sarcoma. Immunohistochemical (IHC) staining was positive for Desmin, Vimentin, and CD10 as well as negative for Myogenin, Cytokeratin, β – catenin, SMA which confirmed diagnosis of UESL. Patient received 6 cycles of chemotherapy. Post-chemotherapy CT scan showed no signs of recurrence at one year.

DISCUSSION

Undifferentiated Embryonal sarcoma of liver (UESL) is a rare and aggressive hepatic mesenchymal tumour. It was first reported by Stocker et al as a Mesenchymoma in 1978. Later it was reported with

other terms including 'mesenchymal sarcoma', 'embryonal sarcoma', 'fibromyxosarcoma' and 'primary sarcoma of liver'. (Zhang, Lei, Zuppan, & Raza, 2016) Right hepatic lobe is frequently affected with UESL then left hepatic lobe or both lobes. (Gao et al., 2013) Patients of UESL usually presents with clinical features including an abdominal mass with or without abdominal pain, fever, nausea, vomiting, lethargy, constipation, respiratory distress and rarely jaundice. Usually fever is associated with haemorrhage and necrosis identified in tumour. Similarly, we also observed persistent high grade fever because of tumour necrosis. (Putra; & Ornvold, 2015)

UESL associated with non-specific laboratory findings. Results of liver function tests, tumour markers including AFP, cancer antigen 19-9 and carcinoembryonic antigen are usually normal. However, it may present with laboratory findings of mild leucocytosis, low albumin, anaemia & elevated lactic dehydrogenises (LDH). In rare cases increased α-fetoprotein and cancer antigen have been reported. Preoperative diagnosis of UESL is difficult because radiological changes of UESL are also non-specific. (Gao et al., 2013; Putra; & Ornvold, 2015) Usually, an ultrasonography reveals a large mass with mixed solid and cystic components. Such a finding of UESL may be mistaken for hydatid disease or benign hepatic lesions or even hepatoma. (Zhang et al., 2016) Pachera et al. reported 23.5% of delayed diagnosed cases of UESL because it appeared as large cystic mass in imaging findings. (Pachera et al., 2008) Often, CT-scan shows a large hypo dense mass with multiple septations with delayed enhancements. Moreover, MRI may detect vascular invasion, biliary obstruction or adenopathy which is helpful for surgical planning. Angiography may show all patterns of avascular, hypo-vascular and hyper-vascular morphology within liver. (Zhang et al., 2016)

The definite diagnosis of UESL relies on pathological analysis and IHC findings. Macroscopically, UESL is 10-25 cm large, single well–circumscribed mass, with a solitary clear boundary. Predominantly, it is solid but frequently it has foci of cystic or gelatinous degeneration, haemorrhage and necrosis. Microscopically, UESL appears composed of medium to large spindled, oval or stellate cells with loose or even myxoid material with variable cellularity. (Putra; & Ornvold, 2015) The IHC of UESL shows the positive expression of SMA, a-ACT, desmin, vimentin, actin and rarely for PCNA, CK 8/18 and p53, while negative expression of AFP, S-100, CEA, CA19-9 and cytokeratin. (Putra; & Ornvold, 2015)

Currently, there is no universal treatment protocol for UESL. Complete resection followed by chemotherapy is the mainstay of treatment. When tumours are unresectable liver transplantation may be the option. In the past, prognosis of UESL has been poor. (Putra; & Ornvold, 2015) Stocker et al. reported poor prognosis of UESL with 80% mortality in a year. The introduction of multimodal therapy including primary resection followed by adjuvant chemotherapy and radiation improved prognosis with long term survival rates.(Stocker & G. Ishak, 1978)

In conclusion, we hereby report a rare case of UESL in a child. He was treated successfully by tumour resection followed by chemotherapy. He was disease free on 1year follow up.

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