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Presentation and outcome of hepatoblastoma: Case series from a tertiary hospital in North Western Nigeria

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Abstract

Hepatoblastoma is a rare hepatic tumour seen exclusively in children. It presents predominantly from zero – 3 years of age. Unusual presentation has been reported particularly in children above 5 years. Adolescents with hepatoblastoma presents unusually late with extensive diseases and unfavourable histologic subtypes, these are associated with poor outcome. Our study, was a case series of three, a neonate and 2 adolescents 13 and 14 years each respectively. All had hepatic masses with elevated alfa feto protein (AFP) and histologic diagnosis of hepatoblastoma. Two of the 3 case series had chemotherapy and two died while one was lost to follow-up.

Keywords: hepatoblastoma, elevated AFP, thrombocytosis, adolescent, poor outcome

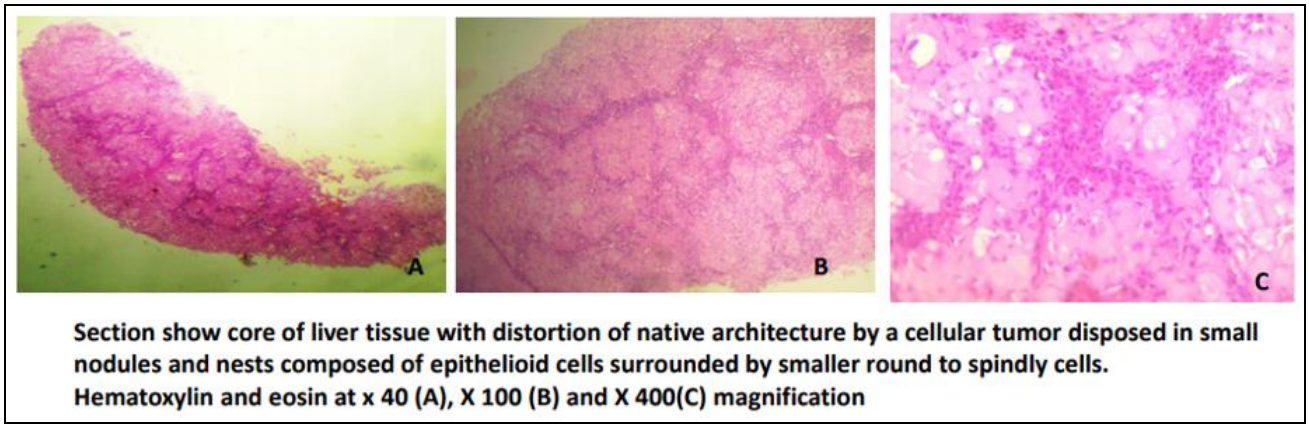
Introduction

Hepatoblastoma is the commonest primary liver malignancy in childhood [1-5]. It is a rare hepatic tumour seen exclusively in children.[3] It constitutes up to 70% of liver malignancy and 1 - 4% of all paediatric malignancy cases [6-7]. Children with hepatoblastoma presents mostly before the age 5 years, predominantly zero – 3 years [4-7]. The diagnosis may be challenging especially in poor resource settings; but should be highly considered in a child with the triad of hepatic mass, thrombocytosis and high serum alpha fetoprotein (AFP) levels [2, 7]. Unusual presentation has been reported particularly in children above 5 years [4-6]. Combined complete surgical resection and chemotherapy have been shown to improve child survival in patients with hepatoblastoma [8-9].

This study report 3 case series of children with hepatoblastoma, highlighting their clinical and morphological features as well as outcome.

Case 1

A 14-year-old girl, presented with abdominal mass for 2 years. Significant finding on examination was a liver mass. Abdomino-pelvic ultra-sound revealed an extensive hepatic mass containing both solid and cystic components. The abdominal CT showed enlarged liver with multiple ovoid hypodense masses. Biopsy of the liver done under ultrasound guidance reported hepatoblastoma mixed (epithelial-mesenchymal) type. [fig.1] Blood tests include WBC: 5.7 x 10^9/L, Hematocrit: 31%, Platelets: 511 x 10^9/L, AST: 48IU/dL, ALT:39IU/dL, AFP:163.08ng/ml. Patient was placed on cytotoxics which comprised of Cisplatin 90mg/m^2, Vincristine 1.5mg/m^2 and 5-Florouracil 600mg/m^2. However, she died 3rd day of commencement of first course of cytotoxics,



Section show core of liver tissue with distortion of native architecture by a cellular tumor disposed in small nodules and nests composed of epithelioid cells surrounded by smaller round to spindly cells. Hematoxylin and eosin at x 40 (A), X 100 (B) and X 400(C) magnification

Fig 1

Case 2

A 13-year-old boy, presented with progressive abdominal swelling for over 5 years. Significant finding on examination were massive hard hepatosplenomegaly and significant wasting. Abdomino-pelvic ultra-sound revealed grossly enlarged liver with coarsened echo texture, grossly enlarged spleen with multiple oval hypoechoic lesions and enlarged intra-abdominal lymph nodes with mild ascites.

