DOI: 10.7860/JCDR/2015/11263.5582

Case Report

Dentistry Section

Alveolar Rhabdomyosarcoma on the Left Maxillary Alveolus: A Unique Presentation

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ABSTRACT

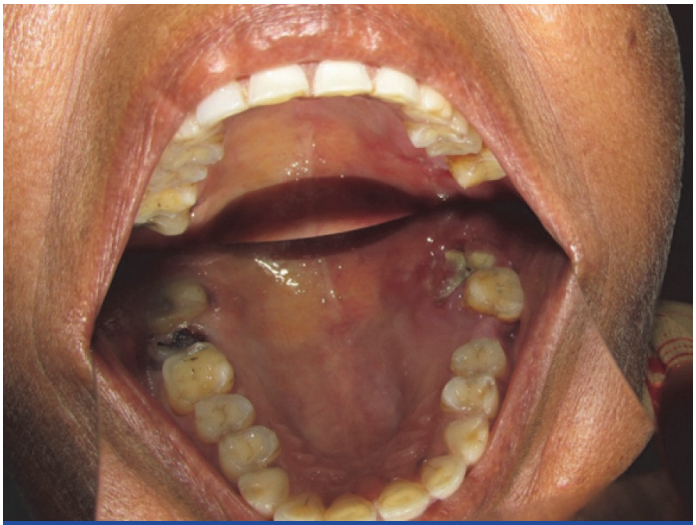
Rhabdomyosarcomas (RMSs) are a group of soft-tissue malignant tumours. They derive from primitive skeletal muscle tissue with head and neck as its principle location. These tumours are extremely rare in adults and it is believed to have a different natural course, treatment response, and prognosis. The invasiveness of tumour, metastasis, lymph node involvement, and the age at diagnosis is a predictor of outcome in patients with RMC. Hence early recognition and histological sub-typing is of critical importance in the therapy of the disease. We report a rare case of RMC in a 50-year-old female patient involving the left maxillary alveolus with a detailed clinical, radiological, histopathological and immunohistochemical findings.

Keywords: Immunohistochemistry, Muscle tumour, Orthopantomograph

CASE REPORT

A 50-year-old female patient presented with a complaint of painful ulcer in the upper left back region of the jaw since 10 days. The patient gave a history of extraction of a mobile tooth in the same region a month ago in a private dental clinic. Following which she had noticed the painful ulcer in the extracted socket. The patient claimed that the pain was severe in intensity, continuous, radiates to the left side of the face and did not subside even after taking a country medication (nature of which not known). The medical and family histories were non-contributory to the complaint.

Extra oral examination showed mild tenderness over the left paranasal sinus region, other facial structures appeared normal and no regional lymph nodes were palpable. Intra oral examination revealed diffuse ulceration of the extraction socket in relation to 28, measuring approximately 1.5x1cm. The edge/margins of the ulcer appeared slightly everted and the floor contained a yellowish slough. The base of the ulcer was not palpable as it was confined within the extracted socket. The surrounding mucosa appeared normal and no secondary changes were observed [Table/Fig-1]. Severe tenderness over the left faucial pillars and pharyngeal region was noticed. Considering the nature and extent of the lesion, a provisional diagnosis of non-healing ulcer of the extracted socket and a differential diagnosis of malignant ulcer was thought of.

