CASE REPORT

Pleomorphic Rhabdomyosarcoma of Gingiva- A Rare Case Report.

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Abstract

Introduction: The most frequent soft tissue sarcoma seen in children and adolescents is rhabdomyosarcoma (RMS), a tumour of skeletal muscle origin. The head and neck region, genitourinary tract, retroperitonium, and, to a lesser extent, the extremities, are the most frequent locations of occurrence. The orbit, paranasal sinuses, soft tissues of the cheek, and the neck are the most often affected areas in the head and neck region. RMS is comparatively infrequent in the oral cavity, and jaw involvement is incredibly uncommon. Rhabdomyosarcoma types include embryonal rhabdomyosarcoma (approximately 60%), alveolar (approximately 20%), pleomorphic (approximately 10%), and spindle/sclerosing (approximately 10%).

Case Presentation: A 30-year-old female Patient complained of fleshy growth in the maxillary right alveolar region since past 15 days and we described the clinical, radiological, histopathological, and immunohistochemical findings for definitive diagnosis.

Management and Prognosis: Rhabdomyosarcoma has a poor prognosis when compared to other oral soft tissue malignant tumors, it is treated with surgical resection when possible, as well as multiagent chemotherapy and/or radiotherapy.

Conclusion: The distribution and diverse histopathologic pattern of RMS possess a challenge to the diagnosis, therefore judicious use of immunohistochemical markers may help in definitive diagnosis.

Keywords: Oral cavity, Pleomorphic rhabdomyosarcoma, Soft tissue sarcoma, Spindle cell lesion, Streaming lesions.

Introduction

Rhabdomyosarcoma (RMS) was first described by Weber in 1854, as a malignant soft tissue neoplasm of the skeletal muscle origin. The histogenesis of RMS is still unclear, but the most widely accepted hypothesis is that RMS arises due to the proliferation of embryonic mesenchymal tissue.[1,2] It accounts for almost 6% of all malignancies in children under 15 years of age. The most commonly affected areas are the head and neck region, genitourinary tract, retroperitonium, and, to a lesser extent, the extremities.[3] The head and neck RMSs are anatomically divided into 2 categories: parameningeal (including RMS of the nose, nasopharynx, paranasal sinuses, middle ear, mastoid, infratemporal fossa, and pterygopalatine fossa) and nonparameningeal (including RMS of the scalp, orbit, parotid gland, oral cavity, oropharynx, and larynx). RMS of the oral cavity accounts for 10–12% of all the head and neck malignancies.[1,3,4] On the basis of the histological findings, 4 broad subtypes of RMS have been identified: botryoid, spindle cell, embryonal, alveolar, and undifferentiated. In this report, we present a case of oral RMS arising within the left gingival mucosa and involving the maxilla in a 30-year-old female, and we describe the clinical, radiological, histopathological, and immunohistochemical features of this RMS.[5,3]

Case Presentation

A 30 year old female patient was referred to our department with a chief complaint of pain and swelling in her mouth since past 15 days. Clinical examination showed no facial asymmetry. Intraoral examination revealed an extensive mass which was soft, tender and seemed to be extending inferiorly, laterally as well as medially involving the alveolar mucosa and buccal mucosa along with that there is presence of vascular prominence.(Figure 1) Buccal sulcus was obliterated, with displacement and mobility of regional maxillary anterior teeth. A provisional diagnosis of osteosarcoma was made. Orthopantomogram showed extensive bone loss in the maxillary anterior region with a faint soft tissue shadow. Computed tomography images revealed ill-defined, heterogeneously enhancing, soft tissue

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mass causing extensive destruction of the maxillary dento-alveolar and palatal bone. (Figure 2 A and B) Differential diagnosis included, Non-Hodgkins lymphoma, fibrosarcoma, malignant peripheral nerve sheath tumor, chondrosarcoma and solitary plasmacytoma. Haematological parameters were within normal limits. Preoperative serology was negative for HIV and hepatitis. Incisional biopsy was performed from the most representative site after receiving informed consent of the patient, and was sent for histopathological evaluation. The section stained with hematoxylin and eosin revealed lesional tissue comprising of hypercellular areas with large, atypical ovoid to round to spindle shaped eosinophilic cells with hyperchromatic nuclei and indistinct cytoplasm.(Fig 3) Few areas comprised of elongated strap shaped cells resembling rhabdomyoblasts arranged in a haphazard pattern. Focal areas showed few multinucleated giant cells, muscle tissue along with areas of necrosis although distinct muscle striations were absent. With the above findings a diagnosis of malignant neoplasm of muscle origin was made. Immunohistochemical studies showed Desmin and Smooth muscle actin were immunopositive but S100 was found to be negative. (Fig 4A, B and C). We further found Myogenin and MyoD1 to be positive (Fig 5 A and B). Thus a definitive diagnosis of pleomorphic rhabdomyosarcoma was established.