The abdominal CT reported massive hepatosplenomegaly with multiple masses of varying sizes on the spleen. Biopsy of the liver done under ultrasound guidance reported hepatoblastoma (epithelial type) with metastasis to the spleen. Blood tests included WBC: 4.4 x 10^9/L, Hematocrit: 33%, Platelets: 177 x 10^9/L, AST: 48.3IU/dL, ALT: 45.4IU/dL, AFP:320.04ng/ml. He had four successful courses of cytotoxic drugs including cisplatin 90mg/m^2, vincristine 1.5mg/m^2 and 5-Florouracil 600mg/m^2 with regression in tumour size before he was lost to follow up.

Case 3

A two-hour old female term neonate presented with abdominal swelling. Clinical findings on examination were respiratory distress, an axillary temperature of 37.8˚C, abdominal distension with a palpable mass in the right hypochondrium extending into the right lumbar region, measuring 12cm below the right coastal margin, firm, non-tender. Abdominal ultrasound showed hepatomegaly, with masses of varying sizes in both lobes of the liver, extending to the paravertebral region, compressing the spleen, in addition, an oval hypoechoic mass was demonstrated in the left hemi thorax posterior to the lower lobe of the left lung with calcifications. Abdominal CT- scan showed a markedly enlarged liver occupying the right and left hypochondria extending into the right iliac fossa and pelvis. AST=31IU/L, ALT= 15IU/L and markedly elevated serum alpha fetoprotein of 500,000 ng/ml. PT and PTTK were prolong. Cytology of liver mass showed cellular smears with few cell clusters characterized by pleomorphic nuclei with uneven chromatin pattern and scanty cytoplasm, the tumour cells had a characteristic pseudo rosette formation in keeping with a small round blue cell tumour in favour of hepatoblastoma. Patient succumbed to the illness prior to commencement of cytotoxics.

Discussion

Hepatoblastoma is the commonest primary liver malignancy in childhood and majority of cases present in the first 3 years of life. It is rare above 5 years and the occurrence of hepatoblastoma above 5 years is insignificant [3].

In this series, 2 cases we had were adolescents, aged 13 and 14 years each respectively. They presented late, the time interval from onset of the illness and presentation to our facility was prolong (in years). This similar unusual and late presentation were earlier reported [4-6, 11]. The 3rd case was the 1st reported case of neonatal hepatoblastoma in our centre [12]. A good number of cases occurring in newborn were earlier studied [9, 13-15].

Some studies reported no significant variations in the incidence of hepatoblastoma between different nations [3, 10].

Hepatoblastoma is more prevalent in males and its predisposition increases in children with Beckwith–Wiedeman, Familial Adenomatous Polyposis, Trisomy 18 as well as very low birth weight, birth weight less than 1.5Kg [2, 5-7]. Other association that increases the risk of developing hepatoblastoma incudes tyrosinaemia, galactosaemia and glycogen storage disease type 1 [16]. It is of note that none of our cases apparently had any of the aforementioned identifiable syndromes or risk factors.

All our patients presented with abdominal distension with palpable hepatic masses which are usual findings in hepatoblastoma.[4] In addition, our second case presented with significant weight loss a feature of malignancy unlike other reported cases with apparently normal weight.

Hepatoblastoma is classified by histology as epithelial (56%) or mixed epithelial/mesenchymal (44%), epithelial hepatoblastoma is further divided into pure fetal, embryonal, macrotrabecular and small cell undifferentiated types [17-19]. All our 3 cases had biopsy for histology except the neonate that had cytology due to deranged clotting profile. The histology reports confirmed hepatoblastoma of epithelial and ephitheal-mesenchymal (mixed) types each respectively. The most useful tumour marker for screening and diagnosis of hepatoblastoma is alpha feto-protein (AFP) [4, 11]. It is also used as a bio-marker to monitor the treatment successes and tumor recurrence, the normal AFP levels are lower than 50 ng/ml in children [4]. The alpha feto-protein results in this series were elevated but markedly elevated in our 3rd case. High values of AFP in hepatoblastoma may suggest massive tumoral extension, and /or presence of metastasis signifying an unfavourable prognosis [2]. Low levels of AFP < 100ng/ml is often encountered in small cell undifferentiated subtypes of hepatoblastoma, because of the decreased number of hepatocytes that secrete this protein. The low level of AFP is also identified as high-risk subgroup with extensive disease at diagnosis, poor response to therapy and poor survival which may suggest poor prognosis [2, 6, 20].

The combination of cytotoxics and surgical resection yield favourable outcome. In this series two of the cases had cytotoxics, up to 4 courses in one of the cases. Two of the cases died while one was lost to follow-up. The lost to follow up case may be due to financial constraints or might have died at home unreported. The 3 cases were also planned for surgery after neo-adjuvant chemotheraphy but none benefit from that. All our subjects had clinical evidence of extensive disease. The extensive nature of the disease, late presentation and probably high-risk histologic subtypes might have significantly contributed to the mortality of our patients.

Conclusion

Hepatoblastoma may presents rarely in children older than 5 years; and should be suspected in the presence of hepatic mass, elevated serum alpha fetoprotein and thrombocytosis. Late presentation and unfavourable histology type may contribute to high mortality. There is need for more studies to evaluate risk factors and treatment outcome of hepatoblastoma in our community.

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