A Peripheral Primitive Neuro-ectodermal Tumor (pPNET) of Larynx

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ABSTRACT

Introduction

Primitive neuroectodermal tumors (PNETs) are malignant tumors comprised of small round cells of neuro-ectodermal origin that affect soft tissue and bone. Though the occurrence of pPNETs in the head and neck region is rare, these are aggressive malignant tumors, and long-term survival rates following diagnosis remain poor.

Case Report

In the present case, a tumour was located in larynx (as globular/cystic mass of epiglottis) of the patient and was diagnosed as pPNET. Immunohistochemical analysis indicated that tumor cells were positive for CD99 and NSE, focally positive for EMA but negative for synaptophysin and chromogranin. The mass was surgically excised with negative margins. In post op period patient was planned for post-op chemotherapy and radiotherapy.

Conclusion

pPNETs are very rare in head and neck region. Significant advances in the neoadjuvant and adjuvant chemotherapeutic regimens, as well as improved facility in diagnosing these tumors through cytogenetic and immunohistochemical analysis improves the long-term disease-free survival.

Keywords

Neuroectodermal Tumors, Primitive, Peripheral; Epiglottis

Primitive neuro-ectodermal tumors (PNETs) are malignant tumors comprised of small round cells of neuro-ectodermal origin that affect soft tissue and bone.PNET mainly occurs in the brain, extremities, pelvis, and the chest wall.Approximately, 9% arise in the upper aero-digestive tract or head and neck region, making it the third most common anatomic site, after the extremities and the thoracic/abdominal region.[1]

Here, we present a case study of PNET originating from larynx in a 11 year old girl.

Case Report

A 11 year old girl presented with a progressively increasing difficulty in swallowing with a mild change in voice. There was no associated complaint of difficulty in breathing. On clinical examination a smooth globular mass was coming out behind the base of tongue (Fig.1). Indirect laryngoscopy was not possible due to large size of mass. CECT scan of neck showed a moderately large mass measuring approximately 2.5 x 4.5 cm at level of hyoid bone, not involving the bone, highly suggestive of neoplastic process (Fig. 2). Magnetic resonance imaging neck showed altered signal intensity mass arising from lingual surface of epiglottis, filling vallecula more on left side. Excision of the mass was planned. Findings were confirmed on operating table before excision. The mass was excised from lingual surface of epiglottis by blunt dissection (Figs. 3,4,5,6) and sent for histo-pathological examination. HPE revealed a tumor composed of small cells having round nuclei. Immuno-histochemistry gave a diagnosis of PNET. The tumor was strongly positive for CD99, NSE, and focally positive for EMA(epithelial membrane antigen). It was negative for chromogranin, synaptophysin, cytokeratin (CK). Immunohistochemistry

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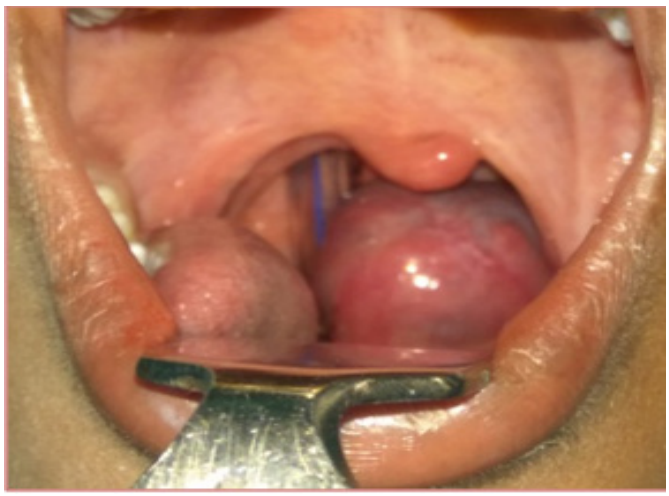

Fig 1. Showing globular mass coming from behind the base of tongue

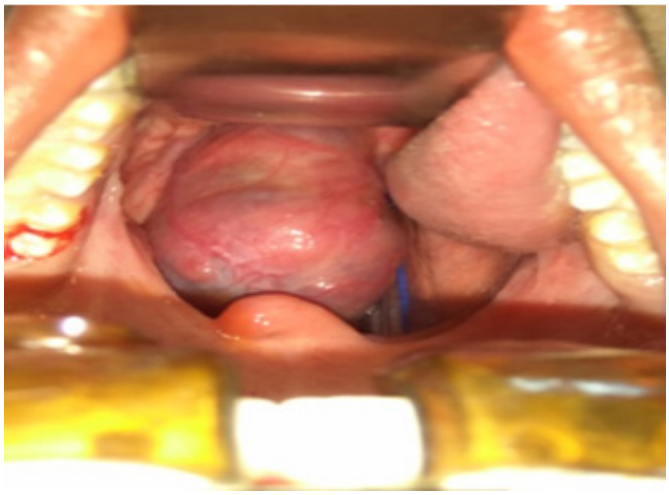

Fig 3. Intra-op photograph showing globular mass after applying Boyle Davis mouth gag.

