PERIPHERAL PRIMITIVE NEUROECTODERMAL TUMOR OF THE LEFT MAXILLARY SINUS WITH ORBITAL INVASION: A CASE REPORT

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Purpose: To report a case of peripheral primitive neuroectodermal tumor (PPNET) with initial presentation of a lacrimal sac mass.

Method: Interventional case report.

Result: A 12-year-old female patient presented progressive erythematous swelling of the left medial canthus with tearing. Computed tomography revealed a huge sinonasal tumor with orbital wall destruction. Endoscopic excision of the tumor was performed, and the diagnosis of PPNET was confirmed by pathological findings. Whole body survey did not reveal distant metastasis. She received chemotherapy including Ifosfamide, Cyclophosphamide, Doxorubicin, Etoposide, Vincristine, Dactinomycin with radiotherapy. The tumor lesion gradually subsided and no recurrence was noted during follow-up of more than 3 years.

Conclusion: We presented a rare case of PPNET originating from the left maxillary area with orbital invasion and successfully treated by chemotherapy, and radiotherapy.

Key words: peripheral primitive neuroectodermal tumor, orbital invasion, maxillary sinus

INTRODUCTION

The primitive neuroectodermal tumors (PNET) are generally described as small round cell tumors that show histologic and immunohistochemical evidence of neuroectodermal differentiation[1]. Such tumors are described as a peripheral primitive neuroectodermal tumor(PPNET) when their origin are outside of the central nervous system, with the chest wall, larynx, abdomen, and pelvis being the most common primary sites[2]. The hard and soft tissues of the head, face, and neck are also potential sites[3]. PPNET is the second most common soft tissue malignancy in childhood[4]. It can occur in any age group, however, the peak age of incidence appears to be the adolescent years[2]. We report

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