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Research Paper

Challenges Encountered In the Management of Congenital Hepatoblastoma in a Resource Constraint Setting

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ABSTRACT

Background: *Malignancies such as congenital hepatoblastoma in neonatal period are uncommon and are often characterised by unknown aetiologies.*

Aims:*We present the first case diagnosed in our neonatal unit in over two decades of the existence of our tertiary institution. This report is aimed at raising awareness of an unfamiliar neoplasm in our resource poor setting and highlight the challenges encountered in making a diagnosis.*

Case presentation: *A 2-hour old female neonate presents with abdominal swelling, eye discharge and refusal to cry since birth.A palpable abdominal mass was felt in the right hypochondrium extending to the epigastrium and right lumbar region, hard, measures 12cm below the right subcoastal margin. The first two abdominal ultrasounds results showed normal findings. Nevertheless, a third abdominal ultrasound showed hepatomegaly, with masses of varying sizes in both lobes of the liver. Abdominal CT- scan showed a markedly enlarged liver.A triple phase abdominal CT and an MRI were requested but were not done due to financial constraints.*

The baby had an elevated serum alpha fetoprotein of 500,000 ng/ml. FNAC of liver mass showed tumour cells with characteristic pseudo rosette formation, in keeping with small round blue cell tumour. Excisional biopsy was contraindicated in our patient as she had a deranged clotting profile. Great importance is given to immediate burial of the dead and though consent was sought for excisional biopsy for autopsy, the care givers did not oblige.

Conclusions:*Hepatoblastoma is an uncommon tumour in our environment but should be suspected in every neonate presenting with abdominal distension. Government policies that support post-mortem autopsies should be put in place in settings such as ours, as this may help in knowing the true burden of congenital hepatoblastoma.*

KEYWORDS: *Congenital;Hepatoblastoma; Neonate; Nigeria.*

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I. INTRODUCTION

Malignancies are often under-reported in sub-Saharan Africa probably as a result of absence of cancer registries, weak health systems, overwhelming burden of infectious diseases and the poor survival outcomes of cancer patients in resource constraint settings like Sokoto, North-western Nigeria [1]. Herein, we present a rare case of congenital hepatoblastoma and the diagnostic challenges we encountered in management.

II. CASE REPORT

A two-hour old female neonate referred to our level one neonatal unit, located in a referral teaching hospital in North Western Nigeria with abdominal swelling, eye discharge and refusal to cry since birth. There was no vomiting and neither was there a delay in passing meconium.

The baby was born at term via emergency Caesarean section on account of prolonged second stage of labour. Though APGAR scores were not provided in the referral note, we were informed that the baby did not cry immediately after delivery and had to be resuscitated by bagging and tactile stimulation. There was no history to suggest maternal febrile illness and orchorioamnionitis.

The mother, a nineteen-year-old primigravida with no formal education attended a minimum of three ante-natal visits in the index pregnancy with no abnormality detected. She received no orthodox or traditional medications apart from her routine ANC medications and had no known prior exposure to ionizing radiation. She was neither diabetic nor hypertensive.Father is a 30-year-old trader with no formal education. Marriage is consanguineous.

On clinical examination she was found to weigh 3.2 kg, with a length of 50 cm and an occipito-frontal circumference of 36cm. She was in respiratory distress, and had an axillary temperature of 37.8˚C. The abdomen was distended more in the area proximal to the umbilicus, it was soft, but abdominal organs were difficult to palpate. Bowel sounds were present and normoactive.

Two serial abdominal ultrasound examinations done for the babytwenty-four hours apartreported normal findings.

A diagnosis of probable intestinal obstruction to rule out necrotizing enterocolitis in a neonate with historical perinatal asphyxia and presumed sepsis was entertained. Surgical consultation was sought and she was placed on nil per oral, nasogastric tube for decompression and commenced on empirical antibiotics while we awaited results of electrolytes, urea and creatinine, blood culture and full blood count and differentials.

On the 3rd day of life she was noticed to be icteric with an increase in abdominal girth (Figure 1) measuring 37.5cm, 6cm from the xiphisternum with visible veins draining upwards, a palpableabdominal mass felt in the right hypochondrium extending to the epigastrium andright lumbar region, hard, irregular, measures 12cm below the right subcoastal margin, non-tender, no bruit, bowel sounds were present and normal.Digital rectal examination revealed good sphincteric tone, an empty rectum with examining finger stained with meconium.Other systems examination revealed normal findings.

