

Case Report

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Neonatal neuroblastoma in a full term Saudi girl: A case report

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Abstract

Neuroblastoma is the commonest malignancy during the neonatal period. It may present as an adrenal mass or with localized or metastatic (4s/Ms) disease, which is typically low risk with an excellent prognosis. We report the case of a 2-day-old neonate with neuroblastoma presenting with right suprarenal mass without evidence of metastasis. This raises the chance of prenatal origin of neuroblastoma, which was missed during a repeat ultrasound done during antenatal care. Although screening for neonatal neuroblastoma is not indicated, targeted screening of infants at risk is acceptable. We hope that this case report will increase the notice of physicians and sonographers about neuroblastoma and other space-occupying lesions within the fetus.

Keywords: Neuroblastoma; Ultrasonography; Adrenal mass; Neonates.

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Introduction

Neonatal tumors are considered rare disorders [1]. Among all pediatric malignancies, neonatal tumors attribute for only 2% with an incidence of 1.58 to 3.65 per 100,000 live births [2]. Neuroblastoma is the most common neonatal tumor accounting for 28–39%, with an incidence of 0.61 per 100,000 live births [3]. Neuroblastoma is originating from neural crest cells which rise to the adrenal medulla and sympathetic ganglia [4]. The adrenal gland is the most typical site of presentation in neonatal neuroblastoma (90%) [5]. Adrenal glands make hormones that control automatic functions of the human body including

digestion, blood pressure, respiration, and heart rate. Other origins of neuroblastoma include nerve tissue in the spinal cord, abdomen, chest, or neck with further spread to other parts of the body [6]. Twenty percent of neonatal neuroblastoma can present with compression of the spinal cord that requires immediate investigations and treatment with steroids and chemotherapy to alleviate the cord compression [7]. Neonates with stage Ms without life-threatening manifestations of adverse genetic features (MYCN amplification or segmental chromosomal abnormalities) is observed for spontaneous regression which can also occur with other localized types of neonatal neuroblas-

tomas [6,8]. Neuroblastoma has a wide range of presentations depending on the site of the tumor and therefore the stage of the disease. Symptoms of neuroblastoma include but are not limited to fatigue, decreased appetite, bulging eyes or dark circles under eyes, pale skin as an indication of anemia, and a lump within the chest, neck, or belly. Treatment modalities can include chemotherapy, radiotherapy, and surgery [5,7]. The prognosis of neuroblastoma depends on the stage of the disease, the child's age, and therefore the risk category. However, the prognosis of neonatal neuroblastoma is good with favorable biological characteristics in most cases. Despite the metastatic invasion that may occur in neonatal neuroblastoma, spontaneous regression is typically expected [7]. Although stage 4S has a favorable prognosis, is often fatal for neonates because of compression manifestation on the liver or inferior vena cava (IVC) [9].

Case report

Our case was a 2-day-old female full-term neonate and was born in Prince Sultan Military Hospital, King Fahad Air Base, Hawiyah, Taif, Saudi Arabia. She was delivered at term by spontaneous vaginal delivery. Pregnancy was uneventful and antenatal obstetric ultrasound scans were done revealing no abnormality. Birth weight was 3.2 kg and there was no history suggestive of perinatal asphyxia. There was a robust case history of congenital renal anomalies. Positive family history of malignancy rather than neuroblastoma.

The baby was vitally stable and asymptomatic. Clinical examination showed normal weight, length, and head circumference. Examination of the chest and heart were normal. Abdominal examination revealed no organomegaly or palpable masses. Skin examination was normal. Neurologically, she was conscious, active, and crying with good neonatal reflexes. The rest of the neurological examination was normal. Examination of the motor system revealed normal muscle tone and strength globally. Routine laboratory investigations, C-reactive protein (CRP), and coagulation profiles were normal. Right suprarenal mass was detected during the routine ultrasound- to rollout congenital renal anomalies- within the 2nd day of life with no reported renal anomalies. It was a small 3.2 X 1.8 cm heterogeneous solid mass, hyperechoic replacing the right adrenal gland with mass effect on the right kidney. The left adrenal gland, liver, spleen, and both kidneys were normal. Colour Doppler showed prominent internal vascularity.

A computerized tomography (CT) scan of the abdomen and pelvis revealed a little mass within the right adrenal gland, solid, nodular, with a central area of calcification. The mass measured 3.1 cm X 2.4 cm X 2.2 cm.

