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# The childhood hepatoblastoma - case report

## Abstract

Hepatoblastoma (Hb) is a rare Pediatrics cancer. The incidence is low and diagnosis very difficult, especially in low income countries where the diagnostic tools may not be readily available and accessible. Here we reported a 30 months (3years and 6months) old male child that was referred with symptoms of 3 month history of recurrent cough, fever, abdominal mass with distention. Child was treated for 8 weeks with anti-tuberculosis therapy with no clinical improvement, hence referred for an expert care. Child was assessed on presentation in our center and immediate work-up investigation results were in keeping with childhood Hb. Immediate definitive medical treatment was commenced to which child responded promptly with resolution of the symptoms and steady decrease in Alfa Fetoprotein level (AFP). Child was discharged and currently on follow-up visit.

Timely diagnosis and intervention of childhood Hb is the hall mark for good prognosis. Need to create awareness of this case in our sub region as it will help to instill the practice of having a high index of suspicion of Hb in any child with abdominal mass and high serum AFP, especially among the clinicians in primary care level. This will enhance prompt referral for proper intervention. This practice will guarantee the needed good prognosis.

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## Introduction

Hepatoblastoma (Hb) is a rare Pediatrics solid tumor arising from the liver.[1,2] Though Hb is uncommon Pediatrics solid tumor, it is the most common form of liver cancer in children.[3] Hb is largely an asymptomatic abdominal mass known to be sporadic with unknown aetiology.[1,2] Some cases of Hb is seen in patients with some genetic syndromes like Goldenhar syndrome, Soto syndrome, Down's syndrome, Beckwith-Weidemann syndrome, hemihyperplasia and some congenital malformations like cleft palate, tetralogy of fallots, renal anomaly, dysplasia of the earlobes etc.[1,4,3] Children with these syndromes are known to have genetic predisposition to develop Hb. Furthermore, low birth weight, prematurity and infection with Hepatitis B virus are risk factors of childhood Hb.[3] Hepatoblastoma originates from immature precursor liver cells, it's usually unifocal affecting right liver lobe more commonly than the left lobe.[2,3] Hb occurs in children within 3 years of age and about 40% of the cases are diagnosed in advanced stages as an asymptomatic abdominal mass.[2] About 20% of the cases already have pulmonary metastasis at diagnosis.[2,3] Childhood Hb occurs in white children 5 times more frequently than black children, though black children have worse outcome when it occurs.[3] Male are more affected than females in the ratio of 1.7:1. Hb is very rare in adolescents and adults with worse prognosis when it occurs.[3]

Diagnosis is made when a child develops abdominal liver mass with markedly elevated alpha-fetoprotein (AFP) level.2 Biopsy of the liver mass is necessary to establish the definitive diagnosis.[2] Hb exist as different histologic types like Epithelial, mesenchymal and mixed or undifferentiated. Subtypes of the epithelial histologic types includes embryonal, fetal, pleomorphic, anaplastic, small cell undifferentiated and cholangioblastic macrotrabecular.[5] Other diagnostic investigations that can be done includes abdominopelvic ultrasound scan, Abdominal CT scan and MRI.[2] This will enhance in pretreatment extent of disease (PRETEXT) staging of the Hb disease. PRETEXT stage Hb liver mass into stages one to four depending on the number of liver lobes that are involved at diagnosis.[3]

Cases of Hb must be referred to a tertiary center for ideal management. This is because proper management of childhood Hb needs multi-disciplinary specialist care ranging from Pediatrician/ Pediatrics oncologist, Pediatrics surgeons, Anaesthetics, Radiologist, etc for proper care.[6] The Pediatric Oncology group in the United States and the International Pediatric Liver Oncology strategy group in Europe recommend surgery combined with chemotherapy as the standard treatment of childhood Hepatoblastoma.[2,7] Combination of these treatment modalities can be done depending on the patients response to initial treatment and when there is re-occurrence of the tumor after a tumor free survival.[2,3] The Chemotherapy usually used includes Cisplatin, Vincristine, 5-fluro-uracil or doxorubicin.[2] The second line chemotherapy that can also be used includes Carboplatin, Fosfamide, etoposide and Irinotecan. Other treatment approaches include cryoablation, radiofrequency ablation, Radiotherapy, and transarterial chemoembolization.[1,2,8] Tumor free survival from childhood Hb can be achieved by combining chemotherapy and surgery. Non-responders to chemotherapy and those children with non-resectable tumor can be treated with liver transplantation with post transplantation chemotherapy.[8,9] Some non-resectable tumor has become resectable after neoadjuvant chemotherapy and survived after post resectional chemotherapy.[10]

