

Rhabdomyosarcoma of Oral Cavity: A Case Report

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ABSTRACT

Introduction: Rhabdomyosarcoma (RMS) of oral cavity corresponds to 10 to 12% of all head and neck lesion and mainly involve the tongue, palate and oral mucosa. This malignant lesion is aggressive in behavior and consists of immature and highly invasive cells. The recurrence rate is high with generalized metastases through the hematogenic and/or lymphatic routes. The tumor is diagnosed by microscopic analysis and auxiliary techniques such as immunohistochemistry, electron microscopy, cytogenetic analysis, and molecular biology.

Case Report: We report here the case of a teenager with Oral RMS managed with wide local excision with adjuvant radio and chemotherapy.

Conclusion: RMS of oral cavity is a rare aggressive malignant tumour with high incidence of recurrence and distant metastasis. Management includes Surgical excision with adjuvant radiotherapy and/ or chemotherapy. Prognosis of RMS depends upon age of patient and extent of lesion.

Keywords: Rhabdomyosarcoma, Immunohistochemistry, Oral Cavity

INTRODUCTION

Rhabdomyosarcoma (RMS) is primarily an intracranial solid tumors which are found mostly in children¹⁻³. They are derived from primitive mesenchymal tissues that exhibit a tendency toward myogenic differentiation and originate from satellite cells produced during embryogenesis of skeletal muscle^{1,4}. The lesion was described first by Weber in 1854. The tumor has predilection towards white and African American children with males being more effected than females. This tumor generally occurs in the first two decades of life⁵. Rhabdomyosarcoma is idiopathic in origin, however possible viral involvement has been suggested. Chromosomal translocations and mutations in oncogenes have been noted but no definitive etiology is established^{2,6}. The head and neck are the most frequently affected regions, followed by the orbit (35% of cases)⁷, trunk and extremities, intra-abdominal organs and genitourinary tract (23%). Rhabdomyosarcoma of oral cavity corresponds to 10 to 12% of all head and neck lesion and mainly involve the tongue, palate and oral mucosa⁵. This malignant lesion is aggressive in behavior and consists of immature and highly invasive cells. The recurrence rate is high with generalized metastases through the hematogenic and/or lymphatic routes. The tumor is diagnosed by microscopic analysis and auxiliary techniques such as immunohistochemistry, electron microscopy, cytogenetic analysis, and molecular biology⁸. Advances in treatment modalities has increased the survival of patients with RMS and its prognosis is linked

with the location, evolutive stage and histological type of the tumor^{9,10}. We report here the case of a teenager with Oral RMS and associated review of the literature, focusing mainly on the clinical aspects, diagnosis and treatment of such tumors.

CASE REPORT

An 18-year-old male reported to our hospital with a fungating solid lesion obliterating two thirds of the oral cavity with associated dysphagia. There was dome shaped swelling in the middle and lower third of face on the left side (fig 1). The swelling had started around two months back as a peanut size growth along the left vestibulolingual sulcus. An incisional biopsy of the lesion was performed which revealed the lesion to be of mesenchymal origin pointing towards rhabdomyosarcoma. Individual was subjected to CT scan and MRI which revealed large space occupying lesion involving the left body and ramus of mandible (fig 2). It confirmed the presence of an extensive infiltrative lesion affecting the left buccal space, submandibular space and masticator space. The lesion was abutting the pterygoid muscles. There was bicortical erosion of mandible and displacement of adjacent structures. PET scan revealed local infiltration and ruled out distant metastasis. The lesion was managed with WLE using chin splitting incision with upper cervical transverse crease incision. The lesion was found to be fungating, necrotic, slough covered dumbbell shaped involving left mandibular body and ramus, gingivobuccal and gingivolingual sulcus extending into suprahyoid region of neck, invading the hyoglossus and mylohyoid. Subcutaneous fat and skin appeared free. Tumor mass was removed en-block including segmental resection of mandible (fig 3-4). Neck nodes were not significantly enlarged. Specimen was sent for histopathological examination. Histopathological analysis of the hematoxylin/eosin-stained material showed Herring bone pattern and proliferation of clear cells compactly arranged in solid masses and cords (Figure 5). Elongated cells which were fusiform in shape having cigar shaped nuclei were seen (Figure 6). Mitotic figures, some of them aberrant, were also noted. Immunohistochemistry

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How to cite this article: Serat Rahman, SK Roy Chowdhury. Rhabdomyosarcoma of oral cavity: a case report. International Journal of Contemporary Medical Research 2018;5(6):F7-F10.