The patient was referred to higher oncology institute and there according to the records obtained she was initially treated with three cycles of chemotherapeutic agent which included injection doxorubicin, and a combination of ifosfamide with mesna injection. She underwent wide excision of left maxilla with clear margins. The patient was further treated with local radiation therapy and she is still under regular follow-up there.

Discussion

After Neuroblastoma and Wilms tumor, RMS is the third most common extracranial malignant tumor that affects children. Of all soft tissue sarcomas, RMS accounts for only 2–5% tumors in adults but approximately 60% tumors in children.[5] The head and neck region is the most common site for RMS, with the orbit being the most frequent primary site. The most common site of involvement in the oral cavity is the tongue followed by the soft palate, hard palate, and buccal mucosa. Pleomorphic rhabdomyosarcoma is a rare variant of rhabdomyosarcoma that almost always arises in adults older

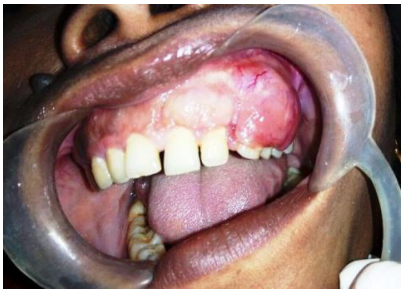

Fig. 1: A fleshy well circumscribed mass present on the left maxillary gingiva

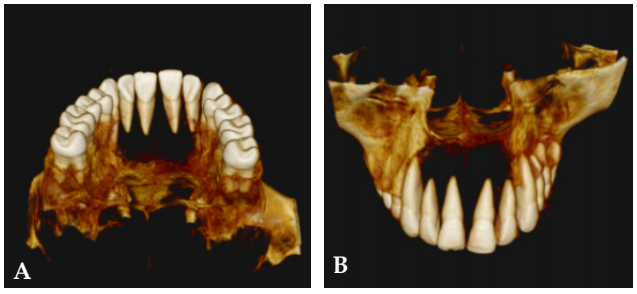

Fig. 2: CBCT - **A.** Palatal aspect and **B.** Labial aspect - showed severe boneloss in the anterior region having floating teeth.

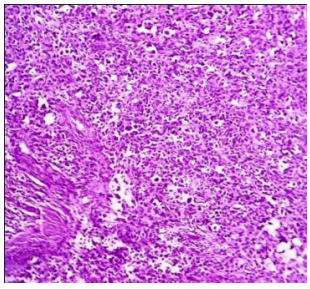

Fig. 3: Ovoid to spindle shaped hyperchromatic cells (H&E, 100X)

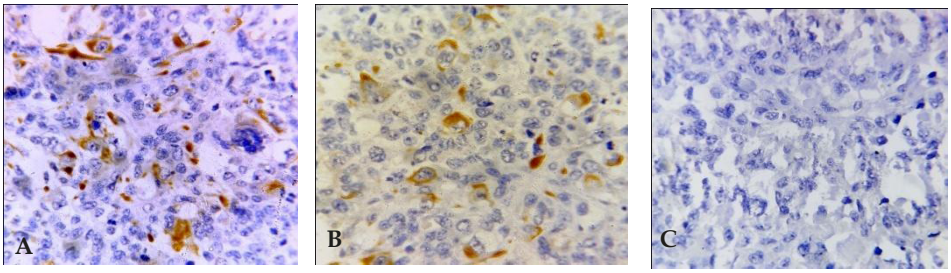

Fig. 4 A: Desmin positive, **B:** SMA positive, **C:** S-100 negative(IHC,100X)

