A Case Report of Congenital Neuroblastoma

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Introduction

It is believed that the incidence rates of neuroblastoma are lowest in developing countries but this could be as a result of under reporting. It is important to remember that the congenital variety of neuroblastomas constitutes the majority of malignant tumors in the newborn and could be missed if awareness and index are low. The prognosis is however excellent with early diagnosis and appropriate treatment.

We report a rare case of congenital neuroblastoma making its first presentation in the paediatric dermatology clinic. Neurofibromatosis type 1 is a close differential which was ruled out by histology. A high index of suspicion is needed to make a diagnosis of congenital neuroblastoma in a neonate that presents with subcutaneous nodules at birth.

Case Report

A 16 month old child presented at the dermatology clinic with multiple subcutaneous nodular swellings, the first of which was the size of a pea, located on the sole of the right foot and first noticed at birth. At about 6 weeks of age, another appeared behind the right ear. Other nodules gradually made their appearances with time. The swellings were firm, non-tender and of varying sizes.

A right abdominal flank swelling was noticed 2 weeks prior to presentation and had been gradually increasing is size. There were multiple hypo pigmented patches and café au lait spots on the skin of the trunk and extremities. The child was otherwise well. The working diagnosis at presentation was neurocutaneous disorder.

Abdominal ultrasound scan revealed an intra-abdominal tumor in the right hypochondrium and suprarenal area with downward displacement of the right kidney. Fine needle aspiration biopsy of one of the nodules showed singly, disposed epithelial and mesenchymal cells in diffuse pattern. The cells were ovoid to spindle-shaped and exhibited mixed pleomorphism, coarse chromatin and prominent nucleoli suggesting a malignant picture but it was not specific for neuroblastoma. Computed Tomography scan was highly suggestive of neuroblastoma**.** Sections of biopsy of axillary lymph nodes showed encapsulated and nodular tissue containing clusters, cords and many nests of tumor cells. The nests were separated by neural stromas that contained some fibrous tissue. Tumor cells were small and hyper chromatic with scanty cytoplasm and non-prominent nucleoli. Homer-Wright pseudo rosettes were seen. Urinary assays of Homovanillic acid and Vanillyl Mandelic acid were not done as a result of financial constraints. The diagnosis at this point was stage IV neuroblastoma.

Chemotherapy was chosen as the best option of management and the nodules decreased in size after the first course of chemotherapy of Vincristine, Adriamycin and Cyclpohosphamide. Unfortunately, the child was lost to followup.

Discussion

Neuroblastoma is the most common extra cranial solid tumor in infancy. [1] Tumors can develop in the abdominal cavity (40% adrenal, 25%paraspinal ganglia) or other sites (15% thoracic, 5% pelvic, 3% cervical tumors, 12% miscellaneous).[2] Neuroblastoma accounts for approximately 7.8% of childhood cancers in the United States of America[3] with an incidence of about 9.5 cases/million children.

Congenital neuroblastoma is the commonest malignant tumor of the newborn, comprising 20% of all malignancies encountered during the neonatal period.[4] A family history of

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neuroblastoma has been reported in 1-2% of patients and follows an autosomal dominant pattern of inheritance.[5] Associations with Hirschsprung disease, congenital hypoventilation syndrome and Neurofibromatosis type 1 (NF1) have been reported.[6] The presence of café au lait spots in our patient suggested the diagnosis of a neurocutaneous pathology either existing alone or as a co-morbidity.

The age of the patient and the stage of the disease at the time of diagnosis are the most important prognostic factors.[7] Patients with localized tumor irrespective of age, have an excellent outcome (80-90% three-year event-free survival rate) while those more than 18 months with metastatic disease fare poorly. Our patient's prognosis may have been better if presentation had been much earlier. More than 50% of patients present with metastatic disease at the time of initial hospital visit: 15% have regional extension and approximately 7% present during infancy with disseminated disease limited to the skin, liver and bone marrow.[2] Fine needle aspiration biopsy though helpful in diagnosis of neuroblastoma is limited in its diagnosis when the tumor is poorly differentiated because of morphologic similarities with other small round cell tumors.[8]

Conclusion

This is a rare case of congenital nephroblastoma being reported in a developing country and making its first presentation in the paediatric dermatology clinic. Neurofibromatosis type 1 is a close differential which was ruled out by histology findings though it could exist in a co -morbid state. A high index of suspicion is needed to make a diagnosis of congenital neuroblastoma in a neonate that presents with skin nodules at birth.

