ECRONICON OPEN ACCESS

EC PAEDIATRICS
Case Report

Neonatal Adrenal Neuroblastoma with Normal Urinary Catecholamines. A Case Report

Satish Kumar KV[1]*, Venkatanarasimhan[2], Asha Thomas[3], Suman Rath[4], Naveen Thomas[5], Balachandra Bhat[6], Hariprasad Gnanavelu[7] and Rahul Krishnan[7]

[1]Visiting Consultant Pediatric Surgeon, Baptist Hospital, Bangalore, India

[2]Head, Department of General Surgery, Baptist Hospital, Bangalore, India

[3]Head/Consultant Pediatric Radiologist, Department of Radiology, Baptist Hospital, Bangalore, India

[4]Consultant Pediatrician, Department of Pediatrics, Baptist Hospital, Bangalore, India

[5]CEO and Consultant Pediatric Surgeon, Department of General Surgery, Baptist Hospital, Bangalore, India

[6]Consultant Pathologist, Department of Pathology, Baptist Hospital, Bangalore, India

[7]Resident, Department of General Surgery, Baptist Hospital, Bangalore, India

***Corresponding Author:** Satish Kumar KV, Visiting Consultant Pediatric Surgeon, Baptist Hospital, Bangalore, India.

Received: September 28, 2020; **Published:** October 19, 2020

Abstract

There is increased incidence of perinatally detected neuroblastomas due to routine antenatal ultrasound. These tumors have excellent prognosis as they are detected in early stages, have favourable biological characters and also known to undergo spontaneous regression. An adrenal mass was detected on routine postnatal ultrasound in a neonate on 2nd day of life. This presentation is unique for neonatal neuroblastoma. It was detected during postnatal evaluation for antenatally detected anomaly (single umbilical artery), the urinary catecholamines were not raised and had no nMyc amplification.

Keywords: *Neonatal Adrenal Neuroblastoma; Urinary Catecholamines; Umbilical Artery*

Introduction

Neonatal adrenal masses can be of benign (hematoma) or malignant pathology (Neuroblastoma). There is increasing incidence of perinatal neuroblastomas and due to their special biological characters, have favourable prognosis. A case of neuroblastoma detected on day 2 of life which was enlarging on serial imaging, the diagnostic dilemma and successful curative laparoscopic resection is presented.

Case Report

A healthy baby girl with antenatal ultrasound suggestive of single umbilical artery was delivered at term with no clinical evidence of external dysmorphism. A routine abdominal ultrasound obtained on day 2 of life as baby had single umbilical artery on prenatal ultrasound. The scan showed a 1.7 x 1.1 cm, right adrenal echogenic mass and the kidneys were normal. The mass had peripheral vascularity (which is not typical of adrenal hematoma), a follow up scan was repeated after 2 weeks. The mass had increased in size and vascularity (Figure 1). A Contrast enhanced CT scan (Figure 2) showed a 1.5 x 1.5 cm right adrenal mass, suspicious of malignancy. The parents were counselled regarding the possibility of malignancy and was decided to follow up with clinical evaluation and follow up scans. The urinary catecholamine assay was normal and further follow up scans and clinical evaluation at 2 weekly intervals were performed. On

serial ultrasounds, the mass was increasing in size (3.4 x 1.7 cm at 2 months) with increasing vascularity. The parents were counselled for further follow up or surgical excision and were keen on surgical excision. The baby underwent elective laparoscopic resection at 4 months of age. The tumor was fleshy, localised to right adrenal and a complete laparoscopic resection (Figure 3) was accomplished with no tumor spillage. Postoperative recovery was uneventful and the baby was discharged home on day 3. The histopathology confirmed poorly differentiated neuroblastoma (Figure 3) and nMyc amplification was negative. As the tumor was completely resected (Stage 1) and of favourable biological characters, further adjuvant chemotherapy was not offered. The baby was followed up at 3 monthly intervals (Figure 3) and a repeat CT scan at 1 year showed no evidence of recurrence. The baby is now 2 and a half years, thriving and clinically well with no recurrence on ultrasound scans.

