Case Report

Embryonal rhabdomyosarcoma: A rare oral tumor

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Abstract Rhabdomyosarcoma is the malignant neoplasm of striated muscle and a relatively uncommon tumor of the oral cavity. Embryonal variety is the most common subtype, observed in children below 10 years of age but occasionally seen in adolescents and young adults. The present report describes a case of embryonal rhabdomyosarcoma in the left posterior buccal mucosa, with extension in the adjacent alveolus, soft palate, oropharynx and nasopharynx of a 17-year-old female.

Key Words: Embryonal, rhabdomyosarcoma, striated muscle

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INTRODUCTION

Rhabdomyosarcoma is a malignant neoplasm derived from primitive mesenchyme that retains the capacity for skeletal muscle differentiation and thus often arises at sites where skeletal muscle tissue is normally absent (e.g. urinary bladder), or in areas where striated muscle is scanty (e.g. nasal cavity and middle ear). The most common site is head and neck (parameningeal and orbit), followed by genitourinary tract. Less commonly involved sites of head and neck regions are nasal cavity and nasopharynx, ear, paranasal sinuses, soft tissue of face and neck and finally the oral cavity, including the tongue lip and palate.[1-3] There are four subtypes: Pleomorphic, alveolar, embryonal and botryoid. Embryonal variety is most common and constitutes approximately 49% of all rhabdomyosarcomas.[4] It mostly affects children below 10 years of age but may also affect adolescents and young adults. In the present case, buccal mucosa is involved in an adolescent, which is very rare. Only 17 cases have been reported in the indexed journals till date involving intraoral mucosa according to our review of the literature.[5-21] Intraoral site involvement in a patient nearer to adult age is extremely rare, and the manner by which panel of immunohistochemical markers had been used to confirm the diagnosis makes this case unique.

CASE REPORT

A 17-year-old female patient reported to the Department of Oral Pathology with a mass involving the left side of the mouth for the last 1½ months. The lesion was stated to be fast enlarging, painless and caused occasional spontaneous bleeding. Left middle third of the face showed slight swelling. On intraoral examination, a diffuse fleshy mass was seen involving left posterolateral part of the palate, left upper retromolar region and adjacent buccal mucosa, measuring about 3 cm × 3 cm [Figure 1]. Overlying mucosa was smooth and streaks of clotted blood was present. On palpation, the lesion was firm, mildly tender and seemed to be extending inferiorly, laterally as well as medially. A provisional diagnosis of connective tissue neoplasm was made. Orthopantomogram showed no bony abnormality except faint soft tissue shadow

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behind maxillary second molar and impacted left maxillary canine. Contrast-enhanced computed tomography showed soft tissue lesion in the parapharyngeal space both causing mass effect on oropharynx and nasopharynx with extension to the cheek causing erosion of mandible [Figure 2]. Incisional biopsy was taken from the lesion after having written informed consent of her parents. Biopsy specimen was processed and sections were stained with H&E. On histopathological examination, normal stratified squamous surface epithelium was present. Underlying connective tissue contained the tumor mass consisting of clusters of primitive spindle and round cells with dark staining nuclei and little cytoplasm arranged in abundant myxoid stroma [Figure 3]. Blood vessel invasion was observed [Figure 4] and hematogenous metastasis was suspected. Mitotic figures and typical tadpole nuclei were also present [Figure 5], but characteristic cross striation was not found. In other areas, primitive oval cells and scanty rhabdomyoblasts with eccentric vesicular nuclei were seen [Figure 6]. The lesion was further provisionally diagnosed as malignant round cell tumor compatible with embryonal rhabdomyosarcoma. For confirmation of diagnosis, immunohistochemical staining was performed.

The lesion showed positive reactivity with desmin [Figure 7], MyoD1 [Figure 8], myogenin [Figure 9][22,23] and muscle-specific actin [Figure 10]. It also showed negative reactivity with cytokeratin, epithelial membrane antigen, Mic-2, synaptophysin, CD34, CD31 and littoral cell angioma.

