

Case Report


Neuroblastoma in 3 months old infant - A rare case report

Shreya Shah^{1*}, Nisarg Champaneria¹, RK Pasle²

¹PG Student, ²Professor and Head

Department of Pathology, SBKS MI & RC, Sumandeep Vidyapeeth, Vadodara, Gujarat, India

*Corresponding author email: shreya27291@gmail.com

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Abstract

Neuroblastoma is one of the most common childhood malignancies. Patients diagnosed under 1 years of age (40%) have a much better prognosis than those diagnosed in later childhood. Neuroblastoma originates from neural crest cells of the adrenal medulla or sympathetic ganglia. Neuroblastomas commonly present as an asymptomatic mass in a child. The treatment of neuroblastoma depends on the stage, and it includes surgical excision, multi agent chemotherapy, and bone marrow transplantation.

Key words

Neuroblastoma, Childhood malignancies, Asymptomatic mass.

Introduction

Neuroblastoma is one of the most common childhood malignancies [1, 2]. Patients diagnosed under 1 years of age (40%) have a much better prognosis than those diagnosed in later childhood [3]. The wide range of clinical presentations makes the diagnosis, clinical management and follow-up a challenge. It can be diagnosed by ultrasound or CT scan. Urine VMA analysis, FNAC or biopsy from the metastatic lesions can confirm the diagnosis. Here we report

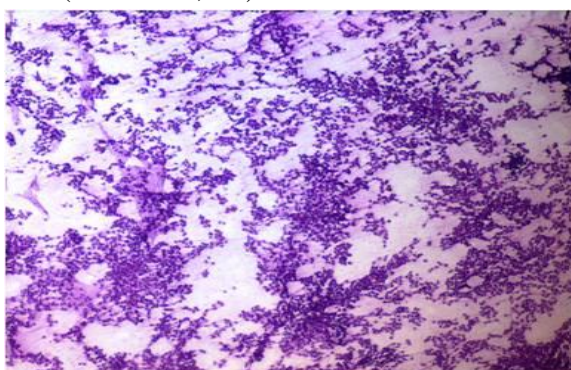
a case of 3 months old infant diagnosed as neuroblastoma with multiple metastasis.

Case report

A 3 months old female baby, born via Cesarean section presented with many cutaneous nodules and fever since 15 days. Mother also gave history of abdominal mass since 2 months. Clinically there were presence of multiple cutaneous nodules and hepatomegaly. Routine investigations showed low Hb, minimally elevated ESR and a positive urine VMA spot

test. USG showed hepatomegaly with multiple hypo echoic lesions having peripheral halo. The left suprarenal area showed a well-defined hypo echoic lesion with central ill-defined area of increased echogenesity. FNAC was performed at cytology clinic using a 22-guage needle attached to a 10 ml syringe after explaining procedure to the patients and obtaining their oral consent for the same [4-10]. The area to be aspirated was cleaned with spirit before aspiration and multiple hits were made within the lesion, with sufficient negative pressure; the needle was removed and the pressure was applied to the area of aspiration to avoid bleeding or hematoma formation. The aspirated material was smeared on glass slide and stained [11-18]. FNAC was taken from the skin nodules. Smears studied showed highly cellular smears with presence of small cells with dark nucei and scanty cytoplasm. There was presence of clusters of cells with presence of typical Homer-wright rosettes composed of several rows of cells arranged around the circular space that is filled with fine neurofilaments (**Photo – 1, 2, 3**). The diagnosis was given as neuroblastoma stage 4.the baby died within 15 days during the hospital stay.

Photo – 1: Highly cellular smear with the tumor cells (H&E stain, 4X).



Discussion

Neuroblastoma originates from neural crest cells of the adrenal medulla or sympathetic ganglia. Neuroblastomas commonly present as an asymptomatic mass in a child [2, 19]. Other mode of presentations are fever, bone pain, opsoclonus, cerebellar ataxia, orbital ecchymosis or intractable diarrhea, and less commonly with

myoclonus [20]. Our patient was also presented with abdominal mass, fever and skin nodules.

Photo – 2: The tumor cells are small with dark nuclei and scanty cytoplasm (H&E stain, 10X).

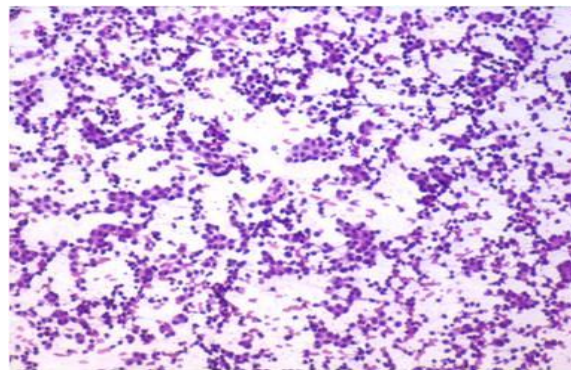
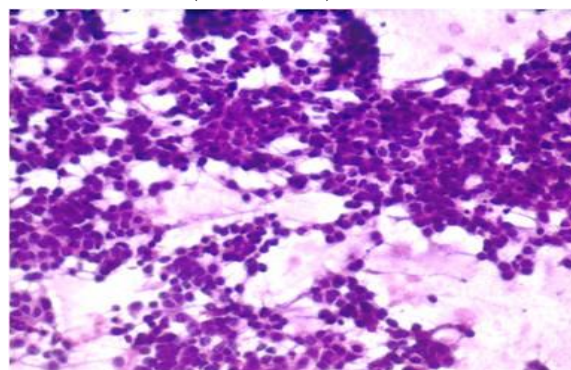


Photo – 3: Tumor cells with fine extracellular neurofilaments (H&E, 20X).



Knowledge of local tumor extent as well as accurate staging of the disease are essential for proper treatment [21]. In resectable stage 1 (confined to the site of origin) and stage 2 (extending beyond the site of origin but not crossing the midline), surgery alone is usually sufficient without the need for chemotherapy or radiotherapy. By contrast, in stage 3 (tumor extending beyond the midline) and stage 4 (remote metastases to bones or lymph nodes), chemotherapy and/or radiotherapy are generally required.

FNAC was taken mainly for extra adrenal sites, such as metastasis to bone, liver or skin. It will be helpful in the diagnosis or the workup for the staging. The smears are usually rich in small tumor cells with homogenous hyper chromatic

nuclei and a tiny rim of cytoplasm. The cells are either dispersed or form small clusters. The clusters must be very carefully examined in the search for typical Homer-Wright rosettes composed of several rows of cells arranged around an approximately circular space that is either empty or filled with fine neurofilaments. This finding is diagnostic of neuroblastoma. The rosettes are often distorted and composed of disorderly clusters of small cells and small, empty, central spaces. The neurofibrils can also be seen outside the rosettes under high power of the microscope as tangled, thin, eosinophilic lines.

The treatment of neuroblastoma depends on the stage, and it includes surgical excision, multiagent chemotherapy, and bone marrow transplantation [22]. Most patients with neuroblastoma have a favorable stage of disease with excellent long-term prognosis.

Conclusion

Neuroblastomas are derived from the neural crest ectoderm, and are the most common solid abdominal masses of infancy. Urine Vanillylmandelic acid (VMA) spot test was positive and biopsy from the skin nodules and liver confirmed the diagnosis of neuroblastoma with metastasis.

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