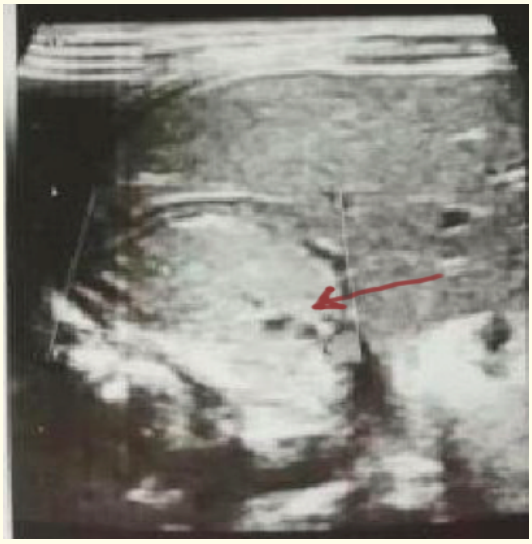

Figure 1: *Ultrasound at 2 weeks showing solid adrenal mass.*

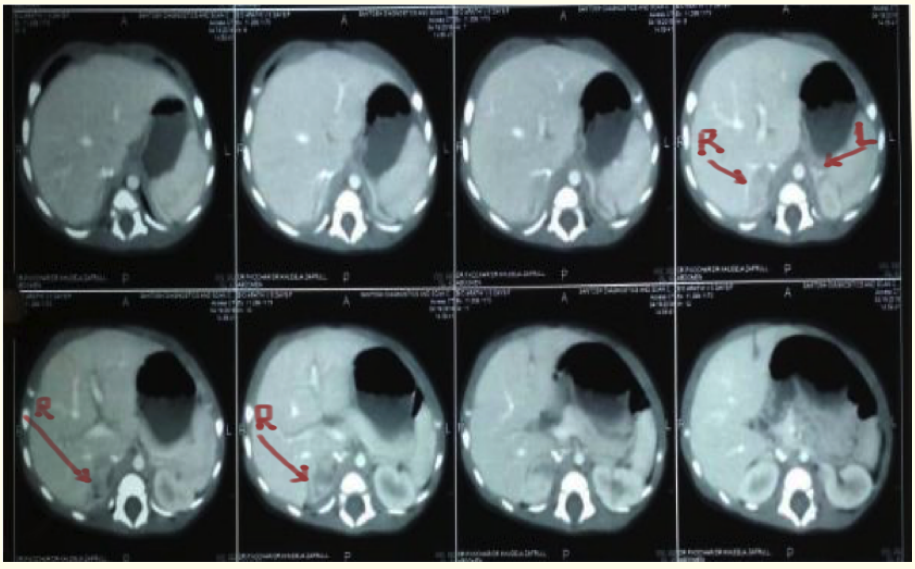

Figure 2: *CT scan of abdomen at 2 months - 1.5 x 1.5 cm adrenal mass with vascularity- Suspicious for malignancy.*

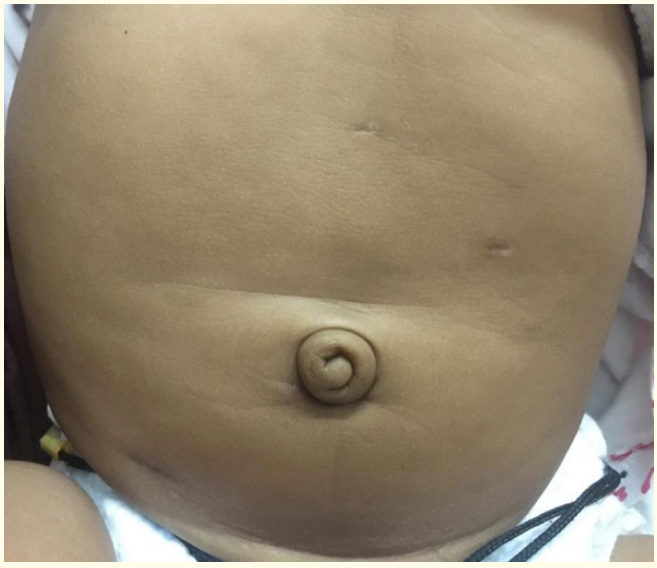

Figure 3: *Appearance of port sites at 3 months. A 5 mm umbilical port, two 3 mm ports and 10 mm right lower quadrant port (for retrieval of tumor in specimen bag) were used.*

Discussion

Neuroblastoma (NB) is by far the most prevalent cancer of infancy with a peak incidence at 18 months. According to a review by Dhir S., *et al*. [1] 16% of infant neuroblastomas were diagnosed during the first month of life (neonatal NB) and 41% during the first 3 months. In our index case, an adrenal mass was incidentally detected on day 2 of life, during sonographic evaluation for anomalies associated with single umbilical artery.

