Giant Sclerosing Oral Rhabdomyosarcoma-A Unique Case

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Received: 15 May 2015 / Accepted: 12 September 2015 / Published online: 22 September 2015
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Abstract We present a case of oral Rhabdomyosarcoma in a 50 years old man presenting with a huge mass in the oral cavity. Pre operative investigations showed the mass to be Rhabdomyosarcoma. The mass was excised and subjected to histopathological and immunohistochemical examinations which confirmed to be Rhabdomyosarcoma.

Keywords Rhabdomyosarcoma (RMS) · Immunohistochemistry

Introduction

Rhabdomyosarcoma, which was first described by Weber in 1854 [1]. It is the most common single variety accounting for approximately 4.5 % of all childhood malignancy with annual incidence of 5.3 per million children under the age of 15 years [2]. The most common sites of occurrence are the head and neck region (40 %), orbital tumour account for 10 %, parameningeal 20 %, other head and neck sites 10 %. It is less frequent in the oral cavity of all head and neck malignancies. The head and neck RMS are anatomically divided into two categories: parameningeal including (Nose, Nasopharynx, Paranasal sinuses, Middle ear, Mastoid, Infratemporal fossa and Pterigopalatine fossa).

And non- parameningeal including RMS of the Scalp, Orbit, Parotid gland, Oral cavity, Oropharynx and Larynx. RMS of the oral cavity account for 10–20 % all head and Neck cases [3].

Histologically three subtypes are recognized: Alveolar (20 %), Embryonal (60 %) and Pleomorphic (20 %). Alveolar and Embryonal is the most common soft tissue sarcoma of childhood and adolescence, usually appearing before age of 20. Pleomorphic rhabdomyosarcoma is seen predominantly in adults. The pediatric forms often arise in the sinuses, head and neck and genitourinary tract, location that normally contain much skeletal muscle [4].

The initial treatment of soft tissue sarcoma is resection. The principles of surgery for sarcomas were described by Simon and Enneking and remain the basis sarcoma management. Adult type sarcoma are considered to be radioresistant. Most clinically detectable tumors are unlikely to be eradicated by radiation doses within normal tissue tolerance, but there is a considerable body of evidence that shows it is capable of destroying microscopic disease.

Case Report

A 50 years old male presented with huge mass over the mouth. When the mass was first noticed by patient it was the size of a marble, and situated in the upper alveolus some 25 years back. In due course, it gradually increased in size, but in the 3 months the rate of increase was very rapid and attaining the present size. Within the last 2 months, he had difficulty in taking solid food and speaking, clinical examination revealed a smooth, pinkish-white, well defined solid to cystic mass approximately 10 × 10 × 8 cm in size extending from the superior

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alveolar ridge to the hard palate with loosening of teeth. The mass was nontender, it can be move side to side. The ear, nose and oropharynx were normal. The tongue was not involved but slight in increased in size noted (Fig. 1).

Fig. 1 Preoperative picture of the patient

Fig. 2 CT scan of neck

Computerized Tomography scan of neck and fine needle aspiration cytology (FNAC) were advised. CT scan neck (Fig. 2; plain and contrast) showing large solid cystic mass lesion with irregular septal enhancement, arising from the hard palate and superior alveolar process with epicenter in the right, causing erosion of the maxillary process with secondary pressure effects on teeth. FNAC not suggestive further advice for histopathological examination. All other hematological and biochemical profile were within the normal range. On the basis of finding the patient was planned for surgery.

An incision was given over the external surface of the mass, capsule separated out by dissection. The mass curetted out from the hard palate and superior alveolar ridge. The specimen was sent for histopathological examination that revealed fibrocollagenous hyalinised and myxoid tissue covered by a hyper plastic layer of squamous epithelial layer. The deeper part tissue section shows pleomorphic cells dispersed in variable numbers. The overall picture is suggestive Sclerosing Rhabdomyosarcoma. The morphologic features of the cells indicated a differential diagnosis comprising Rhabdomyosarcoma, pleomorphic Leiomyosarcoma and Malignant myofibroblastic tumors so immunohistochemistry examination was advised.

Immunohistochemistry shows soft tissue sarcoma with skeletal muscle differentiation, where Vimentin strong positive, Myogenin positive in nuclei of tumor cells 40 % Desmin positive weakly. The patient was discharged on 7th post operative day and referred to radiotherapy department.

Discussion

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 [5].

In 2000, Mentzel and Katenkamp reported a peculiar type of adult rhabdomyosarcoma characterized by prominent hyaline sclerosis and pseudovascular growth pattern. They called it sclerosing, pseudo vascular rhabdomyosarcoma [6].

In 2002, Folpe et al. described four additional cases with similar features, and they proposed to designate the lesion as sclerosing rhabdomyosarcoma. Because of the presence of heavily hyalinized collagenous matrix and occasional pseudovascular growth pattern. Sclerosing rhabdomyosarcoma.

Is frequently misdiagnosed as chondrosarcoma, osteosarcoma or angiosarcoma at initial evaluation. The relationship between sclerosing rhabdomyosarcoma and the conventional rhabdomyosarcomas remains uncertain. Although cytogenetic studies suggest a link with embryonal rhabdomyosarcoma [7].

In most cases, when initially examined, the patients already exhibit large tumors due to rapid growth and the frequent delaying medical consultation.

In our patient though the mass was present since 25 years back, but he came only when it rapidly increased.

There is no lymphadenopathy, except one palpable lymph node in the right level I b level, FNAC was done for this, but came to be reactive lymphadenitis.

The patient had lesions of the hard palate and superior alveolar ridge. There is no pain or paresthesia but loosening of teeth noted due to pressure effect of the mass.

The first line of treatment is radical excision and this is usually supplemented by radiotherapy. It is believed that adjunct combination with chemotherapy may greatly improve the prognosis. Inadequately treated tumours grow in an infiltrative manner and recur in a high percentage of cases.

Conclusion

Despite marked improvement in RMS survival over the past decades, it is known that, in addition to a poor prognosis for patients over 10 years old, delay in diagnosis have a impact on prognosis. The case reports presented were unique in respect to, the clinical appearance and location.

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