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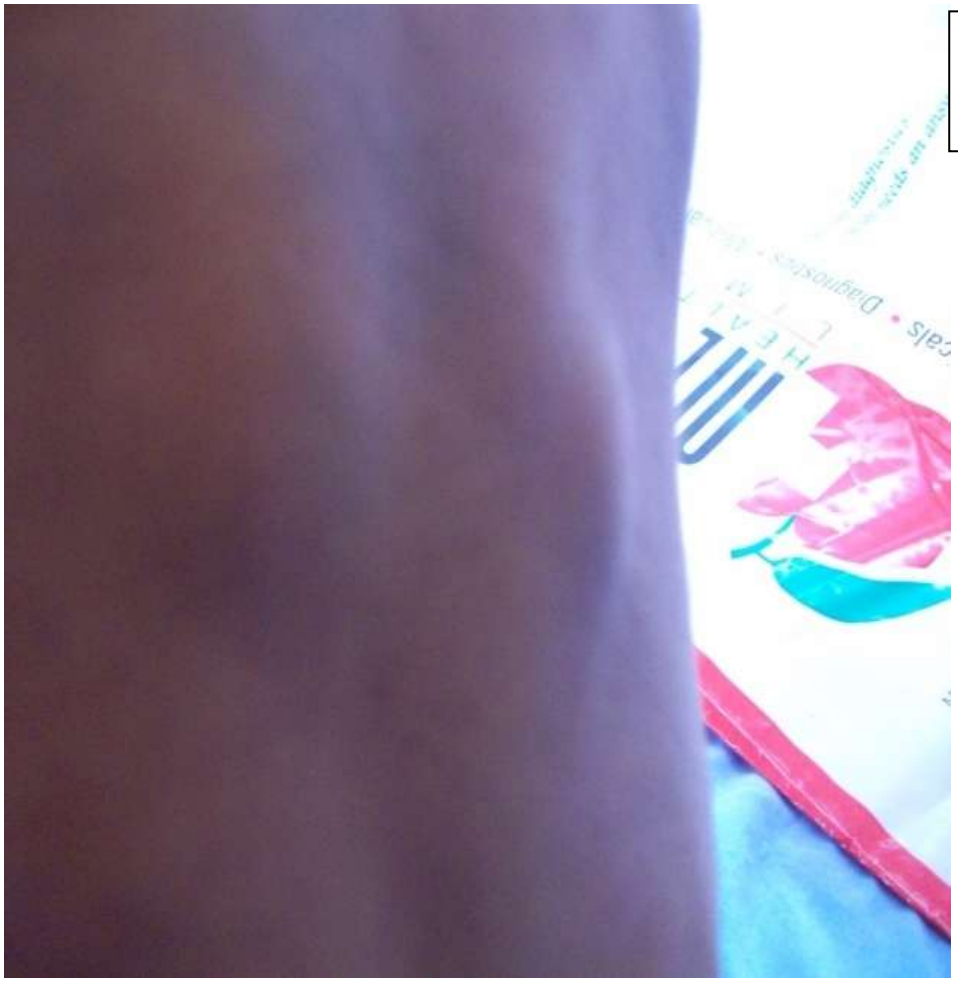

Fig 1. Subcutaneous nodules on the back of the child

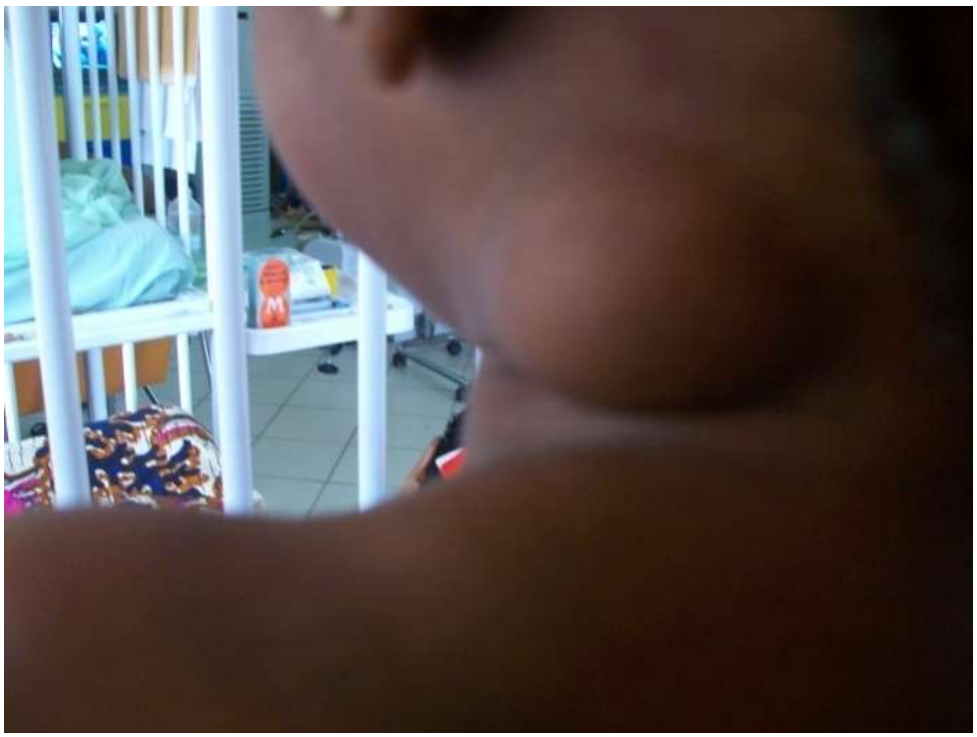

Fig 2. Subcutaneous nodules in the cervical region of