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.

Methods: A Case report and literature review

Results: 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 fasic 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.

Conclusions: Biopsy was taken and pathology diagnosed Angiofibroma. Pre-operative angiographic embolization was performed then complete excision was done. Immunohistochemistry examination revealed vascular component of the tumor. The patient was completely cured and free of the disease in the past three years of the follow up.

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) 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. Larger studies are needed to assess the nature of (PNETs) and to formulate more effective therapeutic protocol.

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. 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.

References