Childhood Hb has 90% survival rate after chemotherapy.[2] Five year survival rate after treatment is about 57-69%.[3] Prognosis of childhood Hb is based on level of AFP, PRETEXT stage and Evans surgical Stage of the disease at diagnosis.[4] rapid reduction of AFP after chemotherapy and surgery shows good prognosis, while persistent high level of AFP or recurrent tumor after initial remission shows poor prognosis.[9,11] Most mortality of childhood Hb is found in cases of recurrent Hb after liver transplantation or liver lobectomy.[9]

Patient that have remained symptom free after 5 years of treatment are considered cured, though long term monitoring of the impact of chemotherapy used on development, growth and organ toxicity is very essential.[3] Hence proper follow-up should be done for the survivals up to 10 years and beyond.

## Case report

A 30 months old male child was referred for an expert care with 3 months history of recurrent cough, fever, weight loss and abdominal mass with distention. Remarkable findings on general examination revealed acute on chronically ill looking male toddler in obvious respiratory distress evidenced by flaring of alae nasi, intercostals and subcostal recession. There was mild pitting pedal oedema bilaterally. Child had moderate pallor, anicteric, acyanosed, afebrile with temperature of 37.0o C with no signs of dehydration. Child weighed 16Kg, height of 108cm and body surface area of 0.70/m2 which are within normal limits. Further systemic examination revealed bibasilar crepitations and rhonchi, dull chest percussion note on the mid and lower lung zones. Heart sounds I and II with gallop rhythm was heard. There was massive ascites demonstrable by fluid trill. The liver mass was palpable per abdomen measures about 8cm below the right costal margin, smooth surface and non-tender. The child was fully conscious and well oriented in time place and person, though moderately irritable. The initial investigations done at Primary health care center before referral were; Full Blood Count (FBC) which revealed lymphocytosis and slight leucopenia. The peripheral blood film shows normochromic normocytic red cells with few hypochromic red blood cells, some bite cells and numerous burr cells. Child's hemoglobin genotype is AA, urine microscopic /culture/sensitivity yielded no growth after 48 hours of culturing. Total serum protein and albumin were within normal range. The serum electrolyte urea and creatinine were normal. Tuberculosis screen test like moutoux test, geneXpert, AFBx3 yielded negative results, screening results for hepatitis B, C and HIV were all negative. The initial chest X-ray done shows massive right pleural effusion with mediastinal shift to the left, the rib cage and cardiac size were normal. The initial abdominal ultrasound done showed liver mass with span of 11.3cm, ascites with no other intra-abdominal mass lesion. Child was further treated for suspected military tuberculosis with sepsis using anti-tuberculosis therapy and antibiotics for 8weeks at the Primary health Center, with worsening symptoms child was then referred to our center for an expert care.

On presentation to our center, the liver function test done showed markedly elevated Alfa Fetoprotein (AFP) of 718iu/ml (<20.0iu/ml) with elevation of all the liver enzymes, alkaline phosphatase and serum total bilirubin. Repeat abdominal ultrasound shows increasing liver span up to 16cm below the right coastal margin, smooth surface with normal echotexture. Other intra-abdominal organs; kidney, spleen, gall bladder and pancreases were normal with normal echotexture and parenchymal differentiation. There was ascites with no focal intra-abdominal lesion. Thyroid function test done were within normal range. Echocardiography done showed an extra-cardiac arising from the right lung lobe impinging on the right ventricle, this mass measured 4.5cmx2.8cmx1.5cm. Further echocardiographic findings showed enlarged right atrium secondary to pulmonary hypertension. There was diastolic dysfunction of the right ventricle. Chest and abdominal CT scan showed asymmetry of the lung fields with interlobular septal thickening and consolidation on the right lung field, mild pericardial effusion, pleural effusion and hepatomegaly. The biopsy of the liver nor of the lungs was done since child was clinically unstable for the invasive procedure.

