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Maxillary Embryonal Rhabdomyosarcoma with Oral and Nasal Extension in an Adult: A Case Report and Review of Literature

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Authors' contributions

This work was carried out in collaboration between all authors. Author RAV wrote the draft of this case report. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Embryonal rhabdomyosarcoma is a malignant soft tissue tumour that recapitulates the biological features of primitive striated muscle. This tumour is rarely encountered in adults. We present a report of maxillary embryonal rhabdomyosarcoma with oral and nasal extension in a 23-year-old man because of its rarity in both location and age.

Keywords: Embryonal rhabdomyosarcoma; rhabdomyosarcoma; maxillary tumour; oral tumour; nasal tumour; soft tissue tumour.

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1. INTRODUCTION

Rhabdomyosarcoma (RMS) is a malignant tumour of primitive mesenchymal origin which recapitulates the earlier forms of the skeletal muscle cell. The diagnostic hallmark of this tumour is, thus, the presence of rhabdomyoblasts [1]. It has been reported as the most common soft tissue tumour in children and adolescents and it is hardly encountered in adults [2]. This tumour usually occur in the extremities, but occasionally also found in the head and neck regions and viscera [3]. We present a report of maxillary embryonal rhabdomyosarcoma with oral and nasal extension in a 23-year-old man because of rarity in both location and age.

2. CASE REPORT

A 23 years old male presented to our institution with a complain of swelling on the left aspect of his face and difficulty in breathing for five months. During the last two months prior to presentation, there was a marked increase in size of the swelling with associated pain of progressive severity. The patient used traditional home remedies that involved making incisions with sharp objects and applying native herbal concoctions on the swelling. The rapid increase in size, uncontrolled haemorrhage from the wounds inflicted during home treatment and difficulty in breathing prompted the patient to seek expert treatment in the hospital. The progressive swelling was accompanied by a progressive nasal obstruction, serosanguinous nasal discharge and one episode of epistaxis. Proptosis of the left eye was noted and had been progressive. There was no limitation of ocular movements but there was frequent epiphora. Facial asymmetry was present on left maxilla. An oval, vaguely circumscribed, firm swelling was present in the left gingival sulcus behind the upper molar teeth. The swelling was non-tender and the maxillary teeth were non-mobile. There was an intra-nasal extension with impaction on the left nasal turbinates and proptosis of the left eye, nasal obstruction, epiphora, paraesthesia of the upper lip and incisus. The inferior left turbinate was engorged and patency test was negative. There was no attachment of the mass to the overlying skin. In addition, there was no cervical lymphadenopathy and the chest was clinically clear.

A plain radiograph of occipitomental view of the skull showed a radio-opacity of the maxillary antrum, no resorption of maxillary teeth and narrowed nasal passages.

The surgeon considered clinical working differential diagnoses of nasopharyngeal carcinoma and carcinoma of the maxillary antrum; to rule out fibro osseous lesion.

The sample specimen obtained, following an incisional biopsy, was fixed in ten percent formalin. Gross examination of the specimen in the laboratory showed a firm grey-white tissue that measured 2 x 3 x 5 cm and weighed 36.5 grams. Cut sections revealed a greyish solid surface with some dark brown foci of necrosis and haemorrhage. The tissue was processed and the 3-5 micrometers thick sections cut from the paraffin blocks were stained with routine hematoxylin and eosin.

Histology showed a loose myxoid stroma with rhabdomyoblasts at different stages of myogenesis (Fig. 1). There were numerous small primitive cells with scant cytoplasm and round nuclei. Larger cells that represent primitive forms of skeletal muscle (rhabdomyoblasts), recapitulating different stages of development, were also seen. Some of these had eccentrically displaced nuclei resembling plasma cells, while others appeared like tadpole cells with tapering cytoplasm, and strap cells. Giant sized cells with bizarre nuclei were also present (Figs. 2 and 3). The tumour was histologically classified as an embryonal rhabdomyosarcoma. Unfortunately, the patient died while still being prepared for a definitive surgery.

