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CASE REPORT

PRIMARY NEUROECTODERMAL TUMOUR OF BASE OF TONGUE – CASE REPORT OF RARE SITE OF ORIGIN

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Abstract

Primitive neuroectodermal tumor (PNET) mostly affects the thoraco-pulmonary region (ASKIN'S tumour), pelvis, abdominal region, and extremities; less frequently, it occurs in the head and neck areas. A 26 year old gentleman presented to the oncology department with the complaints of painful deglutition for 4 months duration and on evaluation with endoscopy, and biopsy and PET- CT scan, he was diagnosed to have proliferative lesion in the posterior third of tongue, and metastatic nodal mass in the neck with disseminated lung, liver and bone lesions. The biopsy was suggestive of PNET with immunohisto-chemistry correlation. He was offered only supportive treatment due to the poor performance status of the patient. Very few cases have been reported in the head and neck region outside the skull and mandible and this is the first case of PNET histology reported in the posterior third of tongue. If non metastatic lesion, the management protocol would include multimodality treatment comprising of chemotherapy, radiation and surgery based on the location.

Keywords: PRIMARY NEUROECTODERMAL TUMOUR, TONGUE, EXTRA SKELETAL ORIGIN, ORAL CAVITY

1 | INTRODUCTION

Ewing's sarcoma (ES) is a highly malignant bone tumor of long bones occurring in children and young adults and was first described by James Ewing in 1921. Extra skeletal ES (EES) is a rare round cell malignant neoplasm with rapid growth and an uncharacterized mesenchymal cell origin, and it is histologically similar to ES arising from bone.[1] Primitive neuroectodermal tumor

(PNET) belongs to the Ewing's sarcoma (ES) family of tumors. PNET mostly affects the thoracopulmonary region (ASKIN'S tumour), pelvis, abdominal region, and extremities; less frequently, it occurs in the head and neck areas.[2,3] Whenever these lesions occur in the head and neck area, they are found arising in the skull or mandible. Very few cases have been reported in the maxilla, nasal cavity, ethmoid, temporal bone, or orbit, parotid.[4,5] Emphasizing on the rarity of the lesion, a unique case of PNET of posterior third of tongue in a young male patient is reported.

2 | CASE REPORT

A 26 year old gentleman presented to the oncology department with the complaints of painful deglutition for 4 months duration. He had noticed growth in the posterior part of tongue 4 months back, which was progressively associated with difficulty in deglutition and he neglected the symptoms initially. Subsequently he developed multiple painful bony swellings over the upper part of sternum, scalp, and right scapula in the last one month and swelling in the left lateral aspect of the neck in the last 2 weeks. He had significant weight loss of more than 30% of his body weight in the last 2 months. On examination, he was emaciated and ECOG performance status – 3. There was a proliferative lesion in the posterior tongue on intraoral assessment and the posterior border of the tumour could not be visualized. There was metastatic matted nodal mass in the left level II, III and IV lymphatic drainage areas. He had multiple painful bony swellings in the right scapular region, manubrium sterni, and the scalp areas.

PET CT scan with 18 F- FDG scan which was done outside, implicated FDG avid lesions in the posterior tongue and the left tonsillar region, in the right posterior parietal bone, right frontal bone, right greater wing of sphenoid bone, multiple cervical, dorsal and lumbo-sacral vertebrae involving body and posterior elements, sternum, head of left humerus, bilateral scapula, anterior aspect of 1^{st} and 5^{th} ribs, bilateral pelvic bones and femur with associated soft tissue components, mediastinal nodes, multiple lung metastases and multiple liver lesions. The endoscopic finding was suggestive of proliferative growth in the left lateral aspect of the posterior $1/3^{rd}$ of tongue. (Fig.2)

Fine needle aspiration of the proliferation growth was highly cellular and showed the presence of tumor cells arranged in sheets with focal rosetting. Individual tumor cells had an indistinct cytoplasm and a centrally placed round to oval hyperchromatic nuclei.(Fig.3A) The biopsy from the primary tongue lesion was highly cellular and shows arrangement of tumor cells in sheets and trabaculae pattern with formation of rosettes. The tumor was predominantly composed of mildly pleomorphic round to ovoid cells with scant cytoplasm and a hyperchromatic nuclei, with striking nuclear molding. (Fig.3B-D) The immunohistochemistry profile of the biopsy was carried out to confirm the diagnosis. The tumor cells were found to be positive for CD99, Fli-1, synaptophysin and negative for Pan CK. (Fig.4 A-C). Based on the above features a final definitive diagnosis of PNET of tongue was given. The diagnosis of disseminated metastatic PNET tongue was confirmed and was offered only supportive treatment due to the poor performance status of the patient.

