

Case Report

Neonatal neuroblastoma in a tertiary health facility in Southwest Nigeria: a case report

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ABSTRACT

Neuroblastoma is a pediatric neoplasm diagnosed in infancy and occurs mainly in the abdomen. There is a paucity of data in developing countries, including Nigeria. Therefore, this study aimed to report the first case of neonatal neuroblastoma in a Federal Medical Centre, Owo, Ondo state, Nigeria. We described a 4-hour-old term female neonate with multiple subcutaneous nodules and a right-sided intra-abdominal mass that crosses the midline. Examination reviewed an ill-looking neonate with multiple, painless anterior abdominal wall subcutaneous nodules and abdominal distension with a palpable right-sided abdominal mass. The abdominal MRI revealed a right abdominal mass separated from the normal-shaped kidney and an enlarged liver with multiple nodules. The liver enzymes and urinary vanillylmandelic acid were elevated. Neonatal neuroblastoma, though rarely reported among Nigerian neonates, should be considered in the differential diagnosis of abdominal mass, and the awareness should be increased among sonographers for early detection.

Keywords: Abdominal mass, Neonatal neuroblastoma, Nigeria

INTRODUCTION

Neuroblastoma is an embryonic group of tumors known as 'peripheral neuroplastic tumors' which includes ganglioneuroblastoma, ganglioneuroma, and ganglioneuroblastoma-nodular that arises from the sympathetic ganglia.^{1,2} Neuroblastoma is one of the most common neonatal malignancies which accounts for 28-39% of all malignancies in the neonatal period.³

It occurs as a result of the neural crest cell transformation from genetic or epigenetic events, though, the exact event of the tumorigenesis is not clear.¹ Neonatal neuroblastoma primary sites include the adrenal glands (40%), the abdominal ganglia (25%), thoracic ganglia (15%), pelvic ganglia (5%), cervical sympathetic ganglia (3%–5%), and

paratesticular region (1%).^{1,4} The location of the primary sites reflects the pattern of migration of the neural crest cells during fetal development.^{1,4} Neonatal neuroblastoma has a good prognosis with a survival rate of 88-91% and the tumor can regress spontaneously despite its malignant nature.^{1,5,6}

We reported a 4-hour-old female neonate who presented with stage 4S neuroblastoma which was the first case of neonatal neuroblastoma presenting in our health facility in the past 20 years.

CASE REPORT

Patient was a 4-hour-old term female neonate who presented on account of abdominal distension and multiple

subcutaneous nodules at birth. There was no vomiting and has passed meconium. He had multiple nodules on the trunk, groin, and upper thigh. The mother had antenatal care at a peripheral hospital and a normal early obstetric ultrasound.

The baby was delivered at term through spontaneous vaginal delivery (SVD) to a non-consanguineous family. Birth weight was 3200 g. The patient is the last of three children and no similar symptoms in other siblings.

Physical examination revealed an ill-looking neonate not in respiratory distress and multiple subcutaneous nodules. The abdomen was distended with a girth of 37 cm (7 cm from the xiphisternum), a nodule on the anterior wall with a palpable intra-abdominal mass extending from the right hypochondria to the right iliac region inferiorly crossing the midline.

The mass was firm, not tender, attached to the underlying but not to the overlying structure, it was possible to get above, and below it (Figure 1).

The skin examination revealed multiple painless subcutaneous nodules on the anterior abdominal wall, back, inguinal region, groin, and upper thigh which were firm, not tender, had no differential warmth, and were mobile with the largest over the groin measuring 5 by 6 cm.

Other systemic examinations yielded no additional abnormal findings. Based on the presentation and intrabdominal mass, a diagnosis of neonatal neuroblastoma was considered.

The investigation result showed an abdominal ultrasound that revealed a huge well-defined oval-shaped heterogeneous vascular mass in the right hypochondriac and lumbar region precisely on the right renal bed measuring 7.8 cm by 4.8 cm.