Adrenal hemorrhage is the most important differential (Table 1) for neonatal adrenal masses and may be difficult to differentiate from neuroblastoma. On ultrasonography, it shows variable echogenicity and absence of doppler blood flow. On serial ultrasounds, the adrenal hemorrhage decreases in size and changes echogenicity within the first week, followed by peripheral increased echogenicity due to calcification. The echogenicity of neuroblastomas is variable, but presence of blood flow differentiates it from adrenal hemorrhage [2].

In our present case, in addition to increase in tumor size, there was increasing vascularity. Though the urinary catecholamines were not raised, suspicion of neurogenic tumor was high. Though the baby was managed non-operatively for 3 months, the patents were keen for surgical excision. It should be noted that though urinary catecholamines are raised in > 90% of neuroblastomas, only 33% of perinatal neuroblastomas have raised urinary catecholamines [3]. Neuroblastomas are categorised into low, intermediate and high-risk groups based on histology, biological characters and presence of allelic loss of 1p and nMyc oncogene amplification. Around 70% of neonatal neuroblastomas fall into this low-risk category [4]. In only two of the 95 patients tested for nMyc had amplification of the gene in a study of prenatal neuroblastomas by Italian study group [5]. Surgical resection is the standard treatment, with excellent overall results. In our index case, the final histopathology confirmed poorly differentiated neuroblastoma, With a stage 1 tumor (post resection) and favourable biological characters (negative nMyc amplification), adjuvant chemotherapy was avoided and baby is well with no recurrence at 2.5 years of age.

In a study by Italian neuroblastoma group [5], 27/134 babies (20.1%) were detected prenatally and localised disease prevailed (65.7%) with an increase of stage 1 patients over a study of 26 years (from 18.8% to 46.5%). All stage 1 patients (48 patients) underwent

resection with no mortality, while 3/15 patients with stage 2 disease died from complications of surgery. The overall mortality for young infants undergoing surgery is greater than or equal to 2% [6]. Careful consideration should be employed during planned surgical resection, as these have favorable biological characters and have tendency to undergo spontaneous regression.

With increasing use of antenatal ultrasound, detection of neonatal neuroblastomas in earlier stages has increased. There is considerable evidence for spontaneous regression of tumors detected through population screening programs for neuroblastoma and spontaneous regression of larger tumors after diagnostic biopsy or partial resection have been observed [7]. Though the rate of spontaneous regression of NB is not known, neurotrophin deprivation, humoral or cellular immunity, loss of telomerase activity and alterations in epigenetic regulation are hypothesized as possible mechanisms for regression. Considering the capacity to undergo regression, several pharmacological, biological or immunological approaches are underway to induce regression as a therapeutic approach in neuroblastomas [8].

Conclusion

The ultrasound features of neonatal neuroblastomas are variable and often difficult to differentiate from adrenal hemorrhage in initial stages. Increasing size and vascularity, necessitates ruling out neuroblastomas. Laparoscopic adrenalectomy is feasible and safe in these small children, who are detected in early stages. As most of these have biologically favorable prognostic features, adjuvant chemotherapy can be avoided.

Bibliography

1. Dhir S and Wheeler K. "Neonatal neuroblastomas". *Early Human Development* 86 (2010): 601-605.

2. Navarro AM and Daneman A. "Caffey's pediatric diagnostic imaging". 3rd edition. 121: 1167-1171.

3. Fishera JPH and Tweddle DA. "Neonatal neuroblastomas". *Seminars in Fetal and Neonatal Medicine* 17 (2012): 207-215.

4. Nuchtern JG., *et al.* "A prospective study of expectant observation as primary therapy for neuroblastoma in young infants: a Children's Oncology Group study". *Annals of Surgery* 256.4 (2012): 573-580.

5. Gigliotti AR., *et al.* "Neuroblastoma in the newborn. A study of the Italian Neuroblastoma Registry". *European Journal of Cancer* 45.18 (2009): 3220-3227.

6. Yao W., *et al.* "Neonatal suprarenal mass: differential diagnosis and treatment". *Journal of Cancer Research and Clinical Oncology* 139 (2013): 281-286.

7. Nuchtern JG., *et al.* "A prospective study of expectant observation as primary therapy for neuroblastoma in young infants: a Children's Oncology Group study". *Annals of Surgery* 256.4 (2012): 573-580.

8. Brodeur GM and Bagatell R. "Mechanisms of neuroblastoma regression". *Nature Reviews Clinical Oncology* 11.12 (2014): 704-713.

Volume 9 Issue 11 November 2020