CASE REPORT

A LARGE HEPATOBLASTOMA IN AN INFANT

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ABSTRACT
A prematurely born, 11 month old male child presented with history of lump over right side of upper abdomen for four months. Hepatomegaly with palpable lump in right hypochondriac and epigastric region was noted. ALT, AST & alpha feto protein were elevated with anaemia. On ultrasonography, ill-defined hypo echoic densely calcified mass lesion with internal vascularity noted in right lobe of liver. CT abdomen shows enlarged liver with heterogeneously enhancing mass lesion with necrotic areas and dense calcifications. Histopathologically diagnosis of hepatoblastoma was confirmed.
Key Words: Paediatric; GIT; Hepatic Tumour; CT Scan

Introduction

Hepatic tumours represent 1% of the child's malignant tumours, most of them being the hepatoblastoma and hepatocarcinoma with an annual occurrence of 1.5 cases to 1 million children.[1,2] The child's hepatic tumours raise diagnosis and therapy problems, one of the reasons being their infrequency: 1-4 % of solid tumours.[3] The suspicion of child's hepatic tumour is based on history, clinical, biological and imagistic data correlated with the alpha-fetoprotein level and referred to the patient's age. The histological, immunohistochemical and molecular biology examinations emphasize certain mechanisms of oncogenic activation with prognosis and therapeutic implications.

Case Report

An 11 month old male presented with history of lump over right side of upper abdomen for four months. The patient was prematurely born, at 34 weeks of gestation, with a weight of 2480 grams. Family history was insignificant. The physical examination revealed a patient with good general status, no fever, weight 7.8 kg, height 65 cm, and pallor. The following were revealed at the abdomen examination: Hepatomegaly with palpable lump in the right hypochondriac and epigastric region, lump was 5 x 5 cm in size with relatively smooth surface without tenderness No evidence of splenomegaly.

On lab investigations anaemia (Hb 8.1 g/ dl), total count of 5400 cells /cu.mm), thrombocytosis (6,50,000 cells / cu.mm.) and mild elevated liver enzyme tests (ALT 94 u/l, AST 106 u/l) were found. The serum alpha-fetoprotein (AFP) showed high values: 426.8 ng/ml (normal values < 20 ng/ml). Serological tests for Hepatitis B, Hepatitis C and HIV were negative.

Routine abdominal ultrasonography revealed the hepatomegaly with liver span of 125 cm. There was an 8.2 x 7.6 cm sized ill-defined heterogeneously hypo echoic mass lesion in right lobe with internal dense calcification with vascularity within. No evidence of dilated intrahepatic biliary radicals. Portal vein source normal colour flow. Few enlarged lymph nodes noted at porta, pre and para aortic region. Chest X-ray did not show metastases.

On CT scan abdomen, hepatomegaly with a 101(T) x 102(AP) x 91 (CC) mm sized large lobulated soft tissue density lesion with coarse calcifications were seen involving right lobe of liver. The lesion appeared heterogeneous on plain images (average density on pre contrast image 42 HU). The lesion showed moderate heterogeneous post contrast enhancement (post contrast average density 95 HU). Washed out were noted in delayed phase. Non enhancing necrotic area was seen within the lesions. The lesion displaced celiac trunk, main portal vein and left branch of portal vein to the left side. The lesion abuts stomach and right kidney with reserved fat plains. No evidence of IHBR dilation. Portal vein and hepatic vein appeared normal. Few enlarged heterogeneously enhancing lesions were seen at porta, pre aortic, aorto-caval and para aortic region largest one 21 x 14 mm sized in at porta.

Histopathological examination showed areas of mesenchymal tissue and foci of osteoid-like material are present, together with areas of epithelial hepatoblastoma.
Based on the clinical features, investigations, imaging studies and Histopathological examination diagnosis of hepatoblastoma was made. Than patient was referred to higher center for further management.

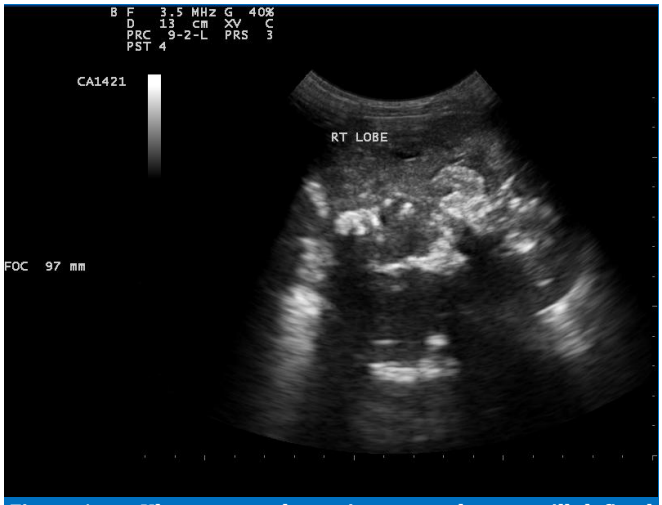

Figure-1: Ultrasonography image shows ill-defined heterogeneously hypo echoic mass lesion in right lobe of liver with dense calcifications within