3 | DISCUSSION

PNET and Ewing's sarcoma are highly malignant tumors of children, adolescents, and young adults. This family of tumors comprise morphologically heterogenous group of tumors that are characterized by a nonrandom chromosomal translocation t(11;22)(q

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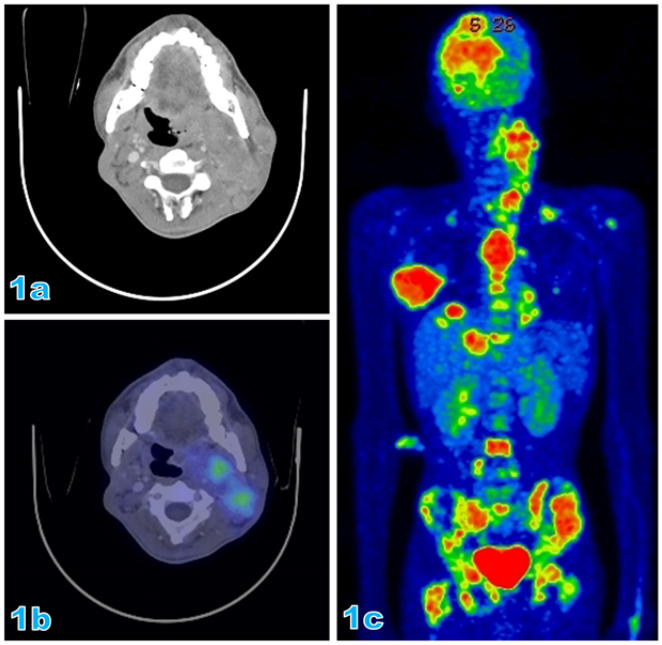

FIGURE 1: 1A:COMPUTED TOMOGRAPHIC AXIAL IMAGE SHOWING GROWTH IN THE BASE OF THE TONGUEEXTENDING TO THE TONSILLAR REGION. FIG 1B: PET CT FUSION IMAGE SHOWING UPATE INTHE BASE OF TONGUE AND TONSILLAR REGION AND NODAL UPTAKE AT LEVEL II LYMPHNODES. FIG 1C: PET CT FUSION IMAGE SHOWING MULTIPLE METASTATIC SITE UPTAKEALONG WITH THE PRIMARY UPTAKE IN THE TONGUE.