3. DISCUSSION

Rhabdomyosarcoma is the most common tumour of soft tissue histogenesis in childhood and adolescents, making up about 60 percent of reported cases with occurance of 4.6/ million children in the United States [3]. Rhabdomyosarcoma is very rare in adults [4]. This tumour is histologically classified into the embryonal, alveolar and pleomorphic sub-types [5]. A series of 277 rhabdomyosarcomas studied by Ahmad et al. [6] the embryonal variant was the most dominant sub-type accounting for 87.4% of the cases. Embryonal rhabdomyosarcoma (ERMS) is a primitive malignant tumour of soft tissue, which recapitulates the biological characteristics of embryonic striated muscle [3]. A great preponderance of 242 cases of the embryonal

sub-type evaluated by Ahmad et al. occurred between the ages of 0-10 years (159, 65.7%) while very few (14, 5.8%) occurred within the age bracket of 21- 30 years [6]. The patient's age (23 years) of the index case reported in this paper falls within this age bracket.

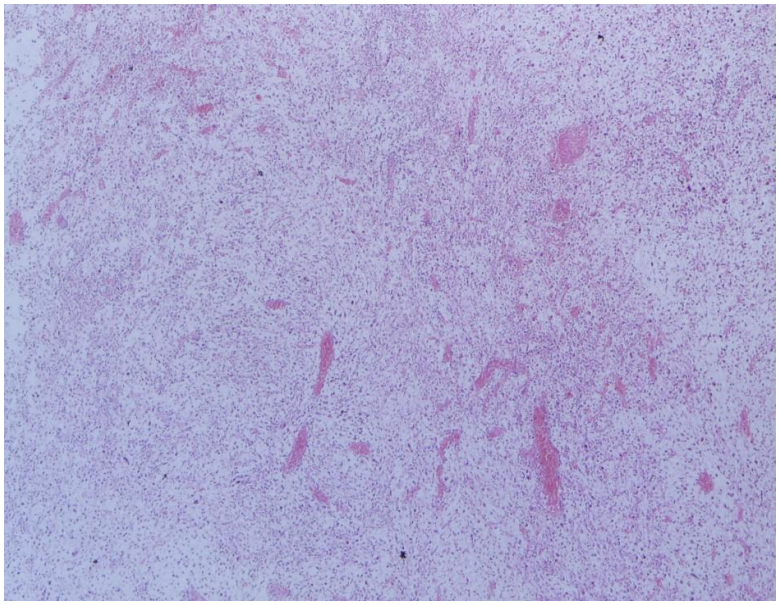

Fig. 1. Embryonal rhabdomyosacoma

This scanning magnification image shows numerous small blue cells scattered in a loose gelatinous matrix resembling primitive mesenchyme. [H & E; x4 objective magnification]