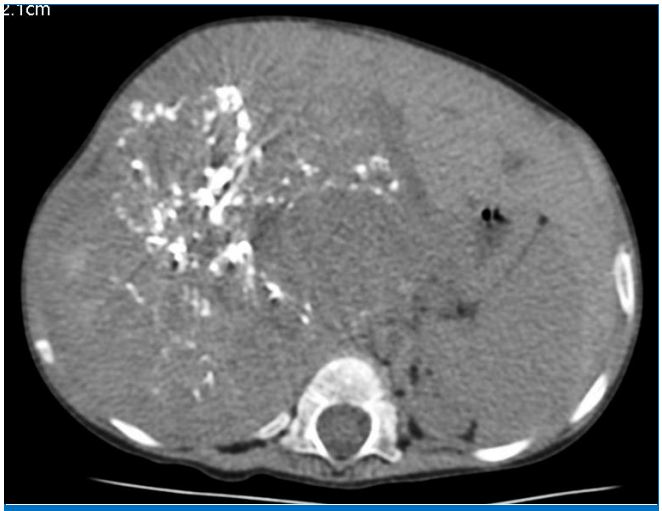

Figure-2: Axial Plain CT abdomen shows grossly enlarged liver with a 101 (T) x 102 (AP) x 91 (CC) mm sized large lobulated soft tissue density lesion with coarse calcifications & hypo dense area within in right lobe of liver. The lesion buts stomach and right kidney with preserved fat planes

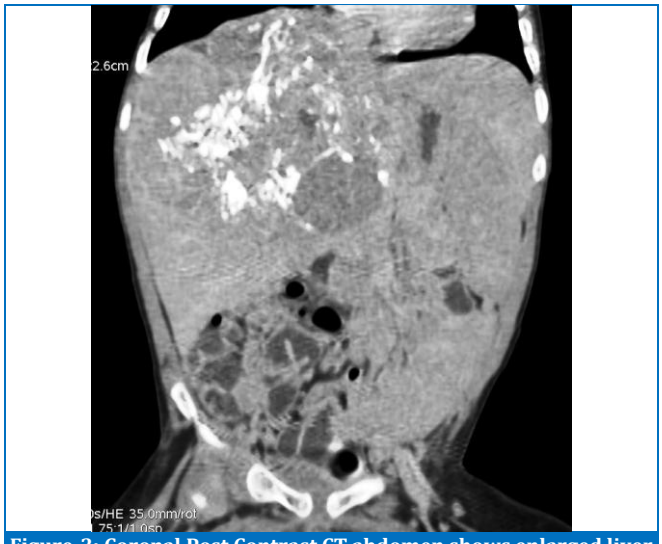

Figure-3: Coronal Post Contrast CT abdomen shows enlarged liver with heterogeneously enhancing mass lesion. Non enhancing necrotic areas are also seen within the lesion. The lesion buts stomach with preserved fat planes

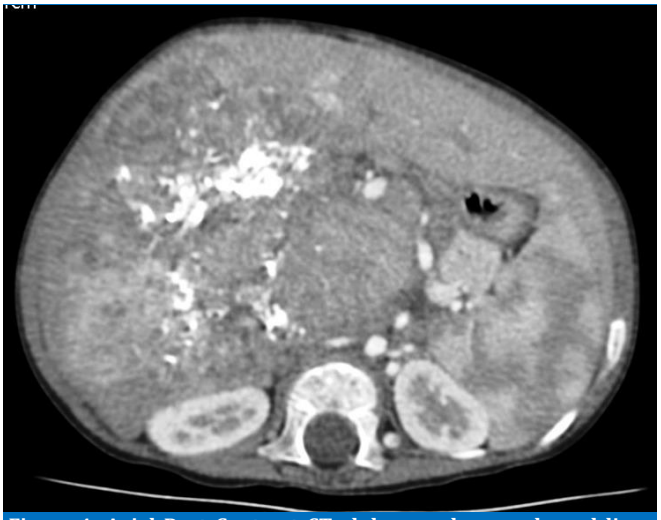

Figure-4: Axial Post Contrast CT abdomen shows enlarged liver with heterogeneously enhancing mass lesion. Non enhancing necrotic areas are also seen within the lesion. The lesion buts stomach & right kidney with preserved fat planes. The lesion displaces celiac trunk, main portal vein and left branch of portal vein to left side