Immunohistochemical reports of this case are given in Table 1 for better understanding.

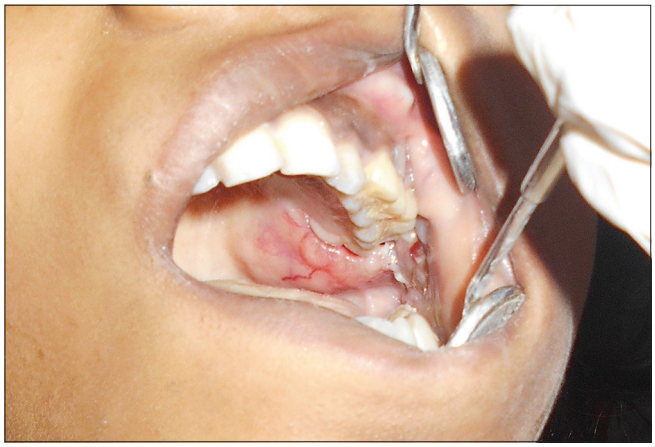

Figure 1: Diffuse swelling involving left maxillary tuberosity, adjacent part of palate and buccal mucosa

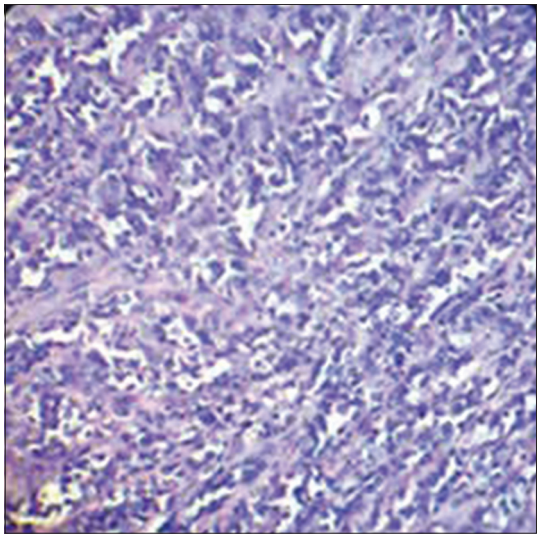

Figure 3: Round and spindle-shaped tumor cells with hyperchromatic nuclei and little cytoplasm.(H&E stain, ×100)

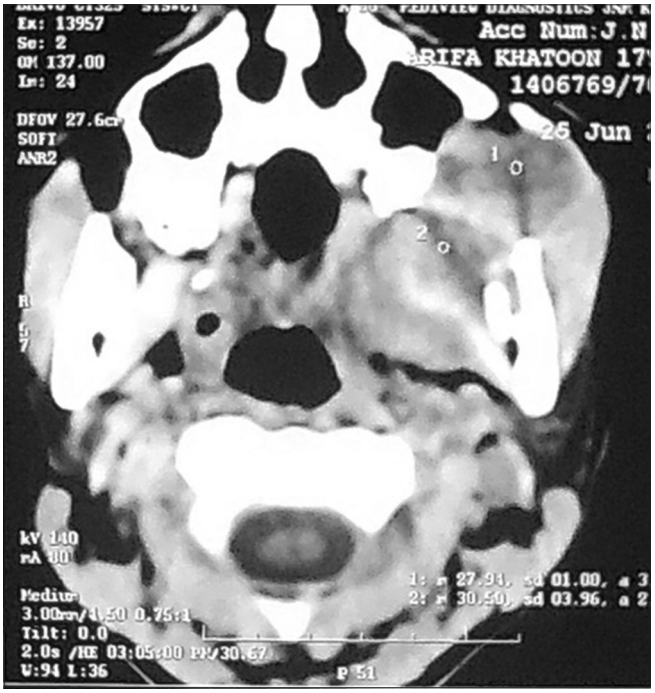

Figure 2: Contrast-enhanced computed tomography shows space occupying lesion involving oropharynx, nasopharynx, parapharyngeal space with extension to the cheek and mandible

