

Case Report

Rhabdomyosarcoma of the maxillary gingiva

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ABSTRACT

Rhabdomyosarcoma is a malignant skeletal muscle neoplasm. The tumor is much more common in children, and the most frequent site is head and neck region. Since this tumor is less frequent than other neoplasms in oral cavity, the clinicians sometimes ignore it, working the patients up. Rhabdomyosarcoma is a high-grade malignancy with poor prognosis. Considering the aggressive behavior and various clinical or histopathologic presentations of the tumor, early diagnosis has a significant impact on the treatment outcome and prognosis of the patients. We highlight the importance of combining the clinical, radiographic, and histopathologic examination to obtain a definitive diagnosis in sarcomas of the head and neck region, especially rhabdomyosarcoma. A case of rhabdomyosarcoma of the maxillary gingiva is presented in a 32-year-old woman in which the primary incisional biopsy was erroneously interpreted as an inflammatory process and consequently, the accurate diagnosis postponed for about 10 months.

Key Words: Head and neck, immunohistochemistry, oral cavity, rhabdomyosarcoma

Received: June 2017
Accepted: October 2017

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INTRODUCTION

Rhabdomyosarcoma is a malignant soft-tissue neoplasm with skeletal muscle origin, first described by Weber in 1845.^[1] This tumor is mostly common in infants and children and less common in adults and teenagers.^[2] In most of studies, there was a slight tendency to male gender.^[3] Head and neck region is the most common site of involvement, then genitourinary tract and the extremities.^[4] The most common sites in the head and neck region are orbit and parameningeal sinuses,^[2] and it is rare in oral cavity.^[5] The rare occurrence in oral and its similarities to benign tumors and inflammatory lesions could delay the diagnosis.^[6] In this article, a rhabdomyosarcoma of maxillary gingiva in a 32-year-old woman is presented.

CASE REPORT

A 32-year-old woman was evaluated for a painful swelling in the maxillary gingiva. She noticed a history of an inflammatory lesion at the same site and antibiotics using 10 months ago. In addition, the size of the lesion did not decrease. After extraction of the upper left second premolar, the lesion kept growing in size. Incisional biopsy suggested a nonspecific inflammatory lesion. She was referred to oral and maxillofacial surgeon.

In extraoral examinations, there was facial asymmetry. Left nasolabial fold was disappeared. Intraoral examinations revealed a firm, sessile, and pink

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How to cite this article: Motallebnejad M, Aminishakib P, Derakhshan S, Karimi A. Rhabdomyosarcoma of the maxillary gingiva. *Dent Res J* 2018;15:80-3.

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exophytic ulcerative mass with a lobulated surface, in maxillary left buccal gingiva, which extended from canine to distal of the first molar [Figure 1a]. On palpation, the lesion was tender and bleeds easily. The first premolar was mobile. In periapical radiography, the PDL widening was seen in teeth number 10, 11 and 12 [Figure 1b]. The incisional biopsy was performed under local anesthesia for histopathology examination.

The microscopic examination of the hematoxylin and eosin-stained sections revealed a neoplastic proliferation of spindle-shaped tumor cells in the adjacent connective tissue. Normal structures were destroyed by the invasion of tumor cells. The highly cellular neoplastic tissue was composed of predominant spindle cells in which some foci exhibited a fascicular pattern [Figure 2a]. In addition, marked cellular pleomorphism and nuclear hyperchromasia with prominent nucleoli, scattered atypical mitotic figures, and bizarre giant cells were identifiable among tumoral cells [Figure 2b]. Therefore, a primary diagnosis of “Spindle cell sarcoma” was rendered.

Using immunohistochemical (IHC) staining, the tumor cells were positively reacted with specific markers for myogenic differentiation such as anti-Desmin, anti-Myogenin, and anti-MyoD1 antibodies [Figure 3]. In addition, a highly positive staining (more than 10%) for Ki-67 was observed.

Finally, based on the histopathologic and IHC staining findings, a definitive diagnosis of the pleomorphic type of rhabdomyosarcoma was made.

The microscopic evaluation of the excised lesion confirmed the previous diagnosis. The excised specimen showed less spindle-shaped cells with more remarkable nuclear pleomorphism and bizarre tumor cells. In addition, scattered tumor cells with abundant eosinophilic cytoplasm and strap-like feature resembled “Rhabdomyoblast” [Figure 2c].

The patient underwent left hemi-maxillectomy. There was small swelling in superior area of the left sternocleidomastoid muscle 10 months after surgery. The initial diagnosis was lymph node metastasis of rhabdomyosarcoma and suggestive treatment was left cervical lymph node dissection after undergoing computer tomography of the head and neck. Neck dissection, removal of 14 supraomohyoid lymph nodes, complementary radiotherapy, and chemotherapy were done.

