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Congenital Neuroblastoma: A Case ReportCongenital Neuroblastoma: A Case Report

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Introduction

Neuroblastoma (NB), a tumor of primitive neuroectodermal cells, is diagnosed exclusively in children and is the third most common childhood cancer and the most common neonatal malignancy that accounts for >20% of neonatal cancer [1]. The presentation of NB can vary widely, as it could spontaneously regress, mature to a benign ganglioneuroma, or metastasize aggressively. NB occurring in the perinatal period frequently carries a good outcome with most of the tumors carrying normal MYCN copy number and hyperdiploid DNA index [2]. The mean age at diagnosis is 17.3 months, however a review of the literature revealed a sparsity of data reported on congenital cases of adrenal neuroblastomas with in-utero liver metastasis. We describe a case of a female infant born at full-term in June 2018 with Congenital Neuroblastoma on the right adrenal along with extensive metastatic deposits on the liver.

Clinical Summary

A second uneventful pregnancy of a young mother resulted in the vaginal delivery of a 2.5kg female baby at full-term. She was delivered in Najran at a primary care hospital. Our patient presented with abdominal distention at time of birth. On examination, she had no signs of jaundice or pallor. Her vital signs were normal. Her abdomen was enlarged and measured at 39cm. Upon palpating the abdomen, hepatic border was palpable 8cm below the costal margin. The abdomen was non-tender and soft in consistency. Other abdominal organs were not palpable. An Ultrasound Abdomen was performed showing heterogeneous increased parenchymal echogenicity of the liver with moderate enlargement, no focal hepatic lesion identified. Right adrenal mass lesion was identified for further evaluation. A CT Chest Abdomen Pelvis was also performed revealing hepatomegaly with diffuse heterogeneous infiltrative lesion with bilateral suprarenal homogeneous soft tissue lesions likely representing adrenal hemorrhage [figure 1]. She was initially admitted at our hospital as an elective case of right sided hematoma for further investigation. During her hospitalization, she developed frequent episodes of hypoglycemia down to 2mmol/L. A liver biopsy was performed, the sample consisted of three irregular-shaped pink-tan soft tissue biopsies, measuring in aggregate 1x0.3x0.2 cm. The sample confirmed metastatic neuroblastoma [figure 2]. Immunohistochemistry showed positive for chromogranin and synaptophysin [figure 3]. Staining for TdT, CD99, CK AND WT-1 were negative. A sample was sent for Bone Marrow Biopsy that showed no evidence of metastatic involvement. A chromosomal analysis revealed no change in copy number of MYCN. Newborn metabolic screen was unremarkable.

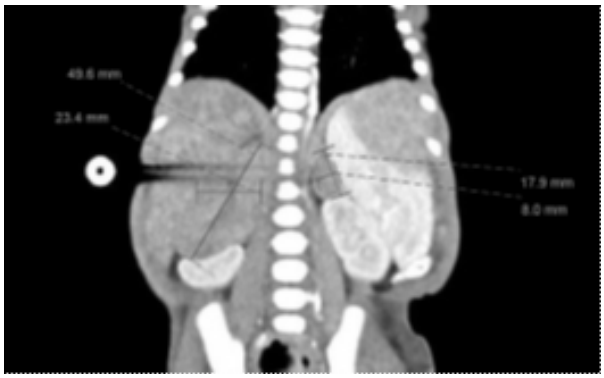

***Figure 1:** Chest Abdomen Pelvis CT scan.*

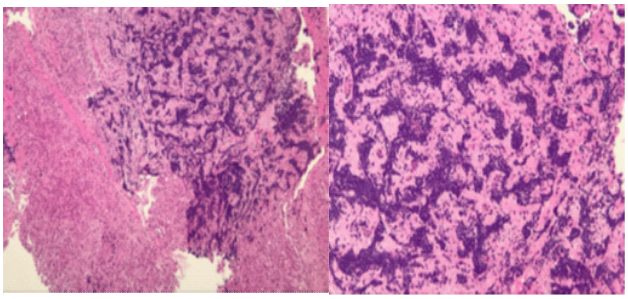

***Figure 2:** The sample confirmed metastatic neuroblastoma.*

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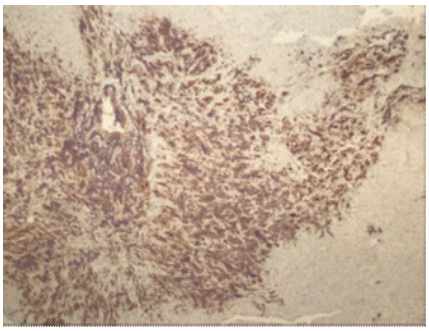

Figure 3: *Immunohistochemistry showed positive for chromogranin and synaptophysin.*

Discussion

Neuroblastoma is the leading malignant neoplasm of the perinatal period and the second most common solid tumor in childhood following brain tumors [3, 4]. Clinical and biological features of NB occurring in fetuses and neonates differ from that in the older age groups, for they have been documented to have a normal MYCN copy number and hyperdiploid DNA index, both of which are associated with a favorable out-come [1]. Neuroblastoma in the older children are more commonly diploid tumors with amplification of MYCN oncogene, connoting an advanced disease stage and a poor outcome. Amplification of the MYCN oncogene is present in approximately 25% of neuroblastomas [4]. Studies have documented that most fetuses present with a favorable stage of the disease with a good prognosis. We hope in future researches to identify risk factors associated with congenital NB, as well as factors associated with a favorable prognosis.

Conclusion

In conclusion, we describe a case of congenital NB present at birth with metastasis to the liver parenchyma. After completion of chemotherapy the patient showed good response to treatment with improvement of the liver size and echogenicity. The patient is still being followed up.

Acknowledgments

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