DOI: <http://dx.doi.org/10.21276/ijcmr.2018.5.6.18>



Figure-1: Pre-op frontal view

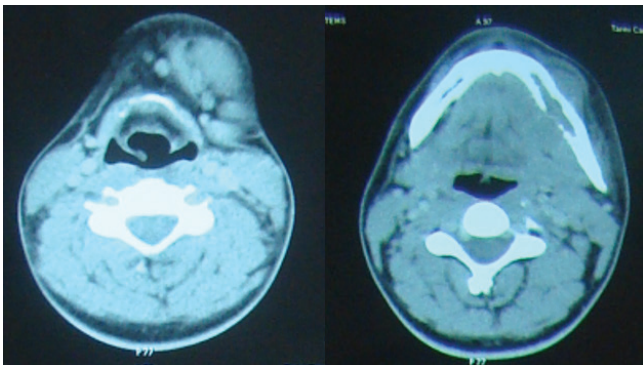


Figure-2: MRI showing extent of lesion

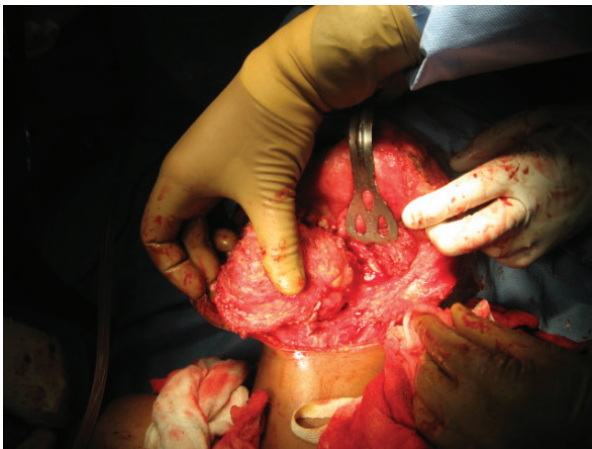


Figure-3: Intra -op view showing tumour mass



Figure-4: Intra-op view post segmental resection of mandible

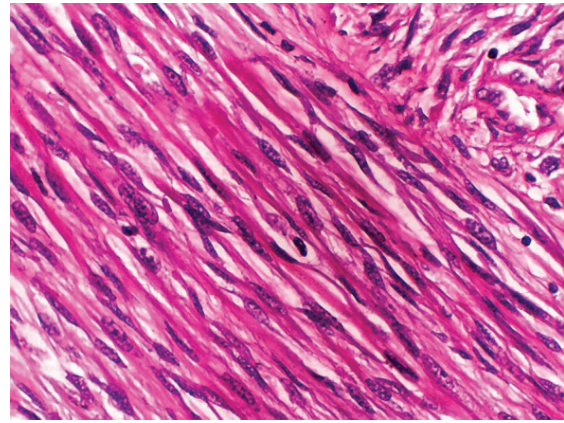


Figure-5: Herring bone pattern in histopathological examination

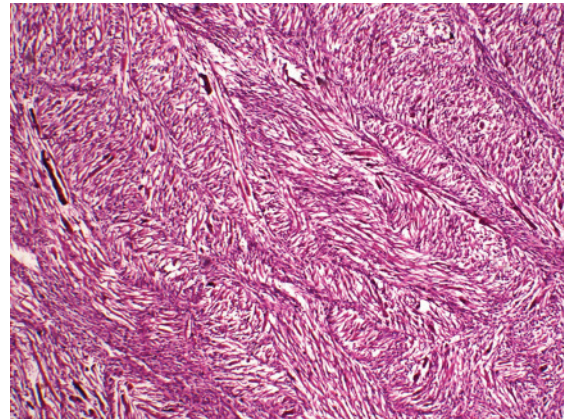


Figure-6: Elongated fusiform cells with cigar shaped nuclei



Figure-7: Post-op frontal view



Figure-8: ST-op intra-oral view

was performed using myogenic and non-myogenic markers (desmin, smooth muscle actin, myoglobin, vimentin and S100 protein). Staining was only positive for desmin, MSA and Vimentin. S-100, Cytokeratin, CD34, EMA, Bcl2 and Mic2 were negative. A diagnosis of Rhabdomyosarcoma (RMS) was established on the basis of the clinical, histopathological and Immunohistochemical characteristics. Post-operative period was uneventful (fig 7-9) and a guiding flange was delivered to patient. Radiation therapy and chemotherapy was given to the patient as adjuvant therapy.

DISCUSSION

Intra oral RMS are rare tumor. Around 30-40% of RMS occur in the head and neck region. These tumors are painless, have rapid growth, reaching large sizes in a short span of time⁷. Childhood RMS of the head and neck region have more favorable prognosis than those arising at other anatomical sites, possibly due to the early evolutive stage of the tumor at the time of its detection at these sites¹¹. The case presented here is in accordance with previous case reports in terms of patient age and the rapid and aggressive growth of the tumor which was also accompanied by bone destruction. According to the literature, clinical establishment of the differential diagnosis of RMS is difficult⁵, which can adversely affect the patient's prognosis. Common signs and symptoms associated with RMS include presence of a space occupying lesion with pain or paresthesia, loss of teeth and possible trismus due to factors like advanced tumor stage, infiltrative growth and site of tumor¹². The present case involved the ramus and body of the mandible, an uncommon location of RMS and therefore making this case unusual. The histopathological diagnosis of RMS is sometimes tricky since the tumor may exhibit nonspecific characteristics similar to those of other neoplasms, a fact requiring staining methods that are more specific than histopathological examination of hematoxylin/eosin stained specimens, particularly