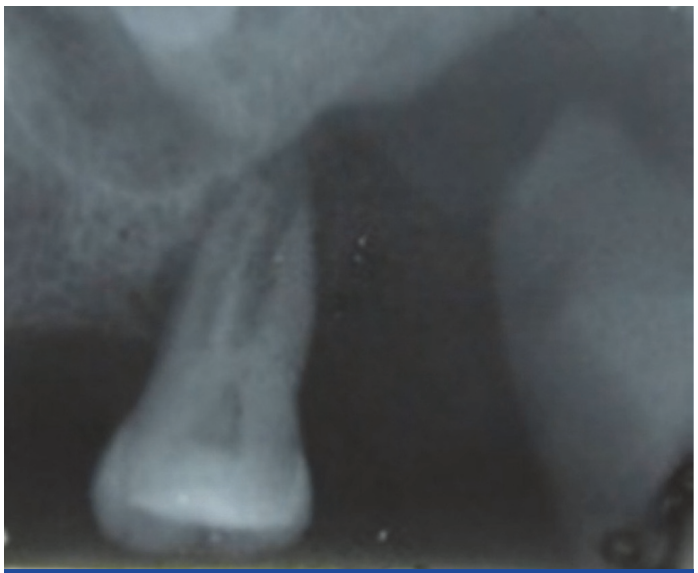


[Table/Fig-1]: Intra oral photograph (mirror image showing) diffuse ulceration of the extraction socket

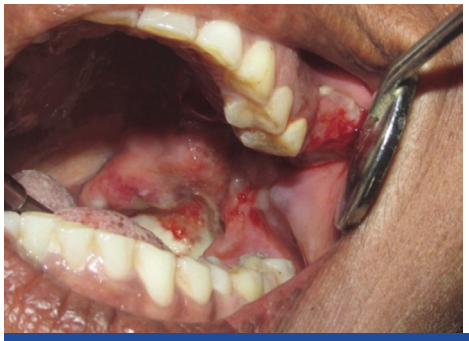
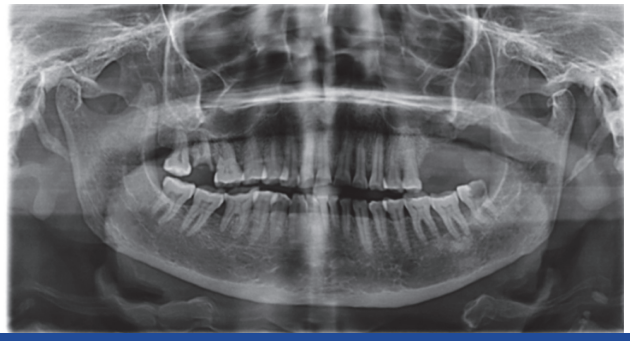
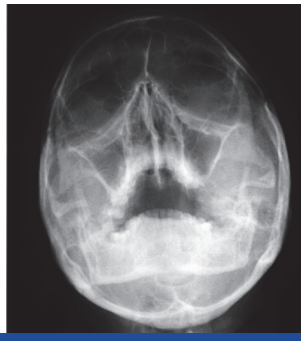
A screening intra-oral periapical radiograph taken in relation to 28 showed missing 28 and complete loss of alveolar bone distal to 27 with destruction of maxillary tuberosity was noticed [Table/Fig-2]. The patient was further advised for an orthopantomograph, Paranasal sinus view and routine blood investigations. The patient however, denied our advice and failed to turn up for the follow-up.

Two months later, patient reported to us with the lesion aggravating into a painful diffuse ulceroproliferative growth causing restricted mouth opening. On examination, no gross facial asymmetry was noticed and no regional lymph nodes were palpable. Intra oral examination revealed the growth extended mediolaterally from the midline of the palate to the buccal vestibule of the molars. Anteriorly from distal aspect of 25 to posterior aspect of the maxillary tuberosity region. Mucosa over the growth appears erythematous and yellowish slough was present. On palpation severe tenderness was noticed with indurated margins [Table/Fig-3].

