Adult Hepatoblastoma in a Female Down’s
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Abstract
Hepatoblastoma is most common primary malignant hepatic neoplasm in children. Median age at diagnosis is one year and mostly present by three years. This is a rare case report of a seventeen years female with Down’s Syndrome presented with hepatomegaly diagnosed as hepatoblastoma.

INTRODUCTION
Hepatoblastoma is a malignant tumor of embryonic or fetal hepatocyte. Mostly occur in paediatric age group and most frequent malignant primitive tumor of liver in children. It accounts about 1-4% of all primary malignancy in children. Peak incidence is in first two years of life though adult presentation reported very rarely. Many congenital anomalies and syndrome may associate with this. We present here a case of Down’s syndrome with hepatoblastoma presenting for first time at the age of seventeen years.

CASE REPORT
Seventeen years female patient admitted with complaint of low-grade intermittent fever for fifteen days associated with dull aching pain in abdomen mainly on right hypochondrium and epigastrium. She also had non-projectile vomiting for last 5 days preceded by pain in abdomen.

She was non-alcoholic, non-diabetic, non-hypertensive, having no history of jaundice, contact with tuberculosis or any major illness. She was 3rd child of elderly mother (39 years), delivered vaginally at term with spontaneous cry. At birth baby was floppy with gross hypotonia and all milestones were delayed.

On Examination
Apparent age was below the normal age. There was a small, flat face, upward slanting eyes, small nose with depressed bridge and protruded furrowed tongue with idiotic look. There was an accessory breast on right side, simian crease in hand with short stubby finger, short 5th metacarpal, clinodactyly and wide gap in 1st and 2nd toes with prominent longitudinal crease, features suggestive of Down’s syndrome (Fig. 1). There was mild pallor but no jaundice or lymph node enlargement in any group. GIT examination revealed firm, tender, huge hepatomegaly without other visceromegaly and there was no free fluid in abdomen. Apart from mild mental retardation, CNS, CVS, Respiratory and Genitourinary system examination revealed no obvious abnormality.

On Investigation
Complete blood count was normal except mild anaemia. Blood sugar, urea, creatinine, electrolytes were normal. In liver function test bilirubin was normal, alkaline phosphatase was raised, serum alfa fetoprotein was elevated, viral marker for Hep B, C was negative. Routine urine examination was normal. Chest X-ray was normal. USG abdomen showed huge hepatomegaly with multiple ill defined relatively echogenic lesion in both lobes of liver. One large hyperechoic mass measuring 11.9 X 10.4 cm was seen in anterolateral aspect of right lobe of liver. CT Scan of abdomen revealed hepatomegaly with multiple poorly marginated heterogeneously enhancing lesions seen in both lobes of liver. The large right lobe mass is diffusely heterogenous and has foci of low attenuation and shows in homogenous enhancement. FNAC followed by biopsy revealed sheets of cells having vacuolated nucleus, arranged in trabecular pattern seen amongst the liver cells, highly suggestive of foetal epithelial type of Hepatoblastoma (Fig. 2). Chemotherapy followed by surgery was planned for treatment.

DISCUSSION
Hepatoblastoma is the most common primary malignant liver neoplasm in children. Most cases occur in children younger than three years. Boys are affected twice than girls. It may be associated with congenital anomalies including Beckwith Wideman Syndrome, synchronous Wilms tumor, alcohol embryopathy, hemihypertrophy, umbilical and diaphragmatic hernia, renal anomalies, Down’s Syndrome, Gardener

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Syndrome, familial polyposis coli etc. There is an association with APC gene defect and most of them are males. Usually presented with asymptomatic enlarging abdominal swelling, failure to thrive, pain, fever, vomiting and weight loss. Pallor though common but jaundice is rare. Few may present with Ips sexual precocious puberty owing to increased ectopic HCG production. Elevated alpha feto protein (90%) parallel the course of the diseases, returns to normal following tumor resection, re elevate with tumor recurrence. Mild anaemia, moderate leukocytosis and significant thrombocytosis are common. SGOT, SGPT, alkaline phosphatase may remain normal. Mostly occur as echogenic single mass (80%) in right lobe (57%) of liver but may occur in left lobe or bilobar (27%) or multicentric. Hepatoblastoma is the malignant derivative of incompletely differentiated hepatocyte precursors. They are classified morphologically as epithelial type (56%) and mixed epithelial mesenchymal type (44%). Epithelial type subdivided into epithelial foetal (31%), foetal embryonal (19%), micro trabecular (3%), anaplastic (3%) and among mesenchymal variant, mixed epithelial mesenchymal without teratoid feature (34%) and with teratoid feature (10%). Usually it metastases in lungs, brain and bone. The prognosis is poor in multiple lobe involvement, embryonal type, decreased P27 gene expression, multifocal dissemination, AFP level < 100 or >100000 ng / ml. In localized solitary lesion surgery may be curative but in inoperable case initial chemotherapy followed by radiotherapy reduce the tumor size making surgery feasible. Liver transplantation is also encouraging. The survival rate of patients with stage I disease after resection approaches 100% but declines progressively with a 27% survival in stage IV.

CONCLUSION

Hepatoblastoma is a common malignant hepatic tumor in children and median age at diagnosis is 1 year. It mostly occurs in male but in our case female Down’s Syndrome presenting at the age of 17 years is certainly a clinical rarity.

REFERENCES