

## Oral Rhabdomyosarcoma: A Case Report

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### Abstract

Rhabdomyosarcoma (RMS) is a malignant soft tissue neoplasm of skeletal muscle origin. The most common sites of occurrence are the head and neck, genitourinary tract, and extremities. Although Rhabdomyosarcoma has a relative predominance for head and neck region, it is less frequent in oral cavity, and accounts for only 0.04% of all head and neck malignancies. Some studies have been showed that soft palate and tongue are the most common sites for oral RMS. We present a case of oral Rhabdomyosarcoma in a 15 year old girl, and demonstrate the clinical, radiological, histological, and immunohistochemical features of this neoplasm.

**Keywords:** Embryonal Rhabdomyosarcoma; Muscle tumor; Oral; Sarcoma

### Introduction

Rhabdomyosarcoma (RMS) is a malignant soft tissue neoplasm of skeletal muscle origin. It is the most common soft tissue sarcoma in children under 15 years of age, and rare among people older than 45 years of age. The most common sites of occurrence are the head and neck (40%), genitourinary tract (25%), and extremities (20%) [1]. Rhabdomyosarcoma are anatomically divided into 2 categories including parameningeal and nonparameningeal.

Parameningeal sites are the nasal cavity, nasopharynx, paranasal sinuses, middle ear, mastoid, infratemporal fossa, and pterygopalatine fossa. The scalp, orbit, parotid gland, oral cavity, oropharynx, and larynx are considered non parameningeal sites [2]. Oral Rhabdomyosarcoma is rare neoplasm, and accounts for only 0.04% of all head and neck malignancies [3].

Three main types histologic appearance of RMS are embryonal, alveolar and pleomorphic. The embryonal type presents the subtypes including classic, spindle cell and botryoid [4]. The embryonal type of RMS is the most common variant, and occurs in children between birth and 15 years old. The embryonal RMS contains a mixture of spindle and undifferentiated round cells and immature striated muscle like cells (rhabdomyoblasts) with abundant eosinophilic cytoplasm either tightly or loosely packed in a myxoid stroma. The botryoid tumor is a morphologic variety or subtype of the embryonal type and because of gross resemblance to cluster of grapes have been named botryoid. Histologically a multilayered band of cells just below the surface epithelium called the cambium layer is a special finding of this variant. The alveolar type presents in the older children (10 to 25 years old) and contains clusters of small round cells with hyperchromatic nuclei and eosinophilic cytoplasm separated by fibrovascular septa. Pleomorphic type occurs in older people between the age group of 50-60 years and is characterized histologically by large bizarre cells of racquet, strap, ribbon and spindle types in a haphazard arrangement [5-7].

The histogenesis of RMS is still indistinct, but the most widely believed hypothesis is that RMS arises due to proliferation of embryonic mesenchymal cells that exhibit a tendency toward myogenic differentiation and probably originate from satellite cells associated with the embryogenesis of skeletal muscle [8-10].

In this report, we present a case of oral Rhabdomyosarcoma in a 15 year old girl, and demonstrate the clinical, radiological, histological, and immunohistochemical features of this neoplasm.

### Case Report

A 15 year old female was referred to the department of oral and maxillofacial surgery of dental school of Isfahan university of medical sciences in 9 mars 2013 with a chief complaint of painless swelling in the mandibular left posterior region of 40 days duration. The swelling was initially small size and quickly increased (progressive growth). She had not a past history of trauma and systemic disease. But, there was a history of weight loss before 2 month ago. According to the patient statement, there was no sign of pain, sensation disorders, pus discharge, and bleeding in the involved area. Extra oral examination showed facial asymmetry, and a diffuse, soft to firm, tender swelling in the posterior region of the mandible (Figure 1).

The left submandibular lymph nodes were palpable, firm, mobile and non-tender. Additionally, she had limitation in maximum opening.



Figure 1: Swelling in left mandibular region.

