Primitive neuroectodermal Tumors (PNETs) of the Nose: A Case report and a Literature Review

Ahmed Awad Hussein M.D. and Hussam ElBosraty M.D.
Department of Otolaryngology-Head and Neck Surgery
Cairo University School of Medicine, Cairo, Egypt

KASR ALAINY
CAIRO UNIVERSITY - FACULTY OF MEDICINE

Abstract

Objectives:
Primitive neuroectodermal tumors (PNETs) are highly malignant tumors composed of small round cells of neuroectodermal origin that affect soft tissue and bone. Rare in the head and neck. Exhibit pathologic similarities with other small, round cell tumors. We report the second case in the literature of PNET arising in the nose with a review of the literature.
Study Design:
A Case report and literature review
Methods:
We report a 17 years old female patient presented with long history of bilateral nasal obstruction. On examination, a firm and non-painful mass occluding both sides of nasal cavity and expanding causing fascial disfigurement with proptosis of the left eye. High resolution CT was performed and showed a mass filling the nasal cavity infiltrating the left orbit with invasion of both maxillary and ethmoidal sinuses and the hard palate.
Results:
Biopsy was taken and pathology diagnosed Angiofibroma. Pre-operative angiographic embolization was performed then complete excision was done. Immunohistopathology examination of specimen was consistent with (PNET). Radiochemotherapy was introduced to the patient. She was completely cured and free of the disease in the past three years of the follow up.
Conclusions:
With aggressive tumors in the nose, (PNETs) should be considered as a differential diagnosis, immunohistochemistry is essential in the diagnosis. Complete excision with negative margins should be done and affect the prognosis. Surgery followed by radiochemotherapy is the best treatment modality, the orbital location seems to be associated with a particularly better prognosis. Larger studies are needed to assess the nature of (PNETs) and to formulate more effective therapeutic protocol.

Introduction

Primitive neuroectodermal tumors (PNETs) are a group of highly malignant tumors developed from primitive (undifferentiated) nerve cells in the CNS and composed of small round blue cells (Fig 1). This has made classifying this family of tumors challenging and controversial. It is a rare tumor, usually occurring in children and young adults under 25 years of age, with a male predominance. PNET outside the central nervous system is called peripheral PNET (pPNET) developing from migrating embryonal cells of the neural crest. These tumors may be difficult to diagnose due to their primitive morphology but With the advent of immunohistochemical (CD99 positive), cytogenetic and molecular genetic techniques (characteristic translocations involving 22q12) the diagnosis is more accurate.

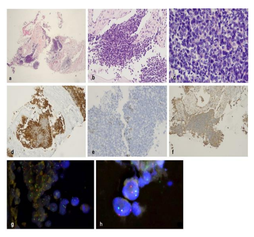

Fig. 1

Fig. 2

Methods and Materials

We report a 17 years old female patient presented with long history of bilateral nasal obstruction. On examination, a firm and non-painful mass occluding both sides of nasal cavity and expanding causing fascial disfigurement with proptosis of the left eye (Fig. 2). There is no any present or past medical history. The patient had no family history of any nasal diseases. The possibility of a tumor of the nasal cavity was considered, for further evaluation of the mass. High resolution CT was performed and showed a mass filling the nasal cavity infiltrating the left orbit with invasion of both maxillary and ethmoidal sinuses and the hard palate (Fig. 3). Wide Transnasal Endoscopic excision of the mass was performed with pathological and Immunohistopathology examination. Literature review was done for this case.

Results

Biopsy was taken and pathology diagnosed Angiofibroma, Pre-operative angiographic embolization was performed then complete excision was done (Fig. 4). The mass appeared grayish white in color (Fig. 5), microscopically the tumor tissue formed of a mixture of fibrocollagenous stroma entrapping variable sized vascular spaces with incomplete or absent smooth muscle wall, which are seen infiltrating bone trabeculae. No evidence of malignancy. Immunohistochemical stain using Envision detection system and DAB chromogen revealed positive staining in the vascular component of the tumor for CD34 and the stromal cellular component for smooth muscle actin. Desmin and S100 are negative which is consistent with PNET. Radiochemotherapy was introduced to the patient. She was completely cured and free of the disease in the past three years of the follow up (Fig. 6). Literature Review of PNETs revealed this reported case is the second case in the literature of PNET arising in the nose.

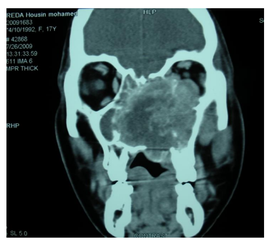

Fig. 3

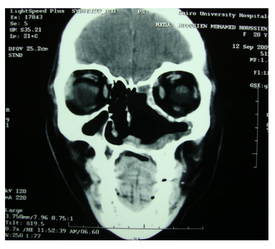

Fig.4

Fig.5

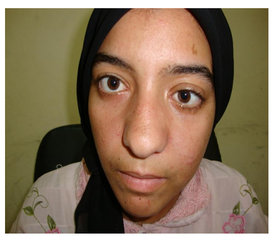

Fig.6

Discussion/Conclusions

With aggressive tumors in the nose, (PNETs) should be considered as a differential diagnosis, immunohistochemistry is essential in the diagnosis. Complete excision with negative margins should be done and affect the prognosis due to the high reliability for local recurrence if incompletely excised. Surgery followed by radiochemotherapy is the best treatment modality, the orbital location seems to be associated with a particularly better prognosis. After reviewing the literature of PNETs, Larger studies are needed to assess the nature of this tumor and to formulate more effective therapeutic protocol.

Contact

Ahmed Awad Hussein
Cairo University
Email: ahmedawadent@gmail.com
Phone:00201000129955

References

1. Weiss SW, Goldblum JR: Primitive neuroectodermal tumors and related lesions. Soft Tissue Tumors. Edited by: Weiss SW, Goldblum JR. 2008, Maryland Heights: Mosby Elsevier, 945-987. 5
2. Shah S, Huh KH, Yi WJ, Heo MS, Lee SS, Choi SC. Primitive neuroectodermal tumor of the maxillary sinus in an elderly male: A case report and literature review. Imaging Sci Dent. 2014;44:307–14.