Case Report
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Embryonal Rhabdomyosarcoma as a Gingival Growth: A Rare Presentation

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Abstract

Rhabdomyosarcoma (RMS) is a malignant tumor of striated muscle composed of neoplastic mesenchymal cells with varying degrees of cell differentiation. It accounts 60% of all soft-tissue sarcomas in children and only 2-5% in adults. Typical pediatric rhabdomyosarcoma variants like embryonal and alveolar sub-types are less frequently occur in adults. It has predilection for viscera followed by the head and neck region. RMS occurs in the head and neck region with 35% and 10-12% in the oral cavity. Establishment of the correct diagnosis and histological sub-type is of very importance in the treatment of this disease, because of the rarity of this neoplasm in the adult population the treatment is not uniform in the literature. Here we present case of embryonal rhabdomyosarcoma as a gingival growth in a 26-year-old female with clinical, radiological, histo-pathological and immunohistochemical aspects.

Keywords: Embryonal, Rhabdomyosarcoma, Sarcomas

1. Introduction

Sarcomas are rare, malignant tumors that can arise from mesenchymal tissues at any site in the body[1]. Soft tissue sarcomas represent less than 1% of all adult solid tumor malignancies[2]. Rhabdomyosarcoma (RMS) is a malignant, soft tissue neoplasm that consists of cells derived from the primitive mesenchyme exhibiting a profound tendency to myogenesis[3]. The first published example of a malignancy of striated muscles i.e. rhabdomyosarcoma was a tongue lesion reported in 1854 and was initially described by Weber[1]. It is the most common soft tissue sarcoma affecting children, adolescents and young adults. The incidence of RMS is only 2-5% in adults and approximately 60% in children of all soft tissue sarcomas[3].

RMS is classified as parameningeal (orbital) and non-parameningeal (non-orbital) anatomically[4]. The sites of parameningeal include nose, nasopharynx, paranasal sinuses, middle ear, mastoid, infratemporal fossa and pterygopalatine fossa. The first case of an aggressive RMS in the middle ear was described by Soderberg. The non-parameningeal forms occur in scalp, larynx oropharynx, oral cavity and parotid gland. The most common oral sites are tongue, palate and cheeks. RMS that occurs in the head and neck region with 35% and 10-12% in the oral cavity[5].

The clinical presentation of RMS is variable and is influenced by the site, from which it origin, the patient's age, and distant metastases is there or not. The clinical presentation includes pain, loss of teeth, paresthesia and trismus depending on site of origin. It may appear as smooth or lobulated surface and in sometimes appear as botryoid or grape cluster–like lesion. At an early stage, it becomes fixed to surrounding tissues[1].

The role of radiography plays an important role to provide valuable information regarding to the size and extension of the lesion, the extension of level of bone

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destruction and critical organs involvement[5]. To confirm the diagnosis, biopsy of such type of mass is mandatory. Surgical treatment with or without radiotherapy, chemotherapy is the standard treatment multimodality for RMS. Adolescents and adults have worse survival rates as compared to younger children[2].

In view of its rarity, here we present case of embryonal rhabdomyosarcoma as a gingival growth in a 26-year-old female with clinical, radiographical, histopathological and immunohistochemical aspects.

2. Case Report

A 26-year-old female patient came to Oral Medicine and Radiology Department with a chief complaint of painless growth over the right mandibular premolar region since 2 months which was non–discharging, non-hemorrhagic. The growth was initially small in size and suddenly increased in size. Patient was giving history of termination of pregnancy 2 times in last year. Patient had associated complain of sudden weight loss since 2 months. Family history and personal history were not significant.

No any significant extra oral findings. Intra oral examination revealed a single ill-defined, approximately 1.5×2cm sized, an ulceroproliferative growth present over mandibular right premolar region extends from the mandibular right canine to distal of second premolar region. Bucco-lingually, the growth extended from the buccal vestibular sulcus to lingually into the linguoalveolar sulcus. Growth has reddish minute papillary like projections that covered with yellowish-white slough (Figure 1). Palpatory findings show that the growth was tender, sessile, as well as indurated and irregular border with everted and raised margin. Grade I mobility was present in the mandibular right first and second premolar. There was no any bleeding or any discharge. Right submandibular lymph nodes were non-tender, palpable, firm, mobile. Considering the age, sex and clinical finding, pyogenic granuloma, tuberculous gingival growth or malignant growths were suspected.