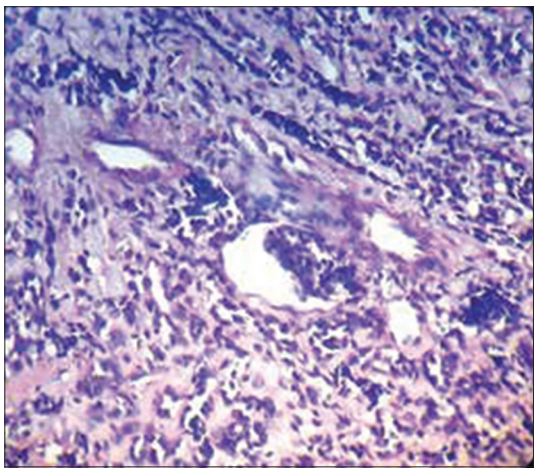

Figure 4: Tumor cell invasion into blood vessel (H&E stain, ×200)

DISCUSSION

According to the available case reports, it is found that mandibular gingiva is more commonly affected than maxillary gingiva. Few reports of intraosseous involvement are there, one of which involved angle of the mandible and ascending ramus.[24] Palatal lesions mostly involve the soft palate, and few of them involved the uvula. Congenital lip[25] and tongue[26] lesions have been reported. Floor of the mouth lesions are extremely rare.[27] This malignancy is most prevalent in the first and second decades of life, gradually decreases with increasing age and rarely affects a person in seventh or eighth decade of life.[28] Findings from Table 1 helped us to confirm the diagnosis as a case of rhabdomyosarcoma. The histopathological report was of

Table 1: Results of immunohistochemical stains performed

IHC marker used	Marker for the tumor	Result obtained
MyoD1	Rhabdomyosarcoma	Positive
Desmin	PNET, neuroblastoma rhabdomyosarcoma, leiomyosarcoma	Positive
Myogenin	Rhabdomyosarcoma	Positive
MSA	Malignant melanoma Rhabdomyosarcoma Breast carcinoma	Positive
Cytokeratin	Carcinoma	Negative
EMA	Carcinoma	Negative
Mic-2	Ewing's sarcoma, PNET	Negative
Synaptophysin	Neuroendocrine tumor	Negative
CD34 and CD31	Angiosarcoma and other endothelial cell tumors	Negative
LCA	Lymphomas	Negative

IHC: Immunohistochemistry, PNET: Primitive neuroectodermal tumor, LCA: Littoral cell angioma, EMA: Epithelial membrane antigen, MSA: Muscle-specific actin

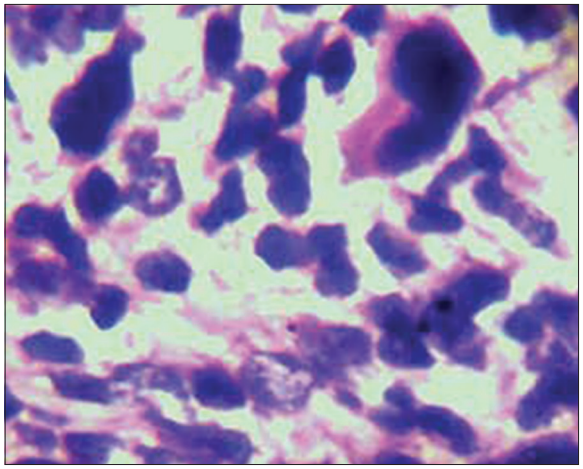

Figure 5: Tumor cells with tadpole nuclei (H&E stain, ×1000)