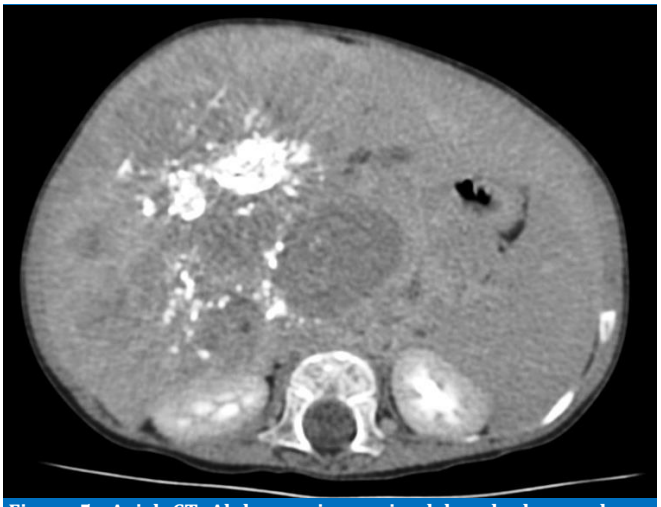

Figure-5: Axial CT Abdomen image in delayed phases shows washout of contrast

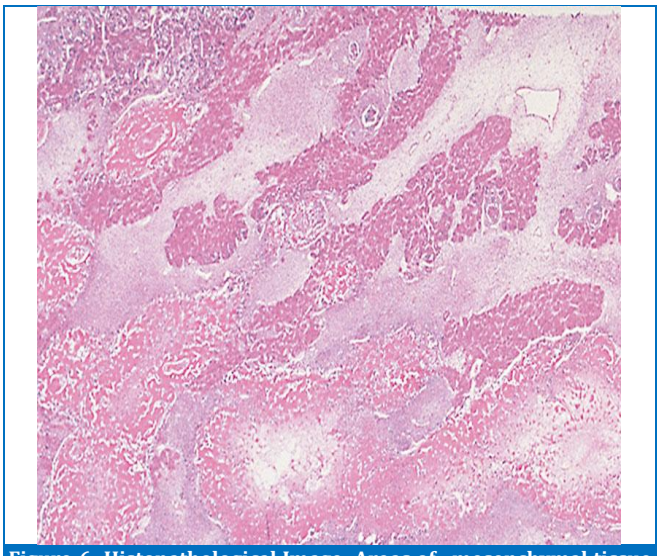

Figure-6: Histopathological Image: Areas of mesenchymal tissue and foci of osteoid-like material are present, together with areas of epithelial hepatoblastoma

Discussion

Hepatic tumours represent approximate 0.5 - 2 % of all the tumours in child, and are responsible for 1-4 % of all the solid tumours.[4] Hepatoblastoma, the most frequent malignant hepatic tumour in child, 74% according to certain studies[5], very rare, presenting less than 1 case to 1, 00 000 births. There is a slight predominance of the tumour in the males (1.4-2:1), difference detectable especially in the child under 5 years old. The diagnosis of hepatoblastoma is suspected in the patient aged between 6 months and 3 years old, in the presence of a hepatic tumour, thrombocytosis and a high level of serum AFP, this association being, almost path gnomic for the diagnosis.[6] Our patient presented all these path gnomic associations.

A high serum level of AFP should be interpreted according to age, very high values indicating hepatoblastoma.[7] Moderately high values of serum AFP can be detected in certain types of hepatoblastoma, tumour of yolk sac, hepatocarcinoma as well as in certain benign tumours (mesenchymal hamartoma, focal nodular hyperplasia and infantile hemangioendothelioma). The complete excision of the hepatoblastoma determines the decrease of the AFP serum level, which will be normalized after 4-6 weeks.

Certain authors have demonstrated the existence of a correlation between hepatoblastoma and prematurity, this representing a possible risk factor.[8,9] Most of the hepatoblastoma cases are sporadic. The hepatoblastoma occurrence in children from families with family adenomatous polyposis is 200-800 times higher than in the general population. Hepatoblastoma develops more frequently in the right hepatic lobe.[10]

The cytogenetic studies on patients with hepatoblastoma show multiple anomalies: trisomy of 20, 2, 8 chromosomes, recurrent translocations: deletion (4)t (1q;4q), rearrangements at the 1q12-21 level.[11,12] Macroscopically, the hepatoblastoma is usually a solitary large tumour, well-defined, multi-nodular, white-yellowish, with fibrous stripes, areas of necrosis and cavities.[5]

The 3-year global survival of the patients with standard risk hepatoblastoma is of 91% with the SIOPEL 2 protocol and of 53% the ones with high risk hepatoblastoma.[13]

Conclusion

Hepatic tumours represent 1% of the child's malignant tumours. The suspicion of child's hepatic tumour is based on history, clinical, biological and imagistic data correlated with the alpha-fetoprotein level and referred to the patient's age. Histopathology is confirmative however CT scan is helpful in characterization of the lesion.

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