Abdominal contrast-enhanced CT was suggestive of right suprarenal neuroblastoma with no evidence of hepatic metastases.

MRI abdomen with contrast and multiple sequences in both T1 and T2 showed a right suprarenal mass, heterogeneous, with low signal intensity in T1 with areas of hemorrhage and high signal intensity in T2 enhancing after contrast injection.

Serum and urinary catecholamines and their metabolites

were not raised and the genetic study failed to nMyc (myelocytomatosis viral related oncogene) amplification.

The baby was subjected to Methyl iodo benzyl guanine (MIBG) scan; the 123-iodinated MIBG compound is specific for neuroblastoma cells, this specialized scan helps to see whether or not the neuroblastoma has spread to other distant areas of the body. Our case showed right suprarenal neuroblastoma with no evidence of spread during the MIBG scan.

Our case was a low-risk and early-stage neuroblastoma in a young baby. A pediatric oncologist was consulted about the case. He recommended a wait-and-see strategy with ultrasonography follow-up. During follow-up, serial ultrasounds were done showing a gradual spontaneous decrease in the size of the right suprarenal mass till complete resolution after 1.5 years.

Discussion

Neonatal neuroblastoma is defined as neuroblastoma identified within the first month of life [10]. Neuroblastoma is considered the most common extracranial solid tumor in infancy and can arise from neural crest elements of the sympathetic chain [11].

Neuroblastoma is more common in males than females, with a ratio of 1.2:1 respectively [12]. About 57% of infants with neuroblastomas were diagnosed early in life [10]. The origin of neuroblastoma is the adrenal medulla and paraspinal or periaortic regions. About 65% of primary neuroblastomas are abdominal mostly—(40%) in the adrenal gland [11]. Antenatal diagnosis is made after 32 weeks of gestation and 93% of tumors are mostly adrenal in origin [12]. The ultrasound is done during antenatal care or postnatal could be a useful screening tool within the detection of neonatal neuroblastoma. Improvements in prenatal imaging have led to an increased rate of diagnostic techniques of fetal neuroblastoma [10]. However, some cases may be missed diagnosis during prenatal ultrasound as in our case report where ultrasonography failed to identify the adrenal mass during antenatal care. Children with a positive family history of neuroblastoma are more likely to develop it. However, the bulk of neuroblastoma is not hereditary. Also, Children with congenital anomalies may have more risk of neuroblastoma [10]. Diagnosis of neuroblastoma is confirmed by histopathology evaluation of tumor tissue or tumor cells in a bone marrow sample. Biochemical studies in infants with neuroblastoma showed increased urine or serum catecholamines or their metabolites [13]. The differential diagnosis of an adrenal mass in neonates includes adrenal hemorrhage, extrapulmonary sequestration, bronchogenic cyst, or renal duplication [14].

In terms of recurrence, neuroblastoma is stratified into low- and intermediate-risk groups [11]. In low-risk patients with neuroblastoma, the utilization of a watch-and-wait approach becomes a recent trend in management because of the high rate of spontaneous regression of this tumor type. Also, an effort to scale back surgical intervention and cytotoxic chemotherapy in some intermediate-risk patients. In a shot to minimize exposure to surgical intervention and cytotoxic chemotherapy is usually recommended [15].

The International Neuroblastoma Risk Group Staging System

(INRGSS) is used to determine the stage of neuroblastoma. The stages of neuroblastoma are:

Stage L1: Tumor is confined to one compartment with no spread. Also, the tumor has the lowest risk with no involvement of vital structures of the body.

Stage L2: Tumor is confined to at least one body compartment with possible spread to regional lymph nodes. Also, there's the involvement of important structures of the body, like large blood vessels.

Stage M: Tumor cells spread to more than one body compartment (distant metastatic disease) with the highest risk.

Stage 4S is a special category of neuroblastoma with metastasis to the skin, liver, or bone marrow. This stage is considered low-risk neuroblastoma with a wonderful prognosis [14,16].

Conclusion

We hope that this case report will increase the awareness of neonatologists, obstetricians, and sonographers about neuroblastoma and other space-occupying lesions in the fetus. Early diagnosis of neuroblastoma during antenatal care offers the opportunity for precautionary cesarean section to prevent possible adrenal hemorrhage during vaginal delivery. Aggressive treatment of neonates with neuroblastoma is not recommended.

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