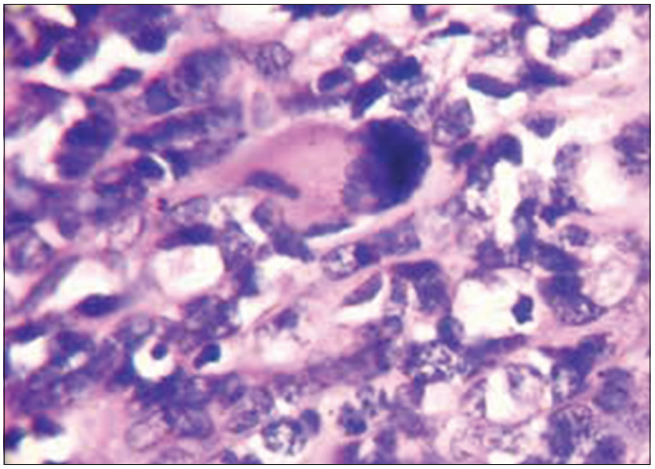

Figure 6: Neoplastic rhabdomyoblast cell (H&E stain, ×1000)

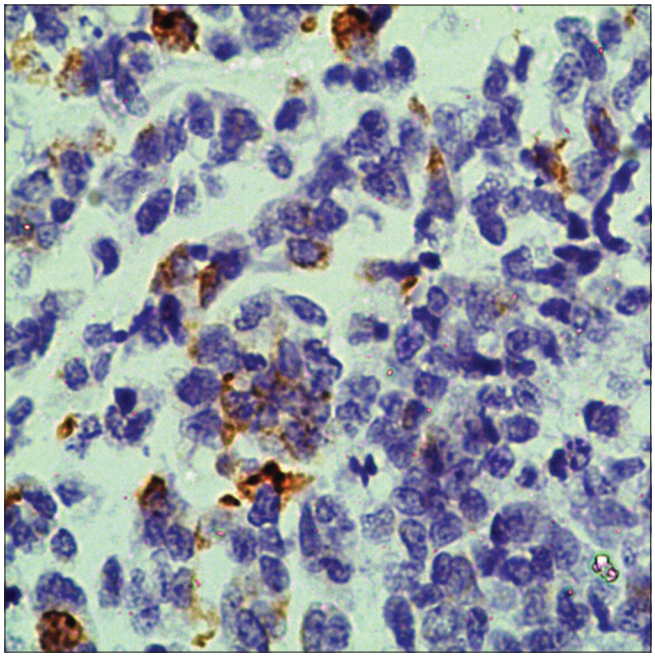

Figure 7: Immunohistochemical staining for desmin shows positive result (IHC stain, ×400)

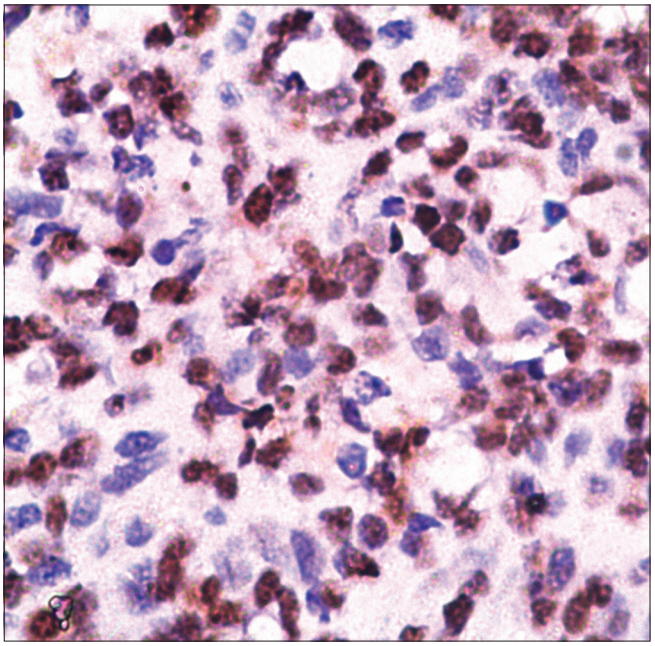

Figure 8: Immunohistochemical staining for MyoD1 shows focal positive result (IHC stain, ×400)