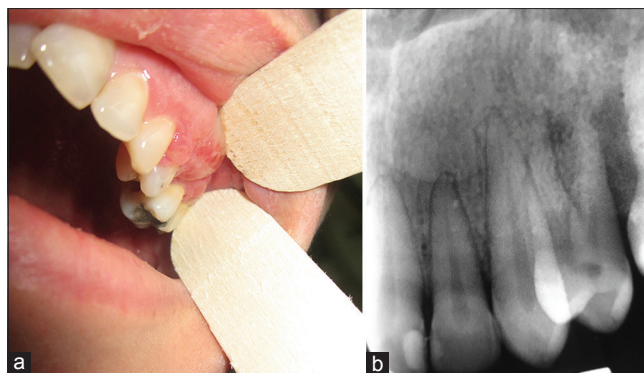


Figure 1: (a) Clinical feature of the lesion. (b) Periapical view of the lesion, note the widening of PDL in lateral incisor, canine, and first premolar.

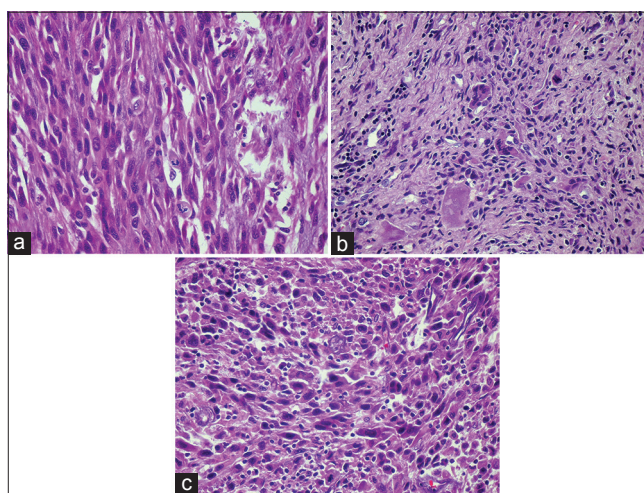


Figure 2: (a) Tumoral cells with abundant eosinophilic cytoplasm resembling “Rhabdomyoblasts” are seen. Note two atypical mitosis at central and mid-low of the picture (H and E, ×400). (b) Neoplastic cells with pleomorphism and scattered bizarre cells (H and E, ×400). (c) Malignant cells with abundant eosinophilic cytoplasm. Note straplike rhabdomyoblasts (×400).

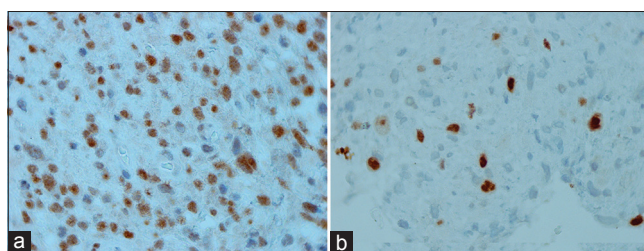


Figure 3: (a) Positive immunoreactivity of tumoral cells with anti-myod1 antibody (×400). (b) Positive immunoreactivity of tumoral cells with anti-myogenin antibody (×400).

DISCUSSION

Rhabdomyosarcoma is an aggressive neoplasm with a rapid growth in children. Oral cavity rhabdomyosarcoma is usually common in male and

in the first two decades of life.^[7] The mean age in diagnosis is 26.9 years.^[2] Although our patient was female and a little older than the mean age.

Thirty-five percentage of rhabdomyosarcomas occur in head and neck region,^[4,8] and oral cavity lesions are not common.^[4] In the present case, the tumor occurred in the buccal maxillary gingiva. Chi *et al.* reported a case in the same region in a 33-year-old woman.^[2]

Clinical intraoral manifestations of rhabdomyosarcoma are various,^[2] and as reported in this case, they could be misleading. In early stages, it usually appears as a painless swelling.^[6] In this case, the lesion was asymptomatic, and then, the patient reported pain as it grew.

