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PRIMITIVE NEUROECTODERMAL TUMOR OF PARANASAL SINUS: A CASE REPORT

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INTRODUCTION: Primitive neuroectodermal tumor (PNET) is a neural crest tumor¹. It is a rare tumor, usually occurring in children and young adults under 25 years of age. The majority of the cells in the tumor are derived from neuroectoderm, but have not developed and differentiated in the way a normal neuron would, and so the cells appear "primitive. Many of these tumors are given a separate niche on the basis of the assumption that these embryonal tumors also could arise from cells already committed to differentiation. Jones and McGill reported 11 of 26 patients with disease in the head and neck². Most other large published series, however, reveal a paucity of cases in the head and neck region^{3, 4, 5}. Clinical symptoms depend on the site of presentation but invariably include pain and swelling of the surrounding structures due to mass effect.

CASE REPORT: A 6yr old female child presented with complaints of protrusion of right eyeball and right nasal obstruction for a period of two months, it was associated with a history of trauma in the past. Patient underwent a complete clinical examination, CECT PNS and orbit followed by routine investigations including, complete haemogram, HIV, HBsAg and B scan. On examination proptosis of right eye was seen, ocular movements were normal, visual acuity was normal. CT reported a 3.3×2.3 ×4.6 enhancing expansile lesion involving right ethmoidal sinus, right nasal cavity, right maxillary sinus medial wall and nasal septum, erosion of the sinus wall and base of skull noted. The lesion was extending to the orbit and compressing the orbit and into the base of skull and sphenoid sinus. We planned for endoscopic sinus surgery under general anesthesia. Intraoperatively, smooth bulge was noted in middle meatus and posterior ethmoid area, uncinectomy was done ethmoid cavity was opened no air cells were seen, biopsy was taken for histopathological examination. It was reported as round cell tumor. Immunohistochemistry was done positive for MIC-2 and neuron specific enolase and reported as primitive neuroectodermal tumor. Patient was treated with multiagent combination chemotherapy. One year and six months of follow up no recurrence was noted it was confirmed by repeat CECT and nasal endoscopy.

DISCUSSION: Primitive neuroectodermal tumors (PNETs) are group of highly malignant tumors composed of small round cells of neuroectodermal origin that affect soft tissue and bone, found mainly in children and young adults. Stout first described PNET in 1918, as these tumors were thought to arise directly from nerves⁶. The pathologic and cytogenetic understanding of these tumors has significantly advanced over the last 25 years. Based on molecular cytogenetic analysis, primitive neuroectodermal tumors (PNET) are known to share the reciprocal translocations, most commonly between chromosomes 11 and 22. Primitive neuroectodermal tumors (PNET) often exhibit aggressive clinical behavior, with worse outcomes than other small, round cell tumors. Clinical symptoms depend on the site of presentation but invariably include pain and swelling of the

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surrounding structures due to mass effect. Other reported symptoms and signs are site specific, including individual cranial neuropathies, exophthalmos, epistaxis, nasal obstruction, anosmia, neck masses, and headache. In our study patient had proptosis of right eye and right side nasal obstruction for a period of 2 months. Kimber et al, in a review of 26 patients with PNET who were treated with chemotherapy, reported that patients with head and neck PNET had an intermediate prognosis⁷. In this study, primitive neuroectodermal tumor was diagnosed after histopathological examination and immunohistochemistry, it was positive for MIC-2 and neuron specific enolase in IHC. Patient underwent six cycles of multiagent chemotherapy inj. Vincristine 1mg i.v. pulse, inj. doxorubicin 30mg, inj. cyclophosphamide 370mg, inj. actinomycin 20 micro gm. Prognosis of the tumor depends on the presence of metastasis, infiltrative nature, unfavourable location that prevents adequate resection, and young age (less than 4 years). In our study after 18 months of follow up, no recurrence was seen which was confirmed with CECT PNS and nasal endoscopy.

CONCLUSION: Primitive neuroectodermal of paranasal sinus is relatively unusual, it is highly malignant and aggressive tumor which is seen more often in children and young adults. Biopsy and immunohistochemistry helps in diagnosing the tumor, it is managed with multiagent chemotherapy. Early intervention is needed for a good prognosis.

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