

# Embryonal Rhabdomyosarcoma Occurring on Mandibular Gingiva in an Adult

GURURAJ PATIL<sup>1</sup>, SANGAMESH HALAWAR<sup>2</sup>, SHITALKUMAR SAGARI<sup>3</sup>, ROOPA BABANAVAR<sup>4</sup>, SHARAD PUROHIT<sup>5</sup>

## ABSTRACT

An embryonal rhabdomyosarcoma (ERMS) is a primitive, malignant, soft tissue sarcoma that recapitulates the phenotypic and biological features of embryonic skeletal muscles. Occurrence of intraoral ERMS in adults is extremely rare. This unique case report highlights the clinical, radiographic, histopathological and immunohistochemical findings of an intraoral ERMS.

**Key words:** Rhabdomyosarcoma, Embryonal rhabdomyosarcoma, Oral, Head and neck

## INTRODUCTION

Rhabdomyosarcoma (RMS) was first described by Weber in 1854. It was sub-classified by Horn and Enterline into embryonal, alveolar, botryoid, and pleomorphic sub-types [1,2]. ERMS is a primitive, malignant, soft tissue sarcoma that recapitulates the phenotypic and biological features of embryonic skeletal muscles [3]. 28% of RMSs occur in head and neck, and amongst them, 0.04% occur as intra-oral tumours [4,5]. Its occurrence in children is common, but in adults, it is extremely rare [2,5]. This case report emphasises the indifferent presentation of an intra-oral ERMS on mandibular gingiva, with clinical, radiographical, histopathological and immunohistochemical findings.

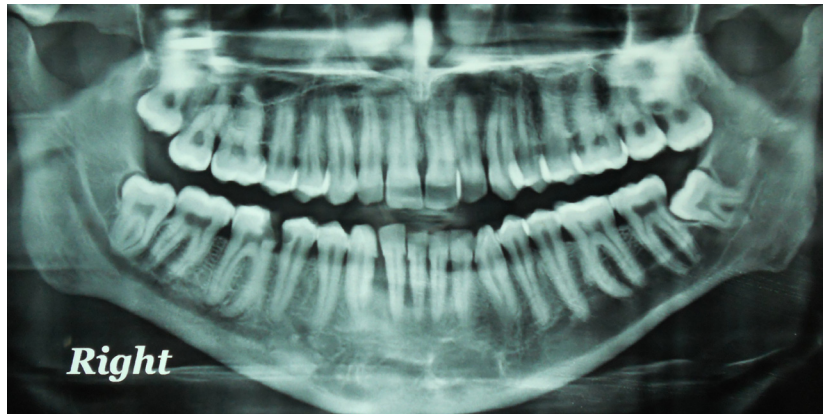
## CASE REPORT

A 34-year-old, male reported with a painless nodule in the lower right, front gingiva. Patient reported a progressive growth of the

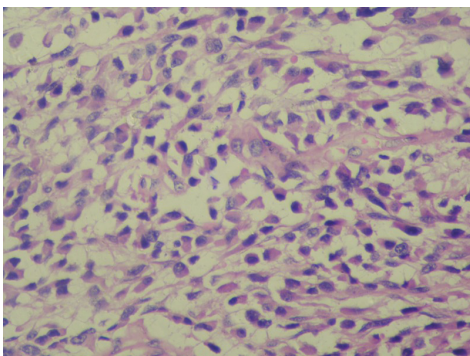
nodule since past six months. On intraoral examination, a swelling with a lobulated surface, which measured 3 cm × 4 cm, was located on mandibular gingiva. Swelling extended from right central incisor to right first premolar and it had a sessile base [Table/Fig-1]. Radiographic examination showed a superficial erosion of the labial cortical plate [Table/Fig-2]. A clinical differential diagnosis of a peripheral ossifying fibroma and a peripheral giant cell granuloma or a healing pyogenic granuloma was considered. After taking informed consent of the patient, an incisional biopsy was performed. Histopathological sections showed prominent primitive cells with amphophilic cytoplasm and central angulated or oval nuclei. Few stellate cells were seen as well [Table/Fig-3]. Presence of tadpole shaped, strap and spider cells was also evident [Table/Fig-4]. An immunohistochemical analysis was done. Vimentin [Table/Fig-5] was strongly positive, with limited expression for desmin and myoglobin. Patient was treated by hemi-mandibulectomy, followed