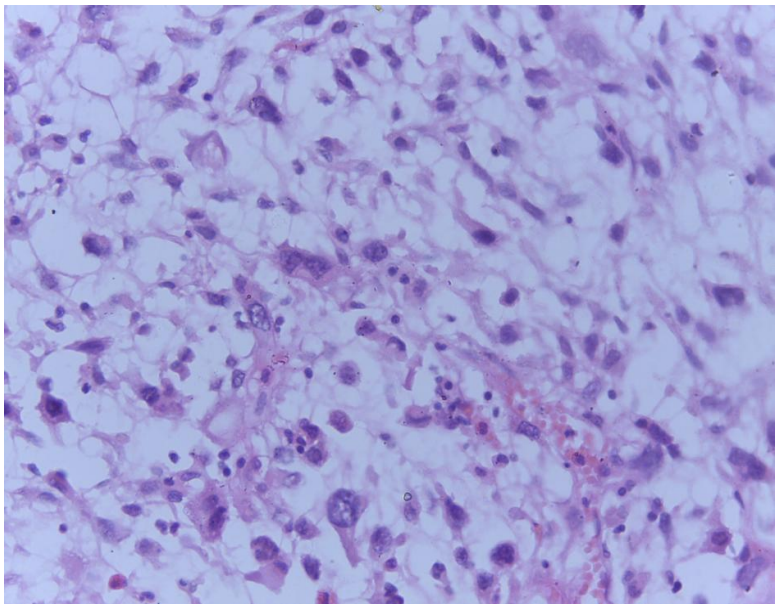

Fig. 2. Embryonal rhabdomyosacoma

This image demonstrates a pale myxoid stroma, a mixture of small primitive round blue cells and giant cells with eccentricaly placed nuclei. The spectrum of the morphological appearances of the rhabdomyoblast, described with different terms such as tadpole, tennis racquet, spider and strap cells are shown. [H&E; x40 Objective magnification]

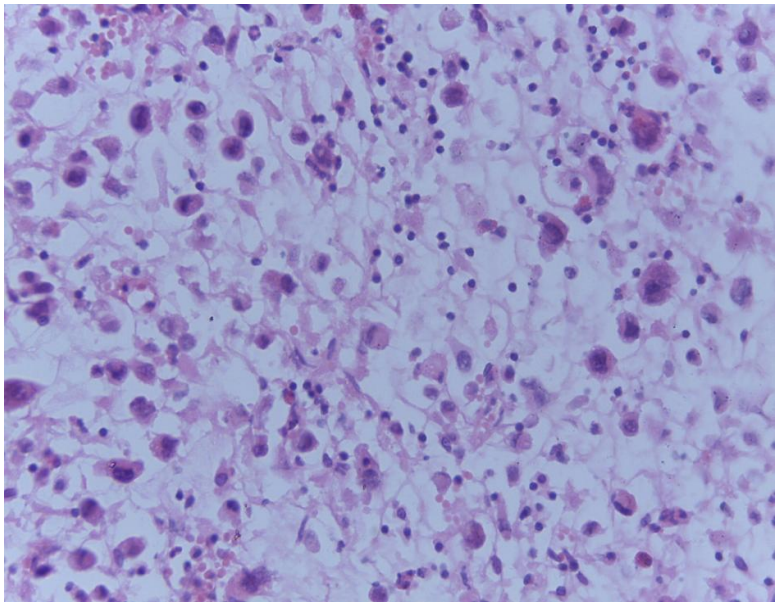

Fig. 3. Embryonal rhabdomyosacoma

Rhabdomyoblasts at different stages of differentiation are present, ranging from the primitive lymphocyte-like cells to the larger cells with eccentrically displaced nuclei. Some of the cells have bizarre nuclear shapes. [H&E; x40 objective magnification]

ERMS occur commonly in the head and neck, and genitourinary regions. Rhabdomyosarcomas in the head and neck region are generally grouped by site of occurrence into those found in the orbit, and parameningeal and non-parameningeal sites. Parameningeal sites include the external auditory meatus, middle ear, nasopharynx and other sites where erosion of bone is a feature. Non-parameningeal sites include the cheeks, oral cavity and oropharynx. The case we reported has the perculiarity of involving two sites, viz: the oral cavity and the nasal cavity. Most reported cases of rhabdomyosarcoma in the oral cavity have been of the embryonal type [6]. The variants of ERMS include the botryoid, myxoid, spindle cell and the round cell types. Just as observed in the case reported, the symptoms of ERMS in the oral cavity are related to mass effects and obstruction. The tumour, visible in the oral cavity, extended as far as the nasal cavity, obstructed respiration and caused proptosis of the left eye.

