International Journal of Research in Medical Sciences
Das AM et al. Int J Res Med Sci. 2017 Dec;5(12):5466-5469
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20175476

Case Report

Head neck neuroblastoma in early childhood: a rare case report

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Received: 27 September 2017
Accepted: 01 November 2017

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ABSTRACT

Neuroendocrine tumors (NETs) comprise a heterogeneous group of malignancies from cells derived from the neural crest with neuroendocrine differentiation. Neuroblastoma is an embryonal malignancy of the sympathetic nervous system arising from neuroblasts (pluripotent sympathetic cells). It can be sporadic or nonfamilial in origin. Despite the differences in the site of origin, nomenclature, biological behaviour, and functional status, NETs share certain immunohistochemical and ultrastructural features. NETs are relatively rare tumors with an annual incidence of 6 new cases per 100000 inhabitants. It is primarily a tumor of abdominal origin from where it metastasizes to lymph nodes, liver, orbital sites, and central nervous system. Head and neck NETs are uncommon. We report a rare case of neuroblastoma in a 3-year-old child presenting with an unusual large fungating mass in left parotid region.

Keywords: Head neck, Immunohistochemistry, Neuroblastoma

INTRODUCTION

The third most common extracranial solid neurogenic tumour of infancy and childhood is Neuroblastoma along the peripheral sympathetic nervous system.[1] Dr. Rudolf Virchow first described it as "glioma" in the abdominal cavity.[2] Neuroblastoma was described as primitive neural cells tumor within the bone marrow by Homer-Wright.[3] Neuroblastoma is detected approximately in 1/7000 live births.[4]

Neuroblastoma may be sporadic or nonfamilial in origin. Exact etiology of neuroblastoma is not well understood, but the recent studies have improved the understanding of genetic susceptibility to neuroblastoma.[5] It originates mostly from the adrenal gland, nerve tissues of the neck, chest, abdomen, or pelvis.[6]

Majority of the neuroblastomas are diagnosed in children younger than 5 years of age.[7] NETs of the head and neck region represent a diagnostic and therapeutic challenge in the routine practice. A complete work-up is necessary to rule out a metastatic origin of the tumor.[8,9] An adequate sub classification of NETs in the head and neck area regarding the degree of differentiation is required to predict the clinical behaviour and to support the treatment decision-making.

Clinical, morphological and immunohistological correlations in large series of cases are necessary to provide clear diagnosis and define the best therapeutic options.[10] The present case reports a rare case of neuroblastoma of cheek in preauricular region of a 3-year-old boy. Age, stage, and biological features encountered in tumor cells are important prognostic factors and are used for risk stratification and treatment.

CASE REPORT

A 3-year-old boy was brought to surgical department of United Nations Hospital in central Africa with the chief complaint of swelling over left side of cheek extending down from left auricular region growing rapidly over last six months. He was treated with some local medications; detailed medical history was not available. He was the second child of a healthy non-consanguineous parent. Physical examination revealed a large fungating mass over left cheek in parotid region (Figure 1).

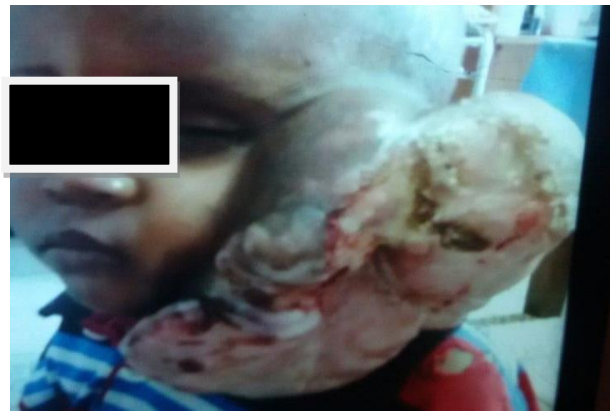


Figure 1: Gross picture of the tumour.

There were no palpable cervical lymph nodes, no intraoral lesions, or facial nerve paralysis. No abnormalities of hand and feet were observed. Intraoral examination was unremarkable. No tumour mass was found in abdominal USG. Based on the clinical features, a provisional diagnosis of malignancy was made. A complete blood profile and biopsy was performed. The complete blood profile was within normal limits. Biopsy under the light microscopy showed monomorphous round cells with salt-and-pepper chromatin arranged mostly in nests with a solid or cribriform pattern that formed frequent rosette-like structures (Figure 2).