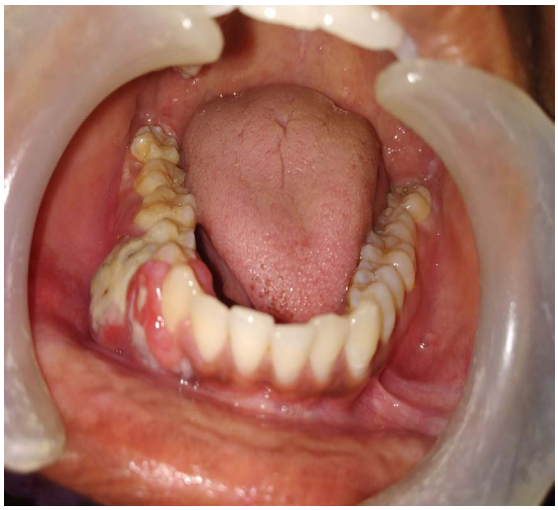


Figure 1. Clinical presentation shows a single ill-defined, approx 1.5×2 cm sized, an ulceroproliferative growth present over mandibular right premolar region with reddish minute papillary like projections that covered with yellowish-white slough.

Conventional radiographs (IOPA & OPG) revealed a single, ill-defined, irregular radiolucency extends from the mandibular right canine to second premolar and alveolar crest to 1cm extending into the alveolar bone (Figure 2). USG of neck revealed a well-defined hypoechoic collection noted in right submandibular region measuring about 10*20*10 mm in size with volume of about 10cc suggested of abscess Koch's in nature. FNAC from submandibular lymph node showed scant lymphoreticular tissue over necrotic background.

The Computed Tomography (CT) of the maxillofacial region showed presence of heterogeneously enhancing soft tissue density lesion noted involving mandibular right buccal space and gingivobuccal sulcus with minimally extends into right side of floor of mouth and abuts ventral aspect of anterior 2/3rd of right side of the tongue and abuts right side of mylohyoid muscle with loss of fat plane suggestive of the infiltrative mass. Presence of few nodes in right level IB, largest one showed internal necrosis (Figure 3).

To rule out distinct metastasis, chest radiography and ultrasonography of the abdomen were done which was insignificant. Incisional biopsy revealed poorly differentiated tumour. Tumor stage was T4N2bM0 (Stage 4). Under General anesthesia, wide local excision of right mandibular alveolus, extended mandibulectomy and right neck dissection II were performed. Chemotherapy with VAC (Vincristine + Actinomycin-D + Cyclophosphamide) for 3 weeks was given.

Histopathological examination of resected tissue was carried out, that shows the spindle cell differentiation with presence of necrosis. There is also presence of sub epithelial edema in squamous epithelium, and vascular proliferation and presence of infiltrative cells with prominent nucleoli and clear cytoplasm and few cells

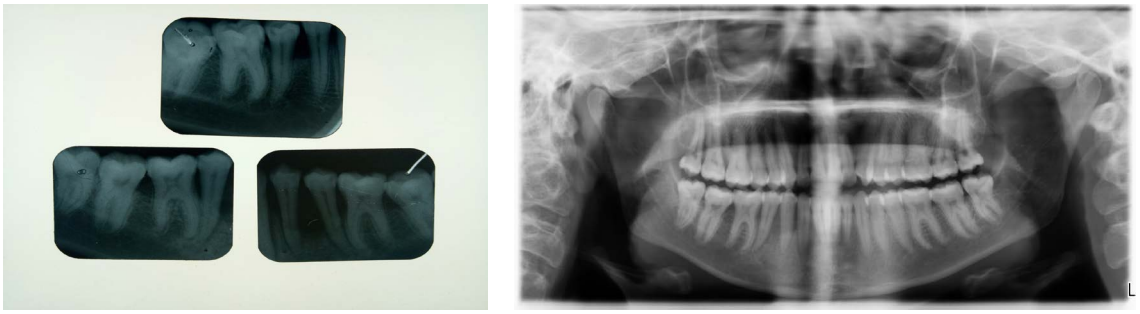


Figure 2. Conventional radiograph shows single, ill-defined, irregular radiolucency extends from the mandibular right canine to second premolar and alveolar crest to 1 cm extending into the alveolar bone.