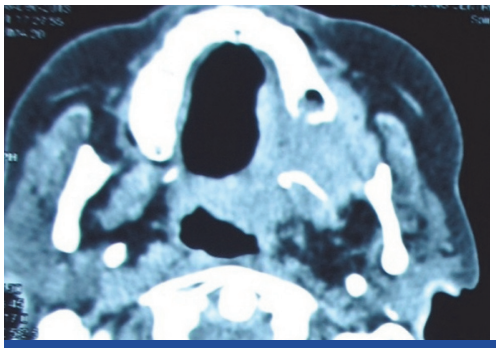
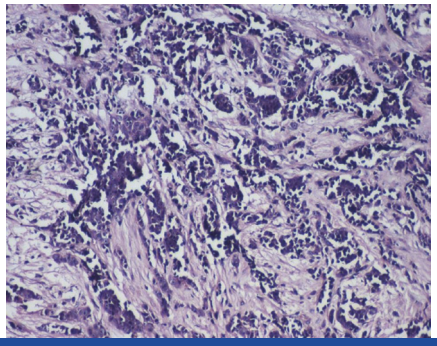
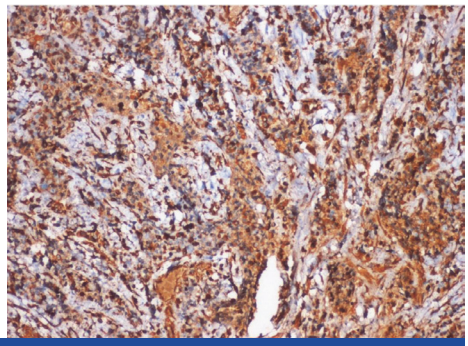
The orthopantomograph showed missing 26, 27 and 28 with complete destruction of bone distal to 25 [Table/Fig-4]. Paranasal sinus view revealed normal sinus architecture [Table/Fig-5]. Computed tomography revealed presence of soft tissue density lesion measuring 5.0 x 3.5 cm involving the alveolar process of left maxilla extending into the adjacent pharyngeal mucosal space



[Table/Fig-2]: Intra oral periapical radiograph showing severe bone loss

[Table/Fig-3]: Intra oral photograph diffuse ulcero-proliferative growth **[Table/Fig-4]:** Orthopantamograph showing severe bone loss distal to 25
[Table/Fig-5]: Paranasal view showing normal sinus architecture bilaterally

[Table/Fig-6]: Axial CT image showing soft tissue density mass involving the alveolar process of left maxilla **[Table/Fig-7]:** Photomicrograph (10x) showing cytoplasm of the tumour cells positive for PTAH **[Table/Fig-8]:** Positive immunohistochemical staining for vimentin (10x)

(lateral pharyngeal wall) and the soft palate [Table/Fig-6]. There was no obvious extension of the mass into the maxillary sinus but slight destruction of the posterolateral wall of the maxillary sinus was noticed.

Incisional biopsy of the lesion showed presence of round cells with large nucleus, prominent nucleoli with clear cytoplasm arranged in a pseudo alvelolar pattern admixed with variable number of large cells with eosinophilic cytoplasm and multinucleated giant cells. Cross striations were evident with few of the strap cells. The cytoplasm of the Tumor cell showed positive staining with PTAH and masson trichome stain [Table/Fig-7]. The tumor cell showed strong positivity for vimentin. S-100 and cytokeratin cocktail showed trace positivity suggestive of its Connective tissue origin. [Table/Fig-8] correlating with clinical, radiographic and H&E sections and special stains, a final diagnosis of alveolar rhabdomyosarcoma was arrived.

The patient was then referred to regional oncology centre for further treatment, unfortunately the patient was lost for follow-up.

DISCUSSION

RMS is a commonest mesenchymal malignant neoplasm seen in children. The peak age of incidence is between two to six years. Histopathology of RMS is analogous to myogenesis in the developing embryo, yielding clues to the biology of these lesions. Thus, RMS is considered as a tumour derived from primitive mesenchyme, exhibiting a profound tendency towards myogenesis than to define it as a cancer arising from skeletal muscle [1].

No clear etiologic factors have been identified to account for the occurrence of this malignant neoplasm. There are however increasing evidences which suggest that gene abnormalities may play a role in the development of RMS. Several non-random chromosome alterations and recurrent reciprocal chromosomal translocations have been identified in RMS. A t (2;13) translocation is detected in 70% of ARMS (alveolar rhabdomyosarcoma) ; a less common t (1;13) variant is found in 10% of ARMS [2].