when the tumor is poorly differentiated. Diagnosis by immunohistochemical findings is not possible in all the cases and may be difficult to interpret within the clinical context². Findings of nonspecific spindle-shaped and round cells, sometimes exhibiting clear cytoplasm and pleomorphism, upon analysis of hematoxylin/eosin-stained material could lead to diagnosis of a malignant neoplasm of possible mesenchymal origin. However, diagnosis was established in our case on the basis of immunohistochemical analysis using antibodies for the identification of the cytological origin of the tumor. In addition, it is known that embryonal RMS exhibits marked cellular pleomorphism. Solid alveolar RMS has been reported by Franca et al⁷. Myogenin and desmin are sensitive and specific immunohistochemical markers for head and neck RMS, as compared to muscle-specific actin which is less sensitive^{1,13}. Myogenic nuclear regulatory proteins such as MyoD1 is another immunohistochemical marker that is helpful with the difficult diagnosis of RMS¹⁴. Expressed by fetal myoblasts, MyoD1 is a marker of the myoid lineage which is necessary for the transition from cell proliferation to differentiation. A number of differentiated cell types can

be transformed into skeletal muscle after transfection with MyoD1 through the activation of muscle-specific genes¹⁵. Myogenin is a myoid differentiation marker. In this respect, it has been described that origin of RMS may be due to the loss of normal proliferation and differentiation control¹⁶. Molecular biology techniques are utilized for the classification of RMS subtypes and may represent an important tool for planning of appropriate therapeutic management and estimation of prognosis⁸. Treatment of RMS consists of surgical resection, when possible, associated with chemotherapy and/ or radiotherapy based on the extent, location and stage of the tumor⁵.

CONCLUSION

RMS of oral cavity is a rare aggressive malignant tumour of skeletal muscle origin associated with high rate of recurrence and distant metastasis. The disease usually manifests within first two decades of life. Diagnosis of RMS is made by clinical examination and histopathological examination including immunohistochemistry. Management includes Surgical excision with adjuvant radiotherapy and chemotherapy. Prognosis depends upon age of patient and extent of lesion.

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Source of Support: Nil; **Conflict of Interest:** None

Submitted: 23-05-2018; **Accepted:** 25-06-2018; **Published:** 06-07-2018