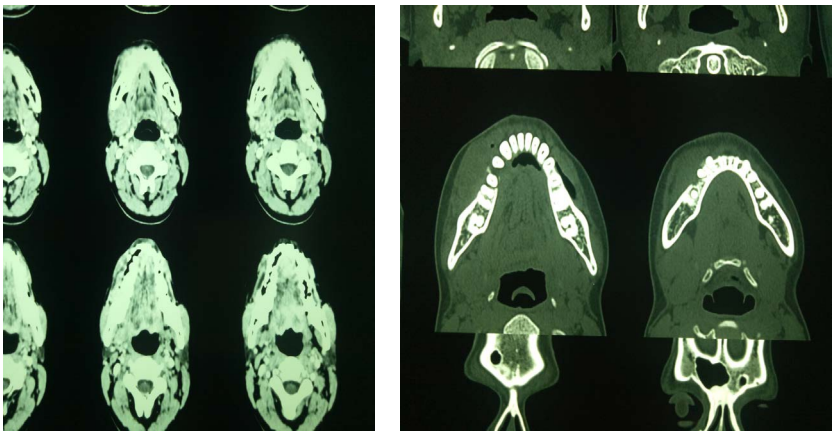
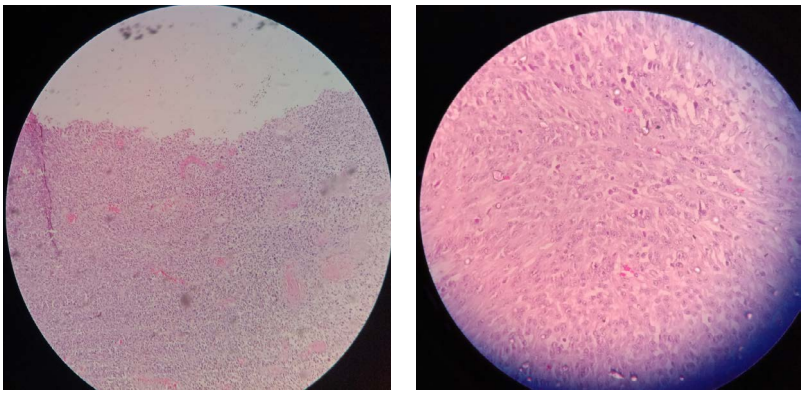


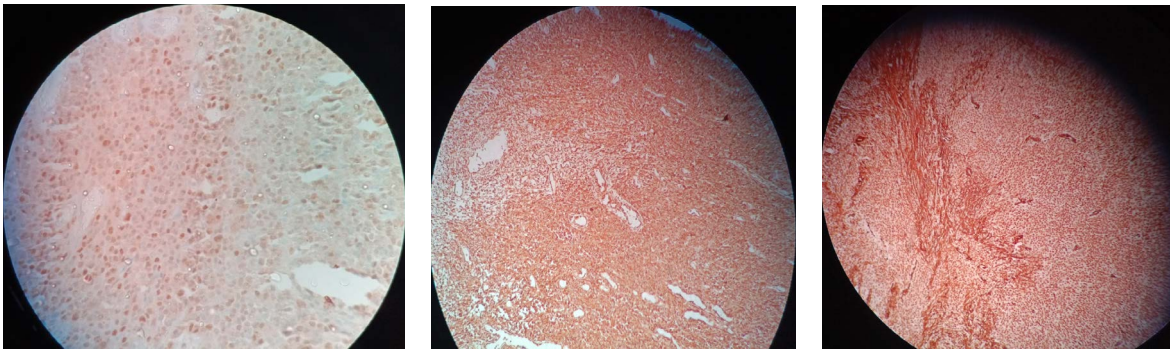
Figure 3. Computed Tomography shows presence of heterogeneously enhancing soft tissue density lesion noted involving mandibular right buccal space and gingivobuccal sulcus with minimally extends into right side of floor of mouth and abuts ventral aspect of anterior 2/3rd of right side of the tongue.

with eosinophilic cytoplasm in surrounding tissue. Mitoses were seen in many cells suggesting malignant small round cell tumor in embryonal rhabdomyosarcoma variety (Figure 4 a, b). Thus, diagnosis was made after histopathological examination of RMS, which was confirmed by doing immunohistochemisrty. Vimentin, desmin and calponin markers were positive (Figure 5 a–c).



(a) (b)

Figure 4. (a) Histopathologic section shows presence of infiltrative cells with prominent nucleoli and clear cytoplasm and few cells with eosinophilic cytoplasm (×20 H and E Section) (b) Atypical cells having nuclear pleomorphism (×40 H and E Section).



(a) (b) (c)

Figure 5. (a) Sheets of mesenchymal cells with cytoplasmic MYOD1 (40X). (b) Sheets of mesenchymal cells with cytoplasmic VIMENTIN stain(20X). (c) Sheets of mesenchymal cells with Nuclear ACTIN stain (20X).