RMS is very rare in adults. It accounts for 60% of tumour in children and 2–5% occurs in adults [3]. It is generally believed that, these tumours are more aggressive in adults when compared to children. Adult RMS shows different biological behaviour and an overall worse survival rate [4-6]. Clinical presentation of RMS is variable and influenced by site, age and the presence or absence of distant metastases. Intraorally, tongue is the most common site followed by the soft palate, hard palate, Buccal mucosa and gingiva [7-9]. Very rarely involvement of the masseter muscle has also been reported [10]. The presenting symptoms for rhabdomyosarcomas of the orofacial region include painful infiltrative growth of short duration, paresthesia, loss of teeth and trismus characterized by fast growth. Pain, proptosis, diplopia, strabismus, decreased hearing, nasal obstruction, dysphagia, cervical lymphadenopathy are other signs and symptoms. In the present case, the tumour initially was seen involving the extraction socket which rapidly infiltrated to the adjacent structures [11,12].

Radiological examination of the lesion may reveal the size and extension of the lesion and they usually show both the elements of bone remodelling and destruction. CT usually demonstrates a soft tissue mass that is isodense to muscle on unenhanced scans and frequently associate with bone erosion. On contrast enhanced CT, these tumours enhance moderately and homogenously. MR imaging include T1 signal intensity similar to muscle, T2 hyperintensity and heterogenous enhancement [13].

Horn and Enterline classified RMS histologically in four subtypes: embryonal, botryoid, alveolar, and pleomorphic. Histologically, the embryonal variety shows a mixture of spindle and un-differentiated round cells and immature striated muscle-like cells (rhabdomyoblasts) with abundant eosinophilic cytoplasm either tightly or loosely packed in a myxoid background. The Botryoid variant abuts an epithelial surface, with condensation of tumour cells in the immediate subepithelial zone. The alveolar variant has aggregates of round to oval neoplastic cells, separated by irregularly shaped fibrous trabeculae forming ill-defined alveolar spaces. The pleomorphic type contain large, atypical, polygonal, pleomorphic rhabdomyoblasts, which may be multinucleated.

Various immunohistochemical markers have been used to identify RMS. The markers include vimentin, myoglobin, desmin, muscle-specific actin, sarcomeric actin, smooth muscle actin, Myo D,

myogenin, troponin, S100 protein and cytokeratin [14]. In the present case, we used a panel of immunohistochemical markers and found positive expression for desmin, vimentin and a faint positivity for S-100 and cytokeratin cocktail.

RMS in adults is an aggressive tumour and has a higher incidence of recurrence. The best possible clinical outcome is therefore achieved via a multimodal approach [3]. Surgical resection followed by chemotherapy is the main stay of therapy. RMS is a radiocurable lesion but results in radiation-induced secondary tumours [15]. Chemotherapeutic agents including vincristine, adriamycin, cyclophosphamide, and actinomycin D reduces micro metastases and malignant cell population [16]. However, the cytotoxic actions of chemotherapeutic agents are not tumour-specific and are not effective in treating advanced and metastatic RMS.

The prognosis of RMS is determined by clinical staging, anatomical site, histology and age at the time of presentation [11]. On the basis of prognosis, RMS is classified into four groups [15]:

- Favorable prognosis: botryoid and spindle types
- Intermediate prognosis: embryonal
- Poor prognosis: alveolar and undifferentiated sarcomas
- Subtypes whose prognosis is not available: RMS with rhabdoid features.

CONCLUSION

Adult ARMS is a rare presentation. As stated earlier, the prognosis of this variant is relatively poor, hence a thorough examination and proper utilization of the imaging modalities, histopathology and immunohistochemistry should be considered to accomplish a correct diagnosis. With regard to treatment, a combined therapeutic approach involving surgery, chemotherapy, and radiation therapy are known to dramatically improve the survival rates. However, lack of cooperation from the patient/family can have a negative impact on the outcome, as seen in our case.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Sep 15, 2014**
Date of Peer Review: **Oct 09, 2014**
Date of Acceptance: **Jan 08, 2015**
Date of Publishing: **Feb 01, 2015**