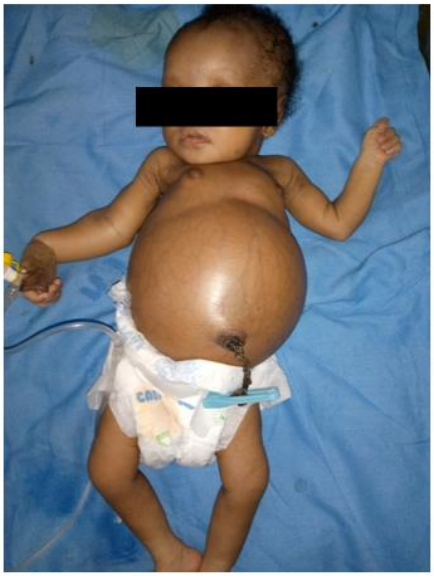

Figure 1: Index case with gross abdominal distension

The first two abdominal ultrasounds results showed normal findings. Nevertheless,a third 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. The biliary tract was normal; Thus, differential diagnosis of neuroblastoma and lymphoma were entertained. Abdominal CT- scan showed a markedly enlarged liver occupying the right and left hypochondria extending into the right iliac fossa and pelvis, the intrahepatic vascular channels and biliary ducts were normal. No abnormal radio-opacity was seen. A triple phase abdominal CT and an MRI were requestedby the radiologist but were not done due to financial constraints.

Our index patient had blood transfusion on account of a hematocrit drop of 14% observed over a period of two weeks. Total serum bilirubin was 18.8mg per dl with a conjugated fraction of 4.84mg per dl.Liver function test showed elevated liver enzymes with an AST value of 31u/L and an ALT of 15u/L. She had an elevated serum alpha fetoprotein of 500,000 ng/ml.

The patient had intravenous fluid, antibiotics, oxygen and vitamin K. Onaccount of the inconclusive imaging studies, a review by paediatric gastroenterologist and histopathologist necessitated an ultrasound guided fine needle aspiration cytology of liver mass which showed a field of moderately cellular smears with few cell clusters characterised as having moderately pleomorphic nuclei with uneven chromatin pattern and scanty cytoplasm, the tumour cells had a characteristic pseudo rosette formation, is in keeping with a small round blue cell tumour in favour of hepatoblastomaSee figure 2A&B below.

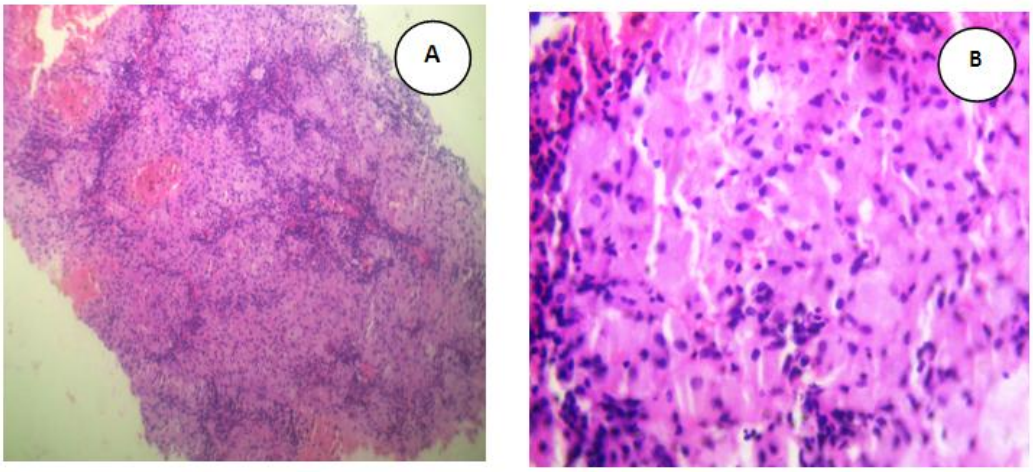

Figure 2 A: Section shows core of tissues composed of malignant hepatocyte. (H & E X40), B: Section shows malignanthepatocyte. (H & E X200)

However, prior to starting therapy, the patient succumbed to the illness. All efforts aimed at convincing the parents to agree to a post mortem biopsyfailed.