Other signs and symptoms of oral rhabdomyosarcoma are tooth mobility, paresthesia, trismus and cervical lymphadenopathy. In this patient, although there was tooth mobility, the initial diagnosis was considered as inflammatory lesion. The tooth was extracted, and according to the diagnosis of inflammatory lesion, the patient received antibiotics. These clinical manifestations usually cause a delay in diagnosis.^[2]

The most common types of rhabdomyosarcoma are embryonal and then alveolar and pleomorphic.^[2,9] The prognosis of pleomorphic type is unfavorable. The embryonal type mostly has a favorable prognosis.^[9] In this case, the histologic variant was pleomorphic type that has relatively poor prognosis. Pleomorphic rhabdomyosarcoma is a rare subtype (about 5% of rhabdomyosarcoma cases) and occurs more commonly in the adult population.^[9] Although pleomorphic rhabdomyosarcoma in adults is rare and reported cases in very few and reported documented cases are in larynx and vocal cord in head and neck area.^[10]

According to histopathologic findings, it is hard to differentiate embryonal type of this tumor from small round cell tumor, neuroblastoma, and extrabony Ewing's sarcoma.^[11] Although any rhabdomyosarcoma may be pleomorphic, the pleomorphic type is limited to the variant that lacks areas showing an embryonal and alveolar pattern.^[12] The main entities to be considered in the differential diagnosis of pleomorphic type of rhabdomyosarcoma, particularly in the head and neck region, are spindle cell carcinoma, melanoma, leiomyosarcoma, malignant peripheral nerve sheath tumor (MPNST) with heterologous rhabdomyoblastic differentiation (known as malignant Triton tumor), and fibrosarcoma.^[9] It is necessary to look for the evidence of regular-appearing carcinoma

or melanoma at first. In addition, these neoplasms will demonstrate positivity for keratins or markers of melanocytic differentiation, such as S-100 protein, HMB-45, and Melan-A respectively.^[9] Cytogenic analysis and IHC lead to a more precise diagnosis. In leiomyosarcoma, very longer fascicles of spindle cells with cigar-shaped nuclei and brightly eosinophilic cytoplasm are evident that these are not a feature of rhabdomyosarcoma.^[9] In this case, in the second incisional biopsy, the markers of anti-Desmin, anti-Myogenin, anti-myod1 and Ki-67 were positive. These skeletal muscle markers are negative in leiomyosarcoma. Rhabdomyoblastic differentiation in MPNST is seen only focally within the main tumor and the dominant feature of tumor aid in the diagnosis. In addition, skeletal muscle markers will be only focally positive in MPNST, and the tumoral cells react with S100 in about 50% of cases.^[9] Finally, fibrosarcoma shows classic "herringbone" pattern and lacks rhabdomyoblasts. In addition, skeletal muscle markers are negative in this tumor.

The surgeon should pay attention to details such as the amount of tissue and its location during the initial biopsy made for biologic and histologic studies^[11]. Unfortunately, in this case, dentist's ignorance caused delaying in the correct diagnosis. This misdiagnosis of common benign swelling can lead to a delay in diagnosis and treatment.^[13] Initial management is critical for a better prognosis of any disease.^[13] Some studies have described that these tumors are more aggressive in adults compared to children and adolescents.^[14,15] Histology was a major treatment determinant and the most important prognostic factor in pediatric maxillofacial rhabdomyosarcoma.^[16] Rhabdomyosarcoma treatments include surgery, radiotherapy, and different regimens of chemotherapy.^[17] The treatments are based on the stage and clinical manifestations of tumor.^[17] Several prognostic indicators have been identified, which include age, tumor location, and histologic type.^[10] For example, in the head and neck region, the treatment of choice for orbital tumors is radiation alone or radiation and chemotherapy.^[15] Localized nonorbital, nonparameningeal head and neck tumors are most often amenable to surgery with low long-term surgical morbidity.^[17] These treatments improve the prognosis of rhabdomyosarcoma.^[2] Initial treatment of rhabdomyosarcoma involves complete gross surgical excision.^[18] The goal of surgical treatment is to remove the tumor without

causing a major functional or cosmetic defect.^[17] Radiation therapy and chemotherapy are used for local control of the primary lesion, regression of tumor size, and treatment of tumors that are not easily accessible to resection.^[17] This case report presented an interesting case of rhabdomyosarcoma because of the anatomical location, sex, and age of the patient, its histopathologic type. A 32-year-old woman with swelling of the maxillary gingiva resembling an inflammatory process reported. Resultant of this misdiagnosis led to suboptimal therapy. Delayed incisional biopsy showed spindle cell sarcoma, and immunohistochemistry confirmed the diagnosis of pleomorphic rhabdomyosarcoma the least common of the other categories of tumor. The patient received surgery, chemotherapy, and radiotherapy. However, because of cervical lymph node involvement, the prognosis was very poor.

CONCLUSION

In summary, rhabdomyosarcoma also occurs in adults, with apparently more aggressive behavior when compared with childhood rhabdomyosarcoma. Early accurate diagnosis and appropriate, usually multimodal, therapy is necessary for improve patient survival.

Acknowledgments

The authors appreciate the personnel of the Department of Oral Medicine and Oral Pathology of Tehran University of Medical Sciences.

Financial support and sponsorship

Nil.

Conflicts of interest

The authors of this manuscript declare that they have no conflicts of interest, real or perceived, financial or nonfinancial in this article.

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