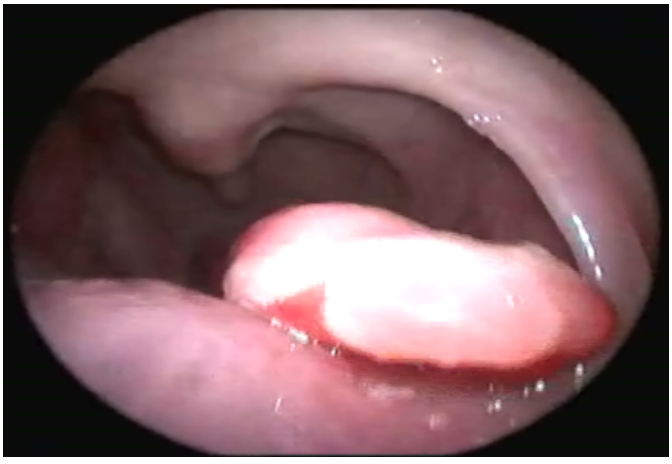

FIGURE 2: ENDOSCOPIC PICTURE SHOWING PROLIFERATIVE LESIONS ATTHE BASE OF THE TONGUE ON THE LEFT LATERAL ASPECT

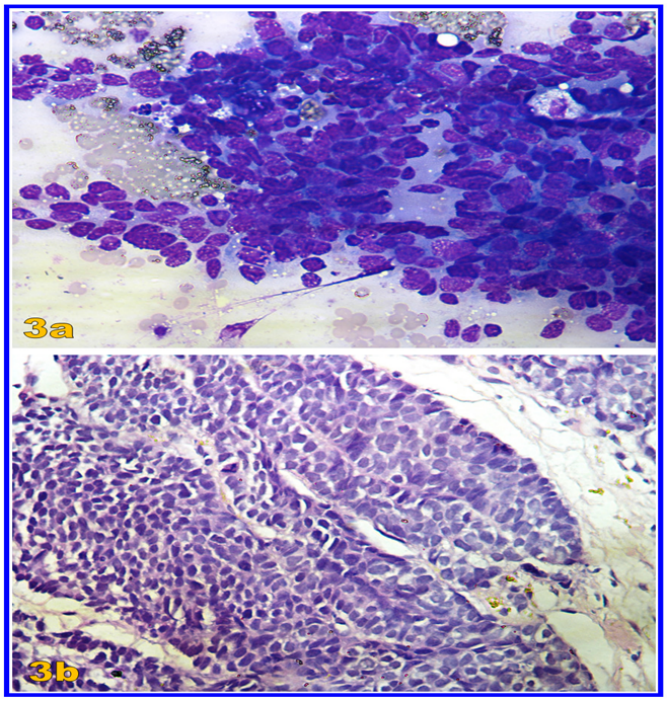

FIGURE 3: 3A: PHOTOMICROGRAPHOF FINE NEEDLE ASPIRATION CYTOLOGY SMEARS OF TONGUE MASS SHOWING ROUND TO OVALCELLS WITH INDISTINCT CYTOPLASMIC BORDERS AND HYPERCHROMATIC NUCLEUS. HOMERWRIGHT ROSETTE IS SEEN TOWARDS THE LEFT CORNER (400X, MGG). FIG 3B: PHOTOMICROGRAPH OF TUMOR CELLSARRANGED IN TRABACULAE PATTERN. INDIVIDUAL TUMOR CELLS HAVE ROUND TO OVALNUCLEUS WITH NUCLEAR MOULDING. (400X,H&E)

24;q 12) leading to the formation of the EWS-FLI-1 fusion protein. This contributes to the pathogenesis of these tumors by modulating the expression of target genes.[6] The peripheral PNET (pPNET) is part of the Ewing's sarcoma family of tumor, which includes Ewing's tumor of bone, extraosseus Ewing tumor (tumor growing outside of the bone), PNET, and Askin tumor (PNET of the chest wall). Approximately 9% of this tumor arises in the upper aerodigestive tract or head and neck region, making it the third most common anatomic site, after the extremities and the thoracic/abdominal region.[7]The actual incidence of PNET in the head and neck region is difficult to ascertain and they are primarily located in the mandible. Very few case reports are available of the tumour originating from the tongue. [8,9]

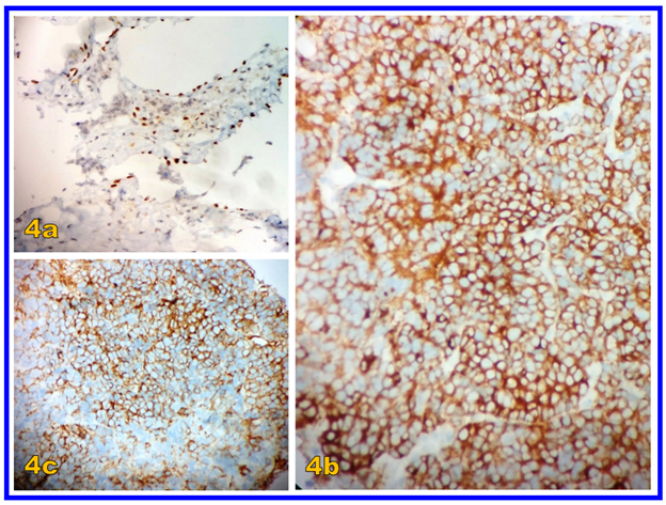

FIGURE 4: 4A:PHOTOMICROGRAPH SHOWING THE IMMUNOHISTOCHEMISTRY OF STRONG NUCLEAR POSITIVITYOF FLI 1(400X, H&E) FIG 4B: PHOTOMICROGRAPHSHOWING THE IMMUNOHISTOCHEMISTRY OF STRONG CYTOPLASMIC AND MEMBRANOUSPOSITIVITY OF SYNAPTOPHYSIN (400X, H&E) FIG 4C: PHOTOMICROGRAPH SHOWING THE IMMUNOHISTOCHEMISTRY OFSTRONG MEMBRANOUS POSITIVITY OF CD99 (400X, H&E).

Nikitakis et al in his observation of 43 patients with pPNET in the head and neck region, revealed a predilection among the children and adolescents.[3] Histologically, the most helpful diagnostic feature is the presence of rosettes, usually of the Homer–Wright type, which indicate neural differentiation.[9] Immunohistochemistry analysis of the tumour helps in the confirmation of the diagnosis. Additional studies such as cytogenetic study often support the final diagnosis. Peripheral PNETs are highly aggressive and have a propensity for local recurrence and metastasis to the lung, bone, and bone marrow.[6] After a tissue diagnosis has been made, the patient should undergo a full metastatic work-up, including a chest x-ray, CT of the lungs, a bone scan, and bone marrow aspiration, to ascertain whether the tumor has metastasized in all cases.