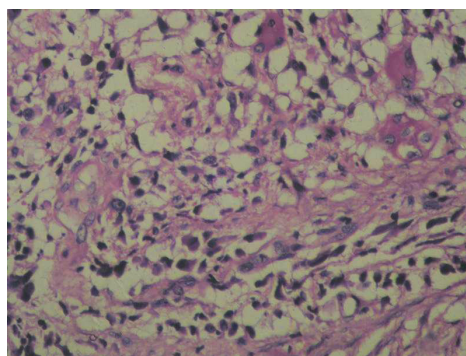
[Table/Fig-1]: Lobulated swelling on gingiva



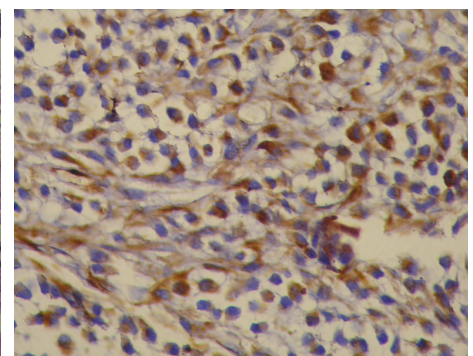
[Table/Fig-2]: Orthopantomograph showing superficial erosion labial cortical plate



[Table/Fig-3]: Primitive cells with light amphiphilic cytoplasm with oval and angulated nuclei



[Table/Fig-4]: Tadpole shaped, strap and spider cell



[Table/Fig-5]: Immunohistochemical expression for vimentine

by radiotherapy. The patient is under follow up since two years, with absence of recurrence.

## DISCUSSION

A recent PubMed search for intra-oral RMSs showed only 25 cases reports which have been published in English language literature. The most commonly affected areas are the head and neck region, genitourinary tract and retroperitoneum. The head and neck RMSs are anatomically divided into 2 categories: Para meningeal and nonparameningeal [1]. 60% of RMSs are of embryonal subtype [2,3,6]. Though they are most common in children, the average age of occurrence is 7.5 years, with a slightly male predilection [5]. Our patient was older than this average age. Previously reported cases had presented with signs of malignancy [5,6], but present case did not have any such signs, except for a superficial erosion of cortical plate. The histologic architecture of ERMS resembles embryonic muscle differentiation stages, which form aggregates of myoblast and, stellate cells with lightly amphophilic cytoplasm and central oval nuclei [1,2,6]. Thus, only vimentin is present in the cytoplasm of the most primitive cells, and desmin and actin are acquired by developing rhabdomyoblasts. Differentiated cells exhibit myoglobin; myosin, and creatine kinase M, markers which correspond to terminal differentiation [1,4,6]. The present case showed strong positive staining for vimentin and variable staining for desmin and myoglobin. Histological and immunohistochemical features were similar to others' findings.

## CONCLUSION

Dental practitioners frequently encounter cases of RMSs or ERMSS in children; however, the occurrence a painless swelling in adults is extremely rare and it can pose a diagnostic challenge. This kind of presentation of the lesion, with superficial erosion of cortical plate as only sign of malignancy as, can lead to a misdiagnosis of common benign swellings. This can lead to a delay in diagnosis and treatment. Initial management is critical for a better prognosis of any disease.

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### PARTICULARS OF CONTRIBUTORS:

1. Reader, Jodhpur Dental College General Hospital, Jodhpur National University, Jodhpur, Rajasthan, India.
2. Reader, Department of oral Pathology & Microbiology, VPDC dental college & hospital, Sangli, Maharashtra, India.
3. Senior Lecturer, Jodhpur Dental College General Hospital, Jodhpur National University, Jodhpur, Rajasthan, India.
4. Senior Lecturer, Jodhpur Dental College General Hospital, Jodhpur National University, Jodhpur, Rajasthan, India.
5. Senior Lecturer, Jodhpur Dental College General Hospital, Jodhpur National University, Jodhpur, Rajasthan, India.

### NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Roopa Babannavar,  
Senior Lecturer, Department of Conservative & Endodontics, Jodhpur Dental College General Hospital, Jodhpur National University,  
Jodhpur, Rajasthan-342001, India.  
Phone: 07597216316, E-mail: rbabannavar84@gmail.com

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