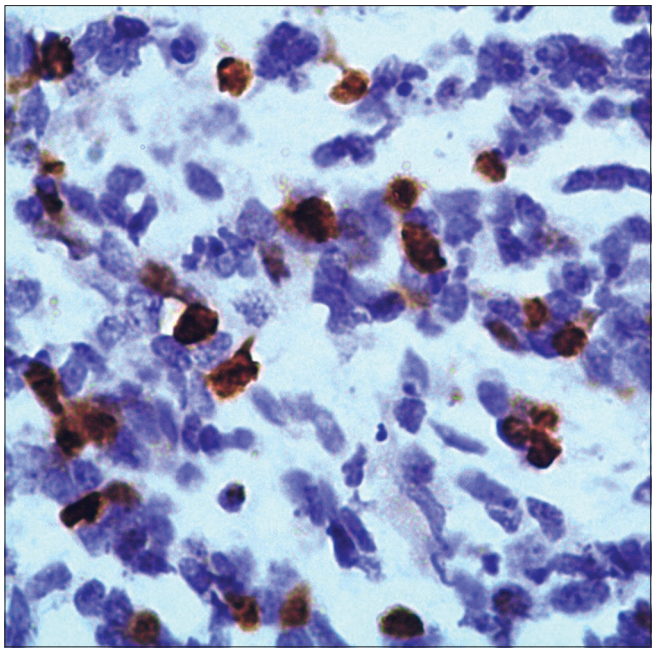

Figure 9: Immunohistochemical staining for myogenin shows positive result (IHC stain, ×400)

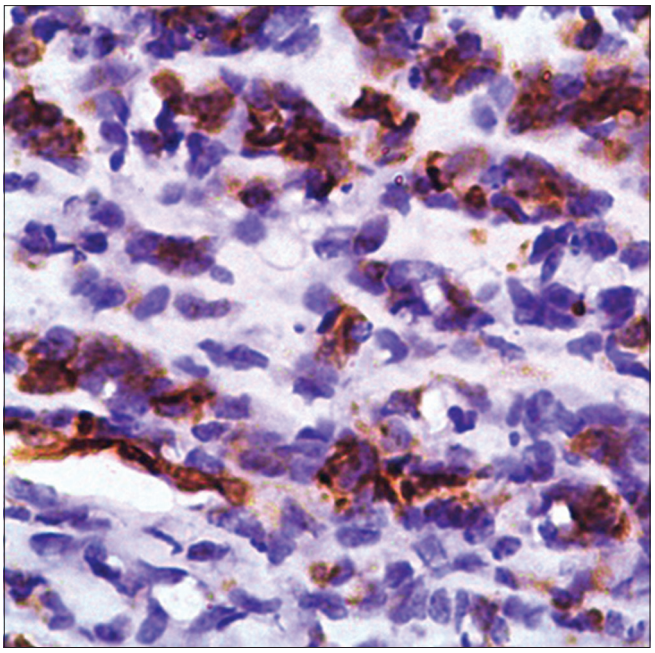

Figure 10: Immunohistochemical staining for muscle-specific actin shows positive result (IHC stain, ×400)

round cell tumor, and so, we had to exclude all other round cell malignancies. Considering the histopathological pattern and age of the patient, we arrived at the final diagnosis of embryonal rhabdomyosarcoma.

Embryonal rhabdomyosarcoma is a rare lesion of the oral cavity. Histopathological (H/P) diagnosis always must be confirmed by immunohistochemical investigation as the histological pattern is variable and poorly differentiated tumors bear resemblance with many other round cell malignancies.

Prognosis of the lesion depends on age of the patient, anatomic site, clinical staging (tumor size, node involvement and metastasis) and H/P subtype. Twenty percent of patients develop metastasis at the time of diagnosis; and lung, lymph node and bone marrow are the common sites.[29,30] Rhabdomyosarcoma is treated by complete resection of the lesion, followed by multi-agent chemotherapy with or without radiotherapy.[31,32] Classic embryonal rhabdomyosarcoma has an intermediate prognosis; 5-year survival rate is around 75%.[33,34] The patient was referred to a cancer specialty hospital.

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Conflicts of interest

There are no conflicts of interest.

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