The abdominopelvic magnetic resonance imaging (MRI) showed an 8.8 by 6.3 cm (L by AP) heterogeneous mass in the right paravertebral region and suprarenal space displacing the right kidney anteriorly and laterally, both kidneys were normal in size, shape, and structure (Figure 2).

The liver was enlarged measuring 17.6 cm span with multiple enhancing nodules (Figure 3). There was an elevated liver enzyme and anemia for which the patient was transfused. The biopsy of the intra-abdominal mass was not done because of the suspicion of high vascularity of the tumor.

The urine vanillylmandelic acid (VMA) was elevated but the homovanillic acid (HVA) was normal. The result of the investigations is shown in Table 1. A diagnosis of neuroblastoma with skin and liver metastasis (stage 4s) was made.



Figure 1: Photograph of a female neonate with abdominal distension and multiple nodules.

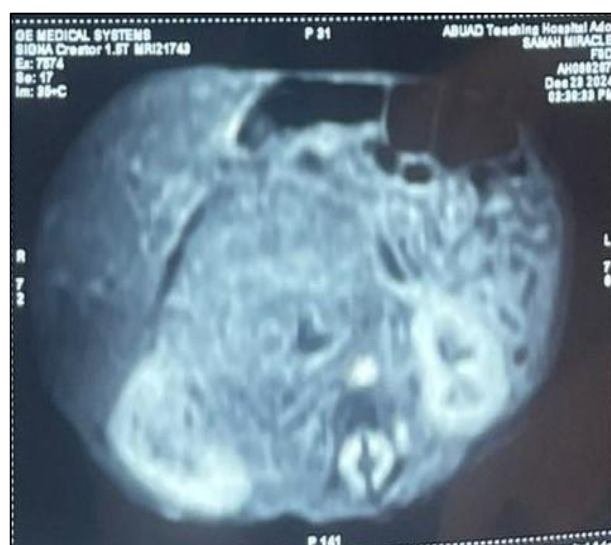


Figure 2: The magnetic resonance imaging (MRI) showing the kidneys with the tumour.

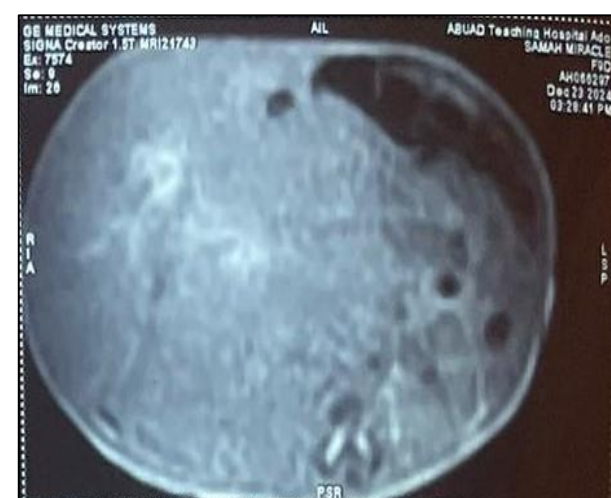


Figure 3: The magnetic resonance imaging (MRI) showing the liver metastasis.

Table 1: Result of investigations.

Liver function test	Result	Normal value
Bilirubin total (µmol/l)	60	<20
Conjugated bilirubin (µmol/L)	32	<5
Alkaline phosphatase (I.U/l)	162	
Aspartate Transaminase (I.U/l)	134	<16
Alanine transaminase (I.U/l)	24	<22
Total protein (g/l)	114.3	55-80
Albumin (g/l)	40.6	35-50
Renal function test		
Bicarbonate (mmol/l)	25	20-30
Chloride (mmol/l)	102	95-110
Potassium (mmol/l)	3	3-5
Sodium (mmol/l)	134	120-140
Creatinine (µmol/l)	44	50-132
Urine test		
HomoVanillic Acid (µmol/ml)	0.7	0.5-2.5
Vanillylmandelic Acid (µmol/ml)	11	<10
Haematological value		
Packed cell volume (%)	25	
Posttransfusion packed cell volume (%)	35	

DISCUSSION

Neuroblastoma is one of the most common neonatal tumors in developed countries but is rarely reported in developing countries including Nigeria.³ Our hospital which is a tertiary health facility is reporting the first case of neonatal neuroblastoma in the past 20 years. The rarity of the disease could be due to low prevalence in developing countries or due to under-reporting of the disease. There is a need for all clinicians to have a high index of suspicion in the diagnosis of the disease.

