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Case Report

Ewing's sarcoma or peripheral primitive neuroectodermal tumor at the base of tongue: a rare location case report

Pinki Devi[1]*, Anjali[1], Nidhi Yadav[1], Sunita Singh[1], Manoj K. S. Tomer[2]

[1]Department of Pathology, Pt B. D. Sharma PGIMS, Rohtak, Haryana, India
[2]Department of Anaesthesia, Noble Heart Hospital, Rohtak, Haryana, India

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***Correspondence:**
Dr. Pinki Devi,
E-mail: drpinkitmr@gmail.com

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ABSTRACT

Peripheral primitive neuroectodermal tumor now termed as Ewings sarcoma/PNET, representing a family of tumors with varying degrees of neuronal differentiation and genetic rearrangements. Most common sites are extraosseous involving trunk and lower limb. Base of tongue is a very rare site for Ewing's/PNET, hence in view of rarity of the location as an uncommon presentation, the case has been reported for the better understanding and supporting the literature with the similar finding. A 68 years old female patient presented with the complaint of difficulty in swallowing since 3 months. On examination growth was identified at the left side base of tongue. Biopsy was processed and stained with H&E and other relevant markers. Differentials on the basis microscopic examination were lymphoma, neuroendocrine tumor and primitive neuroectodermal tumor. Various immunohistochemical markers to rule out the given differential were used. The tumor was positive for CD 99 and vimentin. The case was repored as PNET. Peripheral PNET is a challenging topic. Wide range of extraosseous locations have been observed but head and neck being less reported needs to be studied for understanding the behavior of this highly malignant disease in this rare location so that patient can be benefitted by advanced multimodality treatments including surgery, chemotherapy and radiotherapy.

Keywords: Primitive neuroectodermal tumor, Ewing's sarcoma, Extraosseous, Immunohistochemical

INTRODUCTION

Peripheral primitive neuroectodermal tumor (PNET) has rapid advances in past few years and is now termed as Ewings sarcoma/PNET, representing a family of tumors with varying degrees of neuronal differentiation and genetic rearrangements. Most common sites are extraosseous involving trunk and lower limb. Most commonly seen between the ages of 10 to 30 years. Base of tongue is a very rare site for Ewing's sarcoma/PNET, hence in view of rarity of the location as an uncommon presentation, the case has been reported for the better understanding and supporting the literature with the similar finding. A 68 years old female patient presented to the surgery OPD with the complaint of difficulty in swallowing since 3 months. On examination growth was identified at the left side base of tongue. Biopsy was taken and received in 10% formalin in the department of pathology. Grossly, the growth was gray white measuring 1.0×1.0×0.3 cm. Biopsy was processed and stained with H&E and other relevant markers.

CASE REPORT

Microscopic examination revealed stratified squamous epithelial lined soft tissues showing sheets of round to oval cells with fine chromatin, inconspicuous nucleoli, scant cytoplasm (Figure 1). Differentials on the basis of morphology of tumor cells were lymphoma, neuroendocrine tumor and primitive neuroectodermal tumor. Various immunohistochemical markers to rule out the given differential were used (Table 1). The tumor was

positive for CD 99 and vimentin (Figure 2). The case was reported as PENT.

Table 1: Reactivity of various immunohistochemical markers.

S. no.	IHC marker	Reactivity
1	CD99	Positive
2	Vimentin	Positive
3	CK	Negative
4	EMA	Negative
5	CD3	Negative
6	CD19	Negative
7	CD20	Negative
8	CD10	Negative
9	CD56	Negative
10	Chromogranin	Negative