3. Discussion

Rhabdomyosarcoma is a malignant tumor of the rhabdomyoblasts with similar a microscopic features of striated muscle cells[5]. It is an uncommon lesion but most common cancer of head and neck in young persons[1]. It has a slight predilection for males[1]. Soft palate, buccal mucosa, tongue are most common sites in oral cavity. In some studies, oral RMS presented in the mandibular posterior region[6]. According to Dito and Batsakis, incidence of occurrence of RMS in mandible is 3.5%[7], it is very rare to find RMS in adult female (26-year-old) in right mandibular premolar area as in our case.

RMS is usually present with infiltrative, fast-growing, painless mass. Clinical symptoms like pain, paresthesia, loosening of the teeth, and trismus occurs in the advanced stages of the disease, because of the infiltrative growth[8]. Primary oral RMS is rarely affect the lymph nodes[6,8]. In our case, there was presence of a rapidly growing, painless mass in mandibular premolar area associated with mobility of the involved teeth and enlarge, non-tender, palpable, firm, mobile right submandibular lymph nodes. There was no sign of pain, paresthesia, or trismus were seen.

Considering the age, sex and clinical finding differential diagnosis was given 1) Pyogenic granuloma as patient was young female and history of hormonal imbalance, 2) Tuberculous gingival growth as there was a history of sudden weight loss and USG of submandibular lymphnode suggestive of Koch's abscess and 3) Presence of such type of ulceroproliferative growth with ,indurated and irregular border, raised and everted edge and mobility teeth with irregular bone loss in a young patient may indicate malignancy.

Conventional radiograph, Computed Tomography (CT) scan or magnetic resonance imaging of the primary tumor should advise as an imaging study to determine the size, extension and possible involvement of associated structures. In case of tumors arising from the head, imaging studies of full views of the neck should be carried out to evaluate the involvement of the cervical lymph nodes[2]. In our case conventional radiographs revealed a single, ill-defined, irregular radiolucency. The CT scan revealed presence of heterogeneously enhancing soft tissue density lesion in mandibular right buccal space and gingivobuccal sulcus with minimally extends into surrounding tissue, suggesting the infiltrative mass. There was presence of few nodes in right level IB with internal necrosis.

Histologically, rhabdomyosarcoma has four broad subtypes which includes botryoid and spindle cell RMS, alveolar RMS, embryonal RMS, and undifferentiated RMS[1–6]. Embryonal rhabdomyosarcoma is a commonest variety occurring in the oral cavity with 70-80%[4]. Presented case has embryonal histological variant. The differential diagnosis of embryonal rhabdomyosarcoma by means of histopathological features includes other histological variants of rhabdomyosarcoma and other spindle cell tumors like Non-Hodgkin's lymphoma, Ewing's sarcoma, and neuroblastoma[4]. To differentiate RMS from other spindle cell tumors, electron microscopy and immunohistochemistry are very helpful investigation[7].

It is very beneficial in the diagnosis of RMS to use the immunohistochemistry of the desmin, myogenin, and MyoD1 markers. Positive reaction for vimentin indicates mesenchymal origin of tumor. Positive results for desmin indicated that tumor has muscular differentiation. Desmin

cannot differentiate the skeletal and smooth muscle cell differentiation. Four proteins such as myogenin, MyoD1, myf-5, and MRF-4 are included in MyoD family. MyoD1 and Myogenin proteins have high specificity and sensitivity for skeletal muscle differentiation. In about 90% of the RMS, these proteins have positive results[4–6]. Presented case has positive result for desmin, vimentin, Actin and MyoD1.

Multimodality treatment includes surgical therapy, chemotherapy, and radiotherapy. Surgical therapy is the most reliable treatment for sarcomas of the oral and maxillofacial region according to Yamaguchi et al. study found. Adequate surgical excision with keeping safe surgical margins is an important for better survival[4,7]. Combination with chemotherapy of VAC (Vincristine + Actinomycin-D + Cyclophosphamide) is the gold standard treatment for most of cases of rhabdomyosarcoma[2,4,5]. In our patient surgical excision and chemotherapy given with VAC (Vincristine + Actinomycin-D + Cyclophosphamide) for 3 weeks given.

As compared to other oral malignant tumors, rhabdomyosarcoma has relatively poor prognosis. Prognosis of tumor depends on anatomic site of the tumor, clinical staging, and histological subtype. Embryonal variety has a better prognosis with 83% survival rate[4].

4. Conclusion

Rhabdomyosarcoma is a neoplasm which has a highly aggressive nature with capability of rapid local invasion and infiltration into bone. Hence, early diagnosis and prompt treatment is the needed promptly. Any gingival growth in young female patient should be examine thoroughly and investigated histopathologically to differentiated pyogenic granuloma/pregnancy epulis from malignancy.

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