This index patient presented with a palpable intra-abdominal mass in the right hypochondrium extending to the right iliac region which crosses the midline. Abdominal mass is the most common presentation of neonatal neuroblastoma.^{7,8} Also present in this index patient were subcutaneous nodules, a feature of neuroblastoma described as blueberry muffin spots.^{7,9} The presence of subcutaneous nodules is suggestive of disseminated disease.⁹

About 60% of cases of neuroblastoma present with metastatic lesions that occur through blood vessels or lymphatic drainage.⁹ The index patient has a liver metastasis which was evidenced in the abdominopelvic magnetic resonance imaging (MRI) and the deranged liver enzymes. The other clinical features of neuroblastoma include weight loss, unexplained fever, irritability, and failure to thrive which are frequently seen in older children.^{7,10} This index patient had severe anemia at admission necessitating a blood transfusion. The prevalence of anemia in patients with neuroblastoma is

40%.¹¹ The possible cause of anemia in patients with cancer could be infiltration of bone marrow by cancer cells although bone marrow aspiration for cytology was not done in this patient.¹⁰ Also, the release of inflammatory cytokines by cancer cells which can suppress the bone marrow, and increase hemolysis, coupled with inadequate nutrient intake could have accounted for the anemia.¹⁰ It is not uncommon for patients with cancer in Africa to have a concomitant infection specifically malaria which can also cause anemia in children.¹²

The use of obstetric scans in the early detection of neuroblastoma cannot be overemphasized as 20% of all neonatal neuroblastoma can be diagnosed antenatally through obstetric ultrasound scan.¹³ Although, this index baby had an obstetric scan for the estimated gestational age, weight, and gender but not for anomaly detection thereby having a missed diagnosis in-utero. Hence, the need for more emphasis on anomaly scans during antenatal period which is more likely to detect the lesion and possibly proffer management before delivery.⁸ Similarly, there is an increased risk of bleeding into the tumor during delivery but with antenatal diagnosis, caesarean section could be opted for as a precautionary measure.¹⁴

The role of urinary vanillylmandelic acid (VMA) in the diagnosis of neuroblastoma is very important. Studies have shown that more than 80% of patients with neuroblastoma have elevated urinary VMA.¹⁵ This was similar to the finding in this patient who had elevated urinary VMA although, the urinary homovanillic acid (HVA) was normal. Homovanillic acid is a metabolite of dopamine while VMA is a metabolite of catecholamines that are

produced by the adrenal medulla.¹ The values of the metabolites may be enough to suggest the diagnosis of neuroblastoma especially where tissue biopsy is not advisable.¹

The index neonate was diagnosed with stage 4s neuroblastoma which is a metastatic neuroblastoma presenting in infants aged <12 months with metastasis to the skin, liver or bone marrow.¹ The staging of this index neonate is because of the age of the neonate, the presence of subcutaneous nodules, and the metastasis to the liver which was demonstrated by the abdominal MRI, and the elevated liver enzymes.

The patient was subsequently referred to another tertiary health facility for proximity to the father's location on request by the father. The patient has been commenced on chemotherapy at the referral center. The prognosis is good and sometimes this tumor regresses spontaneously despite the high chance of metastasis.⁶

CONCLUSION

The report shows a rare but possible occurrence of neonatal neuroblastoma among Nigerian neonates. The finding of neuroblastoma in the neonatal period suggested the possibility of the disease starting in-utero which was missed during the obstetric scan in this index neonates. With increased awareness of neonatal neuroblastoma, the incidence of anomaly scans will increase, improve early detection and management in-utero, and proffer the opportunity for necessary precautions during delivery.

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