ERMS grossly appears as a soft yellow brown or grey mass with areas of necrosis and haemorrhage, and exhibit insidious invasion of adjacent structures. The principal identifying feature on microscopy is evidence of myogenesis exemplified by the presence of the rhabdomyoblast at diverse phases of differentiation [3]. However, it is a misconception to assume that these tumours arise from striated muscles because many cases in children have been documented in organs such as prostate, gall bladder and urinary bladder, where skeletal muscles are absent [7]. The less differentiated ones exhibit primitive round blue cells while the better differentiated display brightly eosinophilic cytoplasm, sometimes touting cross striations [1]. As they differentiate, the tumour cells undergo fusion, giant cell transformation, and tandem nuclear displacement, similar to normally developing myoblasts. They characteristically produce alternating foci of hypocellularity resembling primitive mesenchyme with gelatinous loose matrix and areas of areas of condensation resulting in hypercellularity [7].

The histological differential diagnosis of embryonal rhabdomyosarcoma includes desmoplastic small round cell tumour, neuroblastoma, large cell lymphoma, Ewing-sarcoma/primitive neuroectodermal tumors (PNET) and undifferentiated sarcoma. The non-malignant counterpart, rhabdomyoma, may also be considered as a differential diagnosis during histological evaluation. These tumours may be identified by using a combination of morphological pattern and immunohistochemical reactivity to desmin, myoglobin, myosin, vimentin, muscle specific actin (HHF 35), sarcomeric actin, smooth muscle actin, and

troponin T[5]. A positive reaction for vimentin confirms a mesenchymal derived tumor and desmin indicates muscular differentiation. Desmin expressed in myotome of embryo is identified in 75-100% of Rhabdomyosarcomas [8,9]. The cells in Ewing sarcoma characteristically form rosettes and they have pale uniform nuclei which are usually not hyperchromatic. In addition, immunohisto-chemistry show Ewing sarcoma to be CD99+, desmin -, muscle specific actin -, and t(11, 22)+. Neuroblastomas similarly, has small round blue cells which also form rosettes but their cytoplasms tend to have granular chromatin and the patient has elevated levels of urinary catecholamines; and imunohistochemistry show positive reactivity for S100, GFAP and chromogranin. Undifferentiated sarcomas are generally negative for muscle markers. Large cell lymphoma is usually positive for CD45, but just like the undifferentiated sarcoma, negative for muscle markers [10].

Cytogenetic alterations in embryonal rhabdomyosarcoma include hyperploidy with the presence of extrachromosomes 2, 8, 9, 11, 12, 13, 17, 18 and 20. There is also loss of chromosomes 10, 14, 15 and 16. Loss of heterozygosity (LOH) at 11p15.5 which affects the expression of insulin-like growth factor (IGF) has also been described [11,12].

The treatment protocol recommended by the Intergroup Rhabdomyosarcoma Study IV group includes surgical excision, radiotherapy and combination chemotherapy with vincristine, actinomycin-D and cyclophosphamide [9,13]. Unfortunately, the patient passed on within two weeks after the release of the histopathology report; and the preparation for a definitive surgery was thus, cancelled. The age, site, stage and histological classification are the prognostic factors of rhabdomyosarcoma [3]. It is established that embryonal rhabdosarcoma has a worse prognosis in adults than children [14]. The histological subtype and does not seem to influence prognosis in adults. There is involvement of the parameningeal region in the case presented. The literature suggests that the parameningeal tumours tend to have a rather poor prognosis [3].

Not many cases of maxillary, oral or nasal cavity embryonal rhabdomysarcoma in adults have been published. This present report, therefore, adds to the pool of literature on this subject.

4. CONCLUSION

Rhabdomyosarcoma is a malignant tumour of primitive mesenchymal origin exhibiting myogenesis histologically. We report a rare case of intra-oral occurrence of embryonal rhabdomyosarcoma that occurred in a 23 years old man. When encountered in daily practice, it should be adequately investigated and correctly identified.

CONSENT

As per international standard or university standard, the patient's written consent has been collected and preserved by the authors.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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