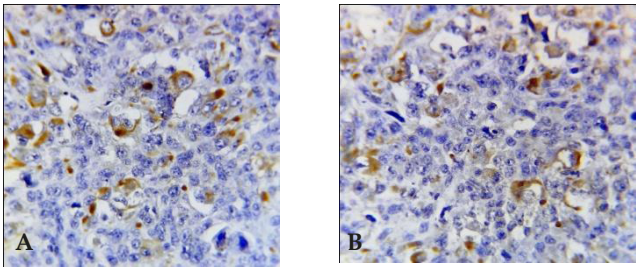

Fig. 5A: MyoD 1 and B) Myogenin positivity (IHC, 100X)

than 45 years.[6]

Studies in the 1960s described the clinicopathologic features of pleomorphic rhabdomyosarcoma, which accounted for 9% to 14% of all soft tissue sarcomas.[3,5] However, with the emergence of the concept of malignant fibrous histiocytoma, many pleomorphic rhabdomyosarcomas were subsequently reclassified as storiform-pleomorphic variants of malignant fibrous histiocytoma, and thus pleomorphic rhabdomyosarcoma became regarded as rare or nonexistent. Subsequently, with IHC and refined recognition of tumors with skeletal muscle differentiation, studies confirmed the existence of pleomorphic rhabdomyosarcoma and delineated criteria by which this sarcoma could be distinguished from other pleomorphic sarcomas.[7,8]

To establish the diagnosis of pleomorphic rhabdomyosarcoma, ancillary techniques are required. IHC identification of sarcomeric differentiation employing antibodies to desmin, muscle-specific actin, sarcomeric -actin, MyoD1, and myogenin is critical in not only detecting but also discriminating pleomorphic rhabdomyosarcoma from other adult pleomorphic soft tissue sarcomas-actin, MyoD1, and myogenin is critical in not only detecting but also distingushing pleomorphic rhabdomyosarcoma from other adult pleomorphic soft tissue sarcomas. However, Furlong et al.reported only modest sensitivity for MyoD1 (53%) and myogenin (56%) in pleomorphic rhabdomyosarcoma.[9,10,11]

Pleomorphic rhabdomyosarcoma may be arranged in a fascicular growth pattern reminiscent of pleomorphic leiomyosarcoma. However, the latter usually has lower-grade areas that display a well-defined fascicular pattern composed of cells with typical smooth muscle features. Both tumors are immunoreactive for actin and desmin, but MyoD1 and myogenin are only present in pleomorphic rhabdomyosarcomas. These markers are also useful in distinguishing pleomorphic rhabdomyosarcoma from all other types of pleomorphic sarcoma, including the undifferentiated form.[11,12,13]

Rhabdomyosarcoma from other more frequent and aggressive lesions affecting the concerned site like Fibrosarcoma, Leiomyosarcoma or Neurofibrosarcoma.[6,8] In our case, histological picture revealing predominantly spindle cell population, marked pleomorphism, fasicular pattern, etc was suggestive if not confirmative for differentiating rhabdomyosarcoma from Ewing's sarcoma and Neuroblastoma. Features favouring rhabdomyosarcoma over soft-tissue sarcomas such as fibrosarcoma, the presence of rhabdoid cells but definitive confirmation requires immunohistochemistry. According to literature, clinical diagnosis rhabdomyosarcoma is difficult; a fact that can markedly affect the patient's prognosis.[7,11,12] In the case reported the lesion was extensive and was involving maxillary bone.

Management and Prognosis

Prognosis of rhabdomyosarcoma is relatively poor compared to that of oral soft tissue malignant lesions. Pleomorphic Rhabdomyosarcoma treatment consists of surgical resection, when possible, associated with multiagent chemotherapy and/ or radiotherapy. It is said that 5 year overall survival rates were reported to be 63% for pediatric patients and 27% for adults.[13] However this case represents a 30 year old female patient who underwent treatment and is alive after 8 months follow up.

Conclusion

Any swelling should be carefully examined, and treatment outcomes should be regularly followed up. High degree of suspicion, early diagnosis, and a multidisciplinary treatment approach would be of great importance in such cases.[14,15] The ubiquitous distribution and diverse histologic pattern of RMS possess a challenge to the diagnosis; judicious use of immunohistochemical markers may help in diagnosis by ruling out other differential diagnosis. Recognition of the correct diagnosis and histological subtype of RMS is of critical importance in the treatment and prognosis of this disease.

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