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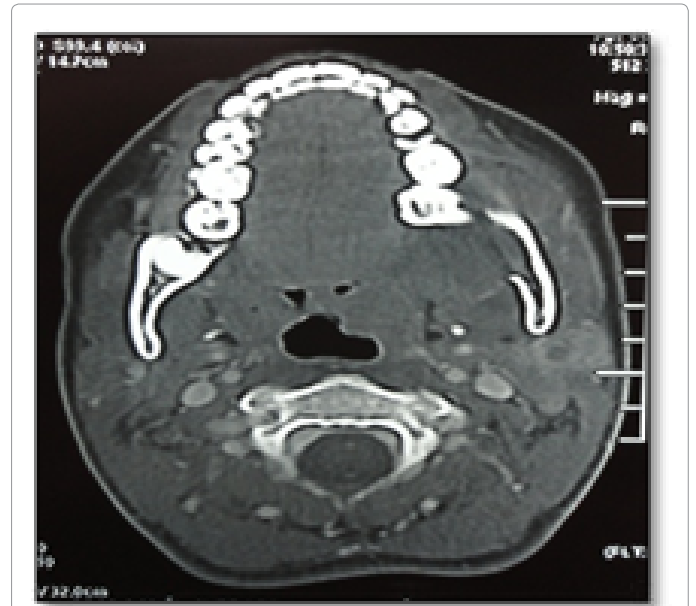
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Assessment of the intraoral cavity showed a 7 × 5 cm, exophytic ulceroproliferative growth, and red to brown, soft to firm, mild tender and diffuse mass. This lesion was covering the buccal and lingual side from the first molar to the retromolar area. Furthermore, the swelling of the left cheek was obliterating the buccal vestibule. The mass was resulting in impingement with occlusion (Figure 2). According to the patient statement, after 30 days of initiation of lesion, the third molar involved in the mass exfoliated. The second molar in the affected region were displaced. The patient could speak, but could not feed comfortably, and she has orthopnea.

The panoramic X-ray revealed a radiolucent lesion with ill-defined borders. The lesion was extending anteroposteriorly from the second molar to the third molar region, inferiorly up to the inferior alveolar canal of the mandible and superiorly to the alveolar ridge (Figure 3).

The Computed Tomography (CT) showed the infiltrative soft tissue mass. The lesion caused facial asymmetry, the marked expansion, and lingual cortical plate perforation. Displacement of the adjacent structures, and second molar was also noted (Figure 4).

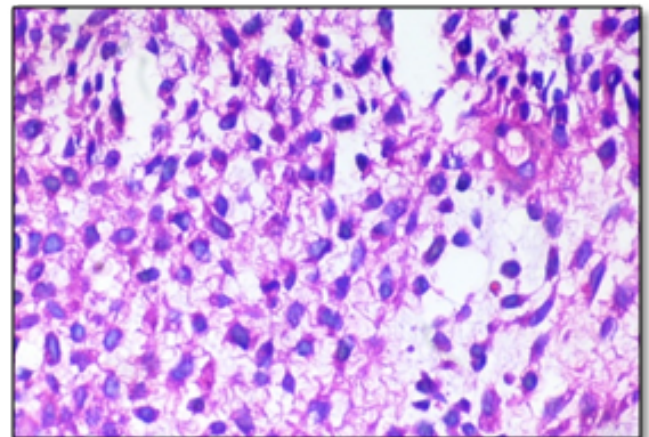
The lesion was subjected for histopathological examination via incisional biopsy under general anesthesia. Histopathological analysis of the hematoxylin/eosin-stained material showed highly cellular mesenchymal tissue containing round or spindle-shaped cells with darkly staining hyperchromatic cellular and nuclear pleomorphism, and cells with abundant slightly eosinophilic cytoplasm (Figure 5A).



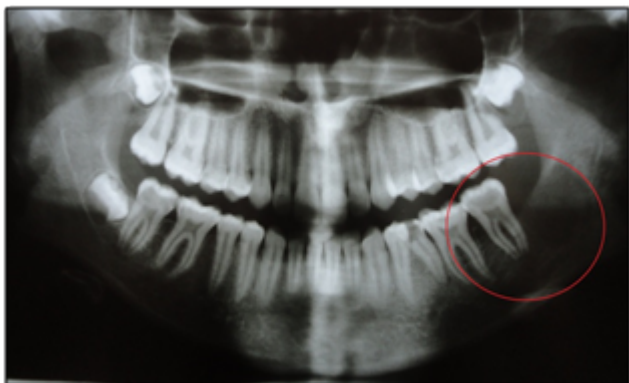
**Figure 4:** Computed Tomography (CT) shows facial asymmetry, lingual cortical Perforation, and teeth displacement.