III. DISCUSSION

Neonatal hepatoblastoma is a rare tumour, our index case being the first to be diagnosed in the over two decades of the existence of our tertiary institution. Our patient is a female in keeping with the report by Wabada*et al,*[2]where the two reported cases from Nigeria were females, this contrast reports by Thomas *et al*[3] and Comert*et al*[4] who reported males.

The aetiology of hepatoblastoma is not fully known. There are evidences to suggest hepatoblastoma is derived from pluripotent stem cells, this could be due to developmental error during hepatogenesis.[5] The age of the mother in this report is 19 years old, this is comparable to the report by Wabada*et al*[2] where the mothers were aged 18 and 19 years respectively. While in another study, the maternal age reported was 28 years old.[4]Despite a dearth of reported cases of congenital hepatoblastoma in Northern Nigeria, the few reported cases in literature reviewed highlight the fact that it is commonly found among infants of young mothers, we hypothesize that this may be due to exposure to an unknown carcinogen, this is because there exists in our setting a cultural practice of using untested sex enhancers in form of herbs '*kayanmata'* in this age group.[5]

Anidentified risk factor for the disease observed in this patient was perinatal birth asphyxia, our patient did not cry at birth and was unable to initiate and sustain respiration at birth. Though it might be reasonedthat congenital hepatoblastomaas a morbid condition in our case might have affected the interpretation of our patients APGAR scores at birth.

As reported, imaging studies were unhelpful in making the diagnosis. Thus, the progressively increasing abdominal distension was pivotal in making a decision to suspect malignancies like: Hepatoblastoma, malignant infantile hemangioma with compartmental syndromes, stage IV neuroblastoma, and giant cell tumor.[6]AFP served as a very useful laboratory marker for the diagnosisthough it is not specific, as levels of AFP can be increased in hepatitis, liver cirrhosis, testicular tumours, germ cell tumours, hepatocellular carcinoma, gall bladder carcinoma and hemangio-endotheolioma.[7]

Being in a resource constraint setting characterised by out-of-pocket spending also limited other diagnostic tests. In addition, excisional biopsy was contraindicated in our patient as she had a deranged clotting profile and we had to make do with cytology. These diagnostic delays led to a non-initiation of chemotherapy and possible surgical resection of the mass, thismay have contributed to the poor outcome observed,as it iswell-known that early commencement of treatment improves survival.[4,8]

In the Muslim dominated Northern part of Nigeria, great importance is given to immediate burial of the dead and though consent was sought for excisional biopsy for autopsy, the care givers did not oblige. [9]

IV. CONCLUSIONS

Hepatoblastoma is an uncommon tumour in our environment but should be suspected in every neonate presenting with abdominal distension. Out of pocket payment systems as epitomized inresource constraint settings like Sokoto,North western Nigeriaresult in a delayof diagnosis and can lead to poor outcomes. Free or subsidised services for diagnosis and management of neonates with malignancy is highly recommended. Detailed check-ups during pregnancy may help in early diagnosis of this tumour. Government policies that support post-mortem autopsies should be put in place in settings such as ours, as this may help in knowing the true burden of congenital hepatoblastoma. Finally, we propose a more methodological robust study to investigate the possibility of a causal relationship between using untested sex enhancers in form of herbs and the occurrence of congenital hepatoblastoma

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Authors contribution:

1. Adamu Asma'u, Design the study, literature search, and final draft of the manuscript
2. Onankpa BO, Gave the necessary guidance and added intellectual content.
3. Jega Muhammad Ridwan, literature search and final draft of the manuscript
4. AtusuAlshatu-Yasmine Alethea, collected data and wrote the first draft of the manuscript.
5. Mohammed Umar, literature search and revision of manuscript
6. Jibril Baba, literature search and revision of manuscript
7. YunusaGarbaHaruna, Gave the necessary guidance and added intellectual content.
8. Musa Nasiru, wrote the first draft of the manuscript.

All authors read and approved the final manuscript.

Consent: Written consent was obtained from the parentsof the patient.

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