The diagnosis of hepatoblastoma with metastasis to the right lung lobe was made. Child was placed on definitive 6 course chemotherapy, three weekly interval using intravenous Cisplatin, Vincristine and 5-flurouracil. Child was also placed on supportive medications with antibiotics, antiviral and diuretics medications. Child had marked clinical improvement after the 2nd course of the chemotherapy, as the pleural effusion and ascites regressed, the liver span became normal not palpable per abdomen. And child was clinically stable as the respiratory rate and pulse rate became normal. Child has completed the 6 course chemotherapy and currently on follow-up. The repeat liver function test result were normal with marked reduction in AFP level. Repeat echocardiography showed absence of the right lung mass with normal systolic and diastolic function of both the right and left ventricles. Liver span has remained normal since on follow-up and no signs of organ toxicity following the chemotherapy use has been reported since on follow-up.

## Discussion

Hepatoblastoma (Hb) is considered a rare heterogeneous tumor arising in an otherwise healthy liver mostly the right lobe with metastasis commonly to the lungs.[1,6] Incidence of childhood Hb is about 1.5cases/million population, though currently, the incidence has been on steady increase over the past 30 years.[12] Childhood Hb is rare accounts for 1% childhood malignancy, though it is the most common childhood primary liver tunour.[12]

Here we reported a case of hepatoblastoma with onset at 30months (3 years, 6months) age, which is within the standard age of onset of 3-5years of childhood Hb.[2,3,12] Though in contrast some cases of hepatoblastoma has been reported in the neonatal period,[13] adolescents and in-utero at 30 weeks gestation.[7,14] This shows that childhood hepatoblastoma is sporadic and can occur in any age group especially when a child with genetic predisposition to develop Hb is exposed to the known risk factors of Hb. Other known risk factors of childhood Hb includes poly or oligo-hydramnions, high maternal pre-pregnancy weight, child of a woman treated for infertility, smoking/tobacco use, and parental occupational exposure to heavy metals.[11] Hence clinicians should have a high index of suspicion for childhood hepatoblastoma in any child with liver mass and high AFP level.

The index case had high level of AFP with metastasis of the tumor cells to the lung, this is same with many cases of hepatoblastoma at diagnosis.[1-3] This findings is in contrast to the discovery of some researchers. A cases of childhood Hb with low AFP at diagnosis has been reported.[15] Another case of high plasma rennin activity , hypertension and normal aldosterone level has been reported in a child diagnosed with Hb.[16] This contrasting reports shows that childhood Hb cells can be involved in excessive production of other hormone like rennin and not just the widely known AFP. This can also affect different signs and symptoms that can be obtained in a child with Hb. These hormones produced in excessive levels by the Hb cells can be used as markers for clinical response to therapy and prognosis ultimately. Clinicians should note that AFP is not the only hormone that is produced in excess by Hb cells, other hormones like rennin and any other hormone can be produced in excess in a child with Hb. Though no other hormones was noted to be produced in excess by the index case report.

This reported case had marked reduction in serum AFP level after the 3rd cycle of chemotherapy. Similar finding was obtained in most reported cases of childhood hepatoblastoma.[2,3] This reduction in AFP has been known to be associated with good prognosis as in the index case. Furthermore the reported case of neonatal Hb also had good prognosis with standard surgical and medical interventions with reduction in serum levels of AFP.[14] The index case had no toxicity with the used chemotherapy. This is in contrast with reported retinal toxicity observed by *Hillard.*[17]

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Though child is still on follow-up and monitoring to ensure adequate growth and development. The managing team is also engaged in serial investigation of the index case for early detection and handling of any long term organ complications that may arise.

## Conclusion

Childhood Hb is rare and involves multi-specialist care when it occurs. It has been shown to occur in any age group in genetically predisposed patients exposed to the risk factors and or some patients with genetic syndromic condition. AFP is produced in excess by Hb cells more often though currently Hb cells has been shown to produce renin in excess. Hence there could be propensity of producing other serum hormones in excess by Hb cells. Therefore Physicians as well as the surgeons should be acquainted with these known current dynamism associated with childhood Hb. Finally a very high index of suspicion is required for prompt diagnosis and management to enhance good outcome.

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## Conflicts of interest

The authors declare no conflict of interest.

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