**Figure 2:** Exophytic mass in left mandibular region.



**Figure 5A:** Round or spindle- shaped cells with hyperchromatic cellular, nuclear pleomorphism, with abundant slightly eosinophilic cytoplasm (H&E staining, original magnification X400).

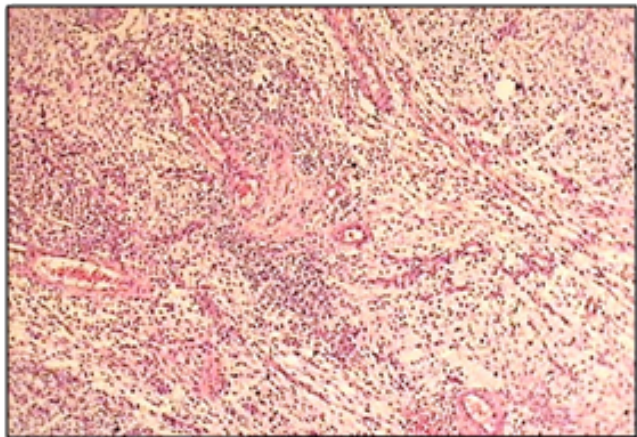


**Figure 3:** Panoramic X-ray shows a radiolucent lesion with ill- defined borders from the second molar to the third molar region after exfoliation of third molar.

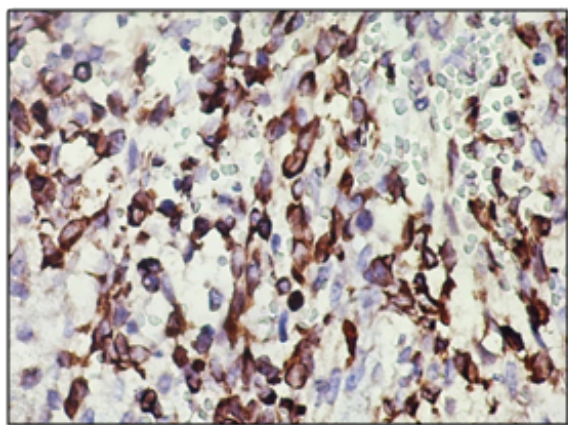
Mitotic figures, and some areas with loosely textured myxoid were noted (Figure 5B). The stroma also displayed severe bleeding, and moderate amount of mixed inflammatory cell infiltrate mainly containing, lymphocytes, plasma cells, and neutrophils. Histopathological features of this tumor indicated a differential diagnosis including Rhabdomyosarcoma, Ewing sarcoma and malignant lymphomas.

Considering the histopathological findings were not characteristic of a specific tumor, immunohistochemical staining was performed for vimentin, desmin, myogenin, MyoD1, Smooth muscle actin (SMA), S100, CD99, Leukocyte common antigen (LCA), and Ki-67. The neoplastic cells were strongly positive for vimentin, desmin, MyoD1, myogenin, and weakly positive for CD99 (Figure 6). Furthermore, the tumor cells were negative for S100, LCA, and SMA. On the basis of all

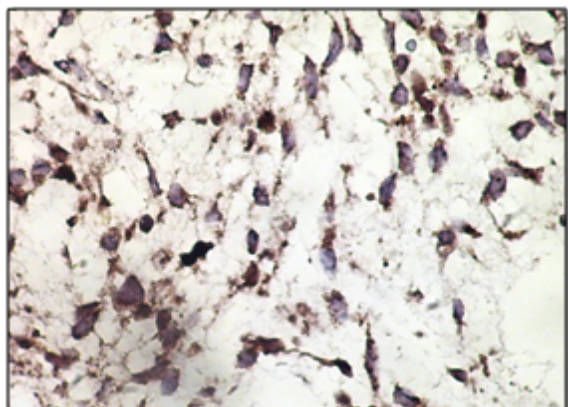




**Figure 5B:** Other aspect of the tumor showing loosely textured myxoid and blood vessels (H&E staining, original magnification X100).