ruled out small round cell tumor differential diagnoses of rhabdomyosarcoma (desmin negative), small cell carcinoma (CK, synaptophysin and chromogranin negative), basaloid squamous cell carcinoma (CK and p63 negative) and extramedullary round cell tumor (MPO and CD34 negative). Patient was screened for bone, abdomen and pelvic metastasis, was found free of any metastasis. Postoperatively, Patient was planned to give three cycles of chemotherapy comprising of ifosfamide, etoposide and mesna, along with granulocyte colony stimulating factor, with 21 days interval. But patient was lost in the follow up.

Discussion

Primitive neuro-ectodermal tumors are a group of highly malignant tumors composed of small round cells of neuro-ectodermal origin, Primitive neuro-ectodermal tumor family of tumors have been classified into the following three groups based on the tissue of origin.[2]

1. Central nervous system (CNS) PNETs — Tumors derived from the CNS.
2. Neuroblastoma — Tumors derived from the autonomic nervous system.
3. pPNETs — Tumors derived from tissues outside the

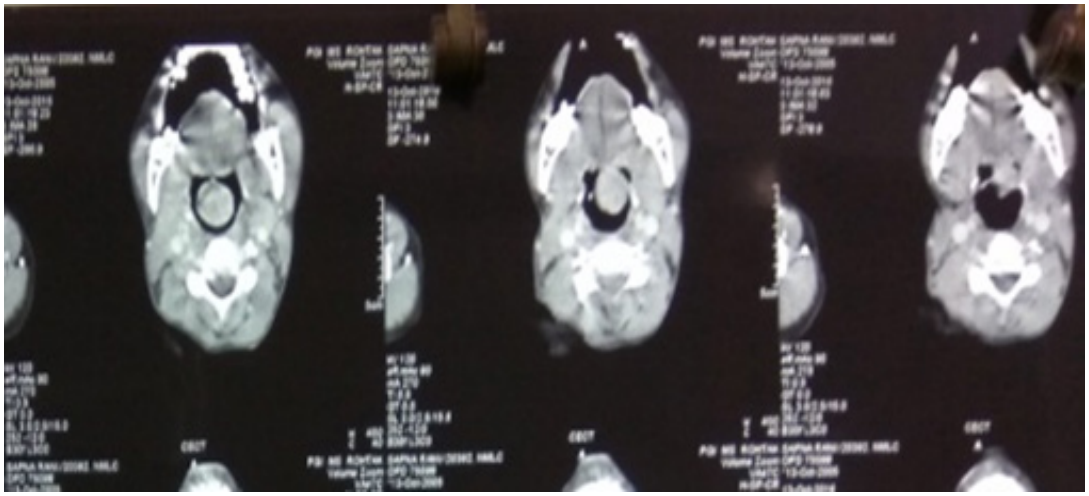

Fig. 2.CECT Scan of neck showing a mass

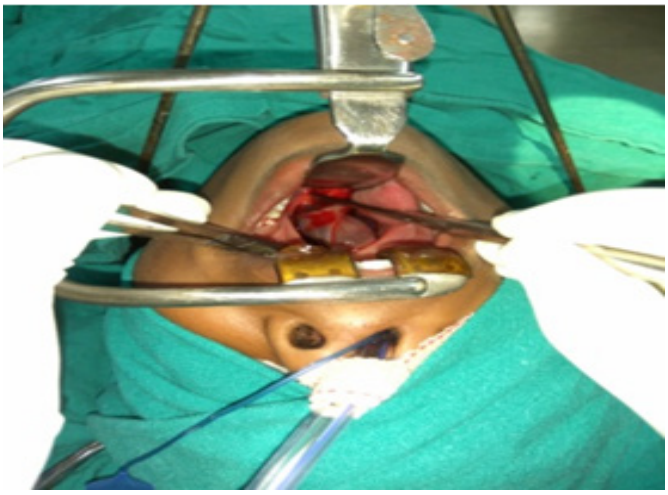

Fig 4. Intra-op photograph showing base of mass while doing blunt dissection.

