INTRODUCTION
Rhabdomyosarcoma (RMS) occurs infrequently in the liver. Rhabdomyosarcomas are malignant tumours that display features of striated muscle differentiation. They are the most common soft-tissue sarcomas among children. In adults however, these are very rare. We report a case of a primary embryonal rhabdomyosarcoma of the liver in a 17 years old boy. This was confirmed by histological examination using immunohistochemical analysis (LCA negative, desmin positive, myogenin focally positive and cytokeratin negative) and site was confirmed by PET CT scan. He received multiple chemotherapies including (doxorubicin, ifosfamide, dacarbazine; gemcitabine, paclitaxel; vincristine, actinomycin D, cyclophosphamide) and longest sustained stable disease was seen with gemcitabine-paclitaxel regimen. The patient died 31 months after the first presentation, secondary to complicated abdominal progressive disease. The poor prognosis and early death of most previously reported cases imply the need for investigation of a more effective treatment method of this uncommon tumour.

CASE REPORT
A 17-year-old boy presented with a 6 weeks history of heaviness in epigastrium and right upper quadrant, associated with anorexia and generalized weakness. He had no history of vomiting, diarrhea, and jaundice or weight loss.

On examination, there was an obvious swelling in upper abdomen with palpable firm mass in the right upper quadrant and epigastrium. Serology for hepatitis B surface antigen and hepatitis C was negative. His ESR showed 26 mm fall at the end of the first hour while the blood complete picture, urea, creatinine, liver function tests, serum total proteins, and serum α-fetoprotein and β-HCG levels were normal.

Ultrasonography and CT scan of the abdomen revealed a huge mass apparently arising from left lobe of liver measuring 20 x 13 cm. There were few satellite lesions in right lobe of liver, largest in segment V measuring 3 cm with proposed differentials of metastasis from a germ cell tumour, primary liver sarcoma, or lymphoma. There were some small left paravertebral lymph nodes at the level of renal hila. His CT scan chest was normal. Subsequently, a Positron Emission Tomography (PET) scan was also performed (Figure 1), which revealed primary hepatic lesion with a lymph node in left paravertebral area. A 4 core ultrasound guided biopsy of hepatic lesion was performed which showed malignant round cell tumour (LCA negative on immunohistochemical staining) and final report showed embryonal rhabdomyosarcoma (desmin positive, myogenin focally positive and cytokeratin negative). Based on these findings, we arrived at a final diagnosis of embryonal rhabdomyosarcoma of the liver.

This patient was discussed in the multi-disciplinary meeting. Based on the decision, the patient was started with chemotherapy VAC (vincristine, actinomycin D and cyclophosphamide) which was stopped after 3 cycles due to evidence of disease progression. Then six cycles of MAID (mesna, doxorubicin, ifosfamide, dacarbazine) were given and again there was progression of the disease seen with ultrasound study. Then he received gemcitabine-paclitaxel chemotherapy with variable
dose, which was continued till disease progression, confirmed on examination and CT scan. Patient had a stable disease for about a year on this regimen. Subsequently his disease progressed and he died of rapid progression at 31 months from the date of diagnosis.

**DISCUSSION**

Rhabdomyosarcoma is the most common of the childhood soft tissue sarcomas, with male-to-female ratio of ~1.5:1. Gender does not appear to carry prognostic significance.

Diagnosis is most common in the childhood and adolescent years, with two peak age frequencies, at ages 2 – 6 years and in adolescence. The great majority of patients are under 10 years of age at the time of diagnosis, and approximately 5% are infants of less than 1 year. Tumours in this younger age group are likely to be of embryonal histology. About 25% of patients are more than 10 years at diagnosis and their tumours are more commonly of alveolar histology.3

Rhabdomyosarcoma (RMS) is rare in adults, accounting for 2 – 5% of adult soft tissue sarcomas.4 Age has been identified as an independent predictor of prognosis, with children < 1 year and > 10 years having inferior survival.3 Experience with treatment of adults with RMS is limited.5,6 Compared to children, adults with rhabdomyosarcoma have an inferior outcome.7,8 Although there is evidence that when treated aggressively using paediatric-type protocols, the prognosis may be similar to that of younger patients.4 Early experience with positron emission tomography scanning indicates that this modality is a valuable component of staging.5

With this case study, we have reached to the conclusion that determination of tumour extent and management is best done with a multidisciplinary approach. An expeditious local and systemic work-up is essential because these tumours have the potential to grow rapidly. PET scan can be helpful in deciding the site of origin, to rule out metastasis and for treatment response assessment. Gemcitabine / paclitaxel regimen is most effective regimen in treatment of adults and needs further studies.

**REFERENCES**


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Figure 1: PET scan showing liver mass.