Aggressive treatment strategies using multi modality treatment including chemotherapy, radiation and surgery play an important role in the treatment of non metastatic PNET. Surgery followed by radiation therapy for ablation of residual microscopic disease and chemotherapy seems to be the appropriate treatment for the patients with localized disease, although the surgical options in difficult access site like in base of tongue are limited. Effective local control requires negative surgical margins. Recommendation for radiation is dependent on the primary site and the size of the tumour, histology, the patient's age and the extent of disease before and after surgical resection. Radiation is generally administered as an adjuvant therapy when surgical excision is incomplete, but it can also be used as a primary treatment for unresectable lesions.[6,] The primary modality of the treatment in the metastatic disease includes the chemotherapy in palliative intent and they are rarely curable except in rare scenarios.[6] ES is unique among sarcoma in that primary and metastatic diseases can respond dramatically to initial therapy with robust initial responses predicting a better outcome. PET/CT can play an important role in treatment response assessment because patients can show metabolically inactive tumor with reduced bulk.[1] In our case, the disease was found to be disseminated and the patient performance status was not sufficient for any form of oncologic treatment.

4 | CONCLUSION

Peripheral primitive neuroectodermal tumors are rare but they do occur at unusual locations like base of the tongue. The possibility of PNET should be kept in mind while evaluating lesions occurring on tongue, which behave differently than the other common tumours in the oral cavity. **(1–9)**

REFERENCES

1. Sandhya KN, Sangeetha KP, Balan A, Girija KL, Bose T. Extraskeletal Ewing's Sarcoma of Floor of Mouth: A 1-year follow-up of the Rare Disease in a Rare Location. Int J Sci Stud. 2017;4(11):220–223.

2. Margaix-Munoz M, Bagan J, Poveda-Roda R. Ewing sarcoma of the oral cavity. A review. Journal of Clinical and Experimental Dentistry.

2017;9(2):0–0. Available from: https://dx.doi.org/10.4317/jced.53575. doi:10.4317/jced.53575 .

3. Ulutin C, Cetinayak O, Aksu G, Fayda M, Ataergin S, Beyzadeoğlu M. Malignant peripheral primitive neuroectodermal (pPNET) of tongue. Auris Nasus Larynx. 2007;34(1):115–118. Available from: https://dx.doi.org/10.1016/j.anl.2006.11.001. doi:10.1016/j.anl.2006.11.001.

4. Sun G, Li Z, Li J, Wang C. Peripheral primitive neuroectodermal tumor of the maxilla. Br J Oral Maxillofac Surg. 2007;45:226–233.

5. Nikitakis NG, Salama AR, O'Malley BW, Ord RA, Papadimitriou JC. Malignant peripheral primitive neuroectodermal tumor-peripheral neuroepithelioma of the head and neck: A clinicopathologic study of five cases and review of the literature. Head & Neck. 2003;25(6):488–498. Available from: https://dx.doi.org/10.1002/hed.10260. doi:10.1002/hed.10260.

6. Gosavi S, Prashant R, Jain D, Mahajan G, Karodpati N, Shinde V. Extraosseous Ewing′s tumor of larynx: A rare presentation. Medical Journal of Dr DY Patil University. 2015;8(4):553–553. Available from: https://dx.doi.org/10.4103/0975-2870.160821. doi:10.4103/0975-2870.160821.

7. Deb RA, Desai SB, Amonkar PP, Aiyer PM, Borges AM. Primary primitive neuroectodermal tumour of the parotid gland. Histopathology. 1998;33(4):375–378. Available from: https://dx.doi.org/10.1046/j.1365-2559.1998.00535.x. doi:10.1046/j.1365-2559.1998.00535.x.

8. Joseph L, Kumar R, Senthil K, Ravindran C, Prathiba D. Primitive neuroectodermal tumor of the maxilla. Journal of Oral and Maxillofacial Pathology. 2008;12(2):64–64. Available from: https://dx.doi.org/10.4103/0973-029x.44580. doi:10.4103/0973-029x.44580.

9. Shah S, Huh KH, W Y. Primitive neuroectodermal tumor of the maxillary sinus in an elderly male: A case report and literature review. Imaging Sci Dent. 2014;44:307–321.

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