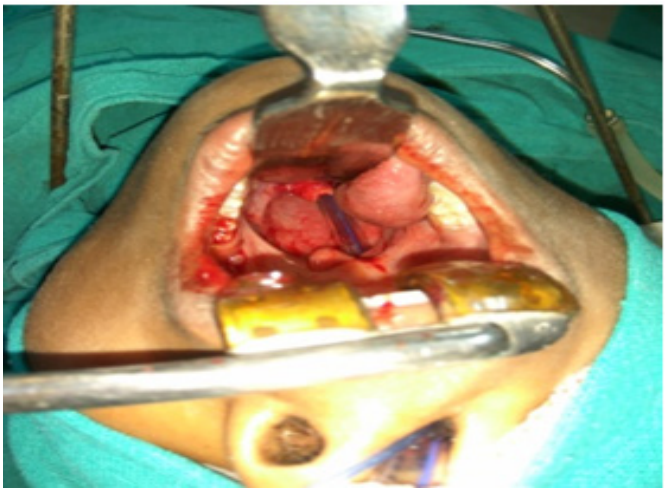

Fig 5. Intra-op photograph showing tip and lingual surface of epiglottis after complete excision of mass.

central and autonomic nervous system. pPNETs are also classified as part of the Ewing family of tumors (EFTs). Primitive neuro-ectodermal tumors and EFTs are referred as synonyms in the literature. Ewing sarcoma, however, is more common in bone, while PNETs are more common in soft tissues.

Immuno-histochemical and cytogenetic studies suggest that all these tumors have a common origin. First described PNETs in 1918, these tumors were earlier thought to arise directly from nerves.[3] Based on molecular cytogenetic analysis, both Ewing's sarcoma and PNETs are known to share the same reciprocal translocations, most commonly between chromosomes 11 and 22. PNETs often exhibit aggressive clinical behavior, with worse outcomes than other small round cell tumors.

Most pPNETs manifest in the thoracopulmonary region (Askin tumor), pelvis, abdomen, and extremities.[4] Rud et al . in a large series of 42 cases of extraosseous Ewing's sarcoma reported few cases in head and neck region.[3] In a series of 26 cases, Jones and McGill reported 11 of 26 patients with disease in the head and neck.

Of the published cases involving the head and neck, the sites of presentation are diverse, including, but not limited to, the paranasal sinuses, jugular foramen, oral cavity, nasal cavity, neck, skull, lingual nerve, parotid gland, larynx, retropharyngeal space, masseter, temporal area, pterygomaxillary space, and orbit. Significant advances in the neoadjuvant and adjuvant chemotherapeutic regimens, as well as improved facility in diagnosing these tumors through cytogenetic and immuno-histochemical analysis, should improve long-term disease-free survival.[4]

Conclusion

Peripheral primitive neuro-ectodermal tumor is an aggressive malignant small round cell tumor that very rarely present in the head and neck, hence timely

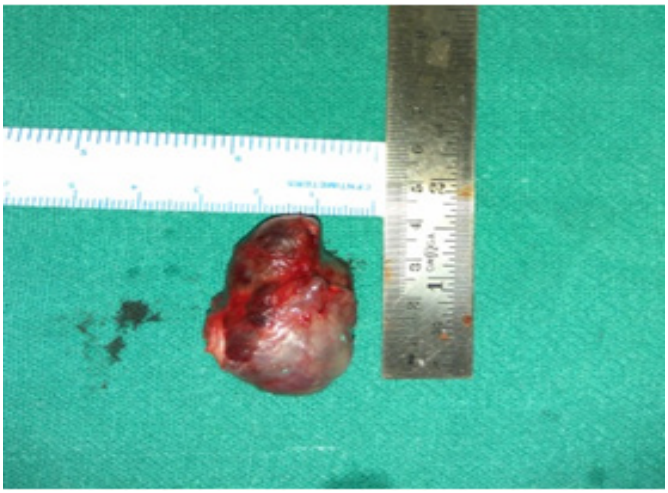

Fig 6. Showing the excised mass.

diagnosis, and treatment is a professional challenge. Long-term survival for patients with PNET is still poor. However, significant advances in the neoadjuvant and adjuvant chemotherapeutic regimens combined with aggressive surgical control of primary disease and in some cases, radiation therapy as well as improved facility in diagnosing these tumors through cytogenetic and immunohistochemical analysis improves the long term disease free survival.[4]

Compliance with ethical standards

FUNDING - no funding.

CONFLICT OF INTEREST - all authors declare that they have no conflict of interest.

ETHICAL APPROVAL - this case report does not contain any studies with animals performed by any of the authors.

ETHICAL APPROVAL - the present clinical report contain a study with human participant.

INFORMED CONSENT - informed consent was obtained from all individual participants included in study.

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