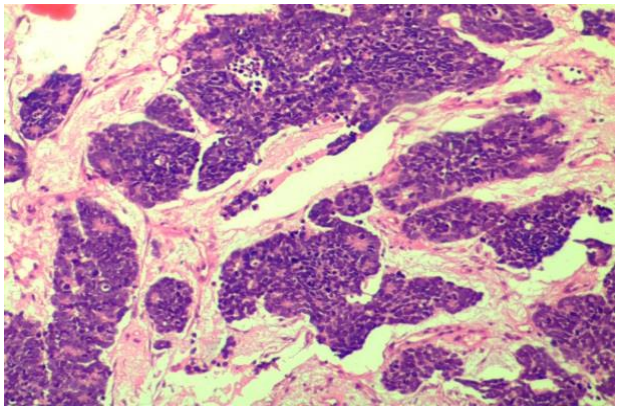
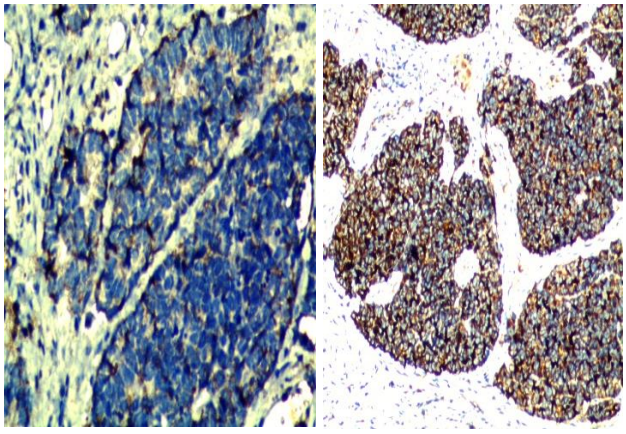


Figure 2: Light microscopy shows monomorphous round cells with salt-and-pepper chromatin arranged mostly in nests with a cribriform pattern that formed rosette-like structures (hematoxylin and eosin stain, original magnification ×100).

These features were suggestive of malignant round cell tumor. Biopsy slide was referred to higher centre for further subtyping of malignant round cell tumor. Immunohistochemical evaluation of the tumour showed a positive immunoreactivity for chromogranin, NSE, and Synaptophysis, (Figure 3) but cytokeratins, desmin, actin, myosin, glial fibrillary acidic protein (GFAP) and calcitonin gene related peptide (CGRP) were negative. The histopathological and immunohistochemical findings conclude a diagnosis of neuroblastoma of the head neck region. Because of remote location of the hospital no further investigations in term of CT scan or MRI could be done. Patient was referred to higher centre in other country hence lost to follow up.



Synaptophysin Chromogranin

Figure 3: Further immunohistochemical analysis shows strong positive staining for chromogranin and synaptophysis. (original magnification ×400).

DISCUSSION

The clinical presentation of neuroblastoma reflects the tumor's primary location and the extent of metastatic disease, if present. The most common primary for neuroblastoma is abdomen which may metastasize to bone, lymph nodes, liver, intracranial, orbital sites, lung, and the central nervous system.[11] Clinically, the neuroblastoma may present with proptosis, periorbital ecchymosis, abdominal distension, bone pain, pancytopenia, fever, anemia, hypertension, paralysis, watery diarrhea, and subcutaneous skin nodule.[12] In children, these signs and symptoms are more severe and widespread (as it metastasize rapidly), whereas in adolescents, there is a greater frequency of metastases to lung or brain.[12] Literature review showed many primary site neuroblastomas spreading to others parts of the body, but none of these patients below 3 years of age presented with large cheek mass.[13] Neuroblastomas have a very broad spectrum of clinical behavior which ranges from spontaneous regression to maturation to a benign ganglioneuroma, or aggressive disease with metastasis leading to death.[14] As per the international classification of neuroblsatoma, the prognosis of childhood neuroblastoma is based on the patient's age at diagnosis.

The best prognosis is awarded to newborn followed by infant and toddler. The children over age five are subjected to poor prognosis.[14]

The clinical presentation in this case are not pathognomonic for neuroblastoma as other lesions such as Ewing's sarcoma, osteogenic sarcoma, histiocytosis X, and osteomyelitis can have similar clinical features. Therefore, for accurate diagnosis emphasis should be made on immunohistochemistry. The pathological diagnosis of the NET in the head and neck area may be difficult just because of the low frequency of these tumors in that location. The diagnosis is based on histological, ultrastructural, and immunohistochemical criteria, which may be overlooked or misdiagnosed especially in small biopsy samples. The diagnosis is based on immunohistochemical proof of a simultaneous epithelial and neuroendocrine differentiation. Immunohistochemical study is also useful to distinguish other malignant small round cell neoplasms that may be considered in the differential diagnosis, as olfactory neuroblastoma, sinonasal undifferentiated carcinoma, basaloid squamous carcinoma, non-Hodgkin lymphoma, and paraganglioma (Table 1).[1,2,9,11,12]

Table 1: Immunohistochemical staining of some malignant round cell tumors.

Stain	Neuroblastoma	ES/PNET	RMS	WIMS
NSE	+++	+	+	+/-
Synaptophysin	+++	+ +	+	-
Chromogranin	++	+	-	-
CD 99	+/-	+ + +	+ + + +	+
Desmin	-	+ + + +	+	+
Myogenin	-	-	+ + + +	+ +
CK	-	-	+ + +	+/-

CONCLUSION

The present case report emphasizes that the proper pathologic identification of primary NET in the head and neck area and their differentiation from squamous cell carcinoma or a metastatic tumor is necessary because prognosis and management of these patients are not the same. Because of the rarity of this entity, an appropriate registry of the cases is highly necessary to gather experience in its management.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Das Ak, Sengupta P, Singh R, Kihemba K. Head neck neuroblastoma in early childhood: a rare case report. Int J Res Med Sci 2017;5:5466-9.