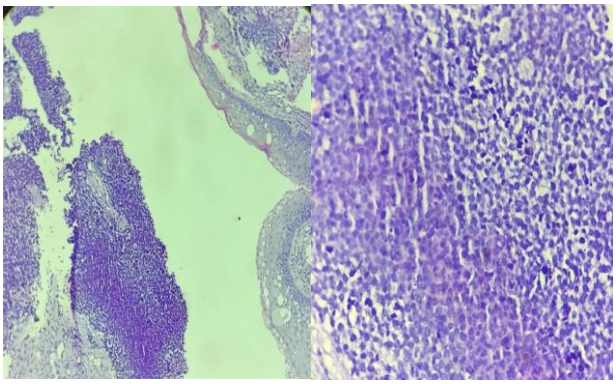

Figure 1: Stratified squamous epithelial lined soft tissues showing sheets of round to oval cells with fine chromatin, inconspicuous nucleoli, scant cytoplasm (H&E, 10X and 40X).

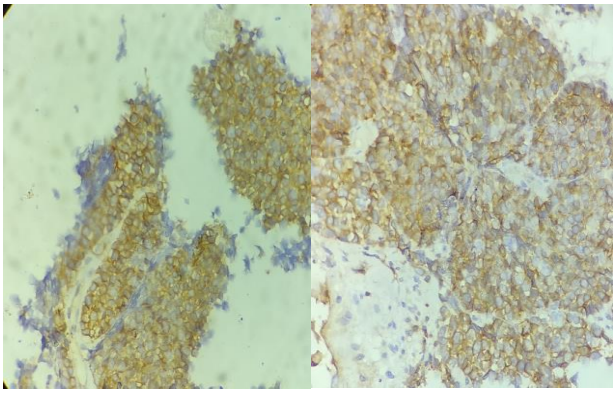

Figure 2: Imunohitochemically, tumor was positive for CD99 and vimentin (40X).

DISCUSSION

Extraskeletal Ewing's sarcoma/PNET is a malignant small round cell neoplasm of undifferentiated mesenchymal origin.[1] James Ewing first described Ewing's sarcoma (ES) in 1921.[2] Later Tefft described its extraosseous variant in 1969.[3] EES/PNET of the head and neck region is extremely rare. Not much literature is available in this context. Sandhya et al has reported a case of sublingual gland showing extra osseous Ewing's sarcoma.[4] We could not find any case of ES/PNET at base of tongue in literature search.

Ewing sarcoma/PNET shows a predilection for males with the ratio of 1.4 to 1, whereas EES/PNET have equal predilection for either sex.[5,6] Nearly 80% of patients are younger than 20 years, and the peak age incidence is during the second decade of life.[5] The most frequent extraosseous site described are extremities; commonly the lower limbs. Other common sites are the head, pelvis and paravertebral region.[7]

Non specificity of radiological features and histological features as are shared by many other round cell tumours makes the diagnosis of this entity more difficult.[8]

Histopathologically, tumour morphology is variable, mostly composed of uniform small round cells with round nuclei containing fine chromatin, scanty clear or eosinophilic cytoplasm, and indistinct cytoplasmic membranes, few showing larger cells, having prominent nucleoli, and irregular contours. Spindle cell morphology of tumor cells is rare in soft tissue ES. Immunophenotypically, CD99 is expressed in membranous fashion. Vimentin stains most tumour cells. Neural markers are frequently expressed and keratin is positive in some cases.[5]

The diagnosis of ESS is challenging, needs to confirmed by advanced diagnostic modalities like molecular analysis along with histopathologic and immunohistochemical studies.

The prognosis in Ewing sarcoma/PNET has improved in the modern era of treatment and current survival rate is estimated to be 41%. Important prognostic features include the stage, anatomic location and the size of the tumour.[5]

CONCLUSION

Peripheral PNET being a challenging topic, a lot of work has been done and rapid advances have been seen in past few years and it was then termed as ES/PNET. Wide range of extraosseous locations have been observed but head and neck being less reported needs to be studied for understanding the behavior of this highly malignant disease in this rare location. Advanced multimodality treatments including surgery, chemotherapy and radiotherapy can be beneficial to patient if the diagnosis of this entity at these rare sites is reported timely with knowledge of this entity being present in literature by case report contribution.

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Ethical approval: Not required

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