**Figure 6A:** Tumor cell showing positive staining for Desmin (immunohistochemical staining, original magnification X400).



**Figure 6B:** Tumor cell showing positive staining for MyoD1 (immunohistochemical staining, magnification X400).

the immunohistochemical staining results, the tumor was identified as embryonal Rhabdomyosarcoma. The result of immunohistochemical staining for ki-67 showed proliferative activity in 70% of tumor nuclei.

She was referred to the oncology department, and the treatment plan was a combination of chemotherapy.

Although, the tumor showed dramatic regression after the initiated 6 weeks of chemotherapy, the computed tomography showed lungs metastasis. Unfortunately the patient denied further treatment because of high cost of treatment and at present her condition is undetermined.

## Discussion

Rhabdomyosarcoma is the third most common extracranial malignant tumor in children after neuroblastoma and wilms tumor [9]. The highest incidence of RMS is in the children aged 1-4 years, lower rate in children aged 10-14 aged and lowest rates between 15-19 years of age [10]. Approximately, 35% of all RMS occur in the head and neck regions. But, oral RMS is rarely encountered [7].

In oral RMS males are affected more commonly than females, and majority are occurred in first two decades of life [11]. In this report, a 15 year old female was presented. A common site of oral cavity has not been well recognized. Some studies have been showed that soft palate, tongue and buccal mucosa most common sites for oral RMS. Some other studies have been presented oral RMS in the posterior mandibular region [9,11]. Our patient involved the left mandibular posterior area.

In general, the patients with oral RMS often have an advanced lesion at the early stage, because of a rapidly enlarging painless mass, local infiltration, pain, ankyloglossia, paresthesia, and trismus. Primary oral RMS is rarely affected lymph nodes [11]. Some studies, showed involvement of the teeth in affected region [10]. In our patient, the painless mass with rapidly growing and involvement of the teeth, but no sign of pain or paresthesia was seen. She had facial asymmetry, and experienced difficulty in opening the mouth.

A careful histopathological analysis is necessary for differentiate the tumor from other small round cell tumors, which are also malignant or aggressive lesions. On the basis of some viewpoints, Rhabdomyosarcoma histologically has been divided in to four broad subtypes: botryoid and spindle cell RMS, alveolar RMS, embryonal RMS, and undifferentiated RMS [9]. The most common type of head and neck RMS are of embryonal type. In our patient, the presence of round cells with eosinophilic cytoplasm and immunohistochemical staining ruled out the diagnosis of malignant lymphomas and Ewing sarcoma. The use of immunohistochemistry for desmin, myogenin, and MyoD1, may only recommend the skeletal muscle differentiation, and is mainly beneficial for the diagnosis of RMS [12]. Positive result for vimentin showed that the tumor has a mesenchymal origin. Positive reaction for desmin indicated muscular differentiation. Although it is very sensitive it was not very specific for skeletal muscle. Desmin could not distinguish between skeletal and smooth muscle differentiation [13]. The MyoD family include four proteins myogenin, MyoD1, myf-5, and MRF-4. Antibodies to Myogenin and MyoD1 have high sensitivity and specificity for skeletal muscle differentiation [13]. They are positive in about 90% of the RMS and the diagnosis of RMS was established only if at least one of these proteins were positive [12]. In our case, desmin, myogenin, and MyoD1 were positive. Immunohistochemical staining for CD99 mainly used for the diagnosis of Ewing sarcoma, but it is also present in other tumors, such as lymphoblastic lymphomas, and some Rhabdomyosarcoma [13]. In our case, immunohistochemical staining showed weakly positive for CD99.

Metastases develop during the progression of the disease in about 20% of cases. Major metastatic sites are the lungs, lymph nodes, and

bone marrow. The lungs are involved in at least two thirds of patients' metastasis [5]. In our patient, it was found to have metastasized to the lungs at the time of initial presentation.

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