TURKISH JOURNAL of ONCOLOGY

Turk J Oncol 2023;38(2):254–56
doi: 10.5505/tjo.2023.3893

LETTER TO THE EDITOR

A Rare Case of Adult Embryonel Rabdomyosarcoma Including the Mandible

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Dear Editor,

Rhabdomyosarcoma (RMS) is a mesenchymal malignant neoplasm that exhibits skeletal muscle cells with varying degrees of differentiation, first described by Weber in 1854.[1] It accounts for <3% of adult soft-tissue sarcoma, but is the most common soft-tissue sarcoma histological subtype and the 4th most common childhood cancer before the age of 10.[2] Oral RMSs are classified as non-orbital and non-parameningeal and represent 28% of head and neck RMSs.[3] Dört alt tipi vardır: pleomorfik, alveolar, embriyonal ve botryoid. It has four subtypes: pleomorphic, alveolar, embryonal, and botryoid. Embryonal diversity is the most common and accounts for approximately 49% of all rhabdomyosarcomas.[4] Moreover, the clinical presentation of oral RMS can sometimes mimic a benign neoplasm or inflammatory process and, therefore, delay diagnosis.[5]

A 46-year-old male patient was admitted to our clinic with complaints of a mass growing over time in the right anterior mandibular region, increased mobility in the teeth in that region, and severe pain. Extraoral examination revealed swelling in the right anterior mandible and palpable submandibular lymph nodes. Intraoral examination of the patient revealed a mobility, exophytic, lobulated, sessile, and painless mass in the affected teeth. The lesion was located in the region of the right anterior teeth, approximately 3×3.5 cm in size, with indistinct borders, hard but bleeding on palpation (Fig. 1). Intraoral ultrasonographic examination revealed a highly perfusion hypoechoic lesion and destruction in the buccal cortical bone. Cone-beam computed tomography showed complete destruction of the cancellous bone in the buccal and lingual cortical bone surrounding the affected teeth and in the relevant region. It revealed that the teeth in the relevant area appeared "floating" (Fig. 2). After obtaining the patient's informed consent, an incisional biopsy was performed and the sample was sent for pathological examination. Under

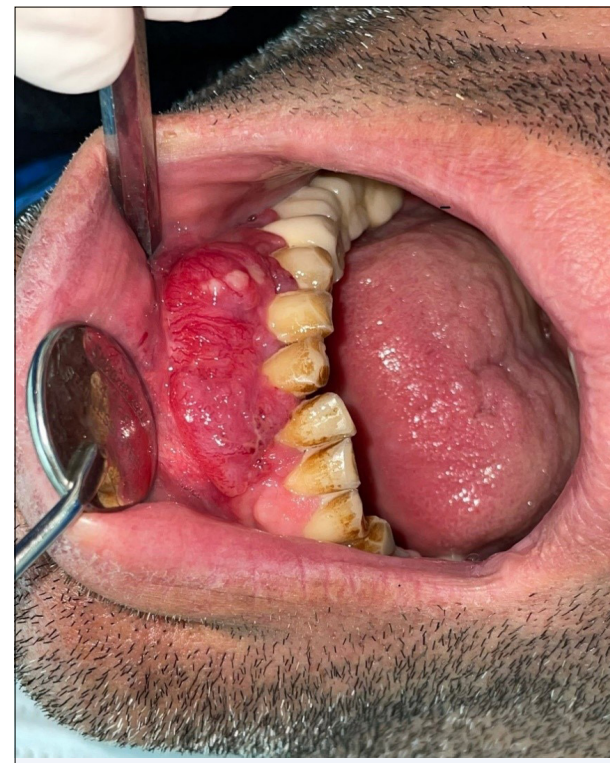

Fig. 1. Intraoral view and clinical feature of the lesion.

Received: December 13, 2022
Revised: January 08, 2023
Accepted: February 03, 2023
Online: February 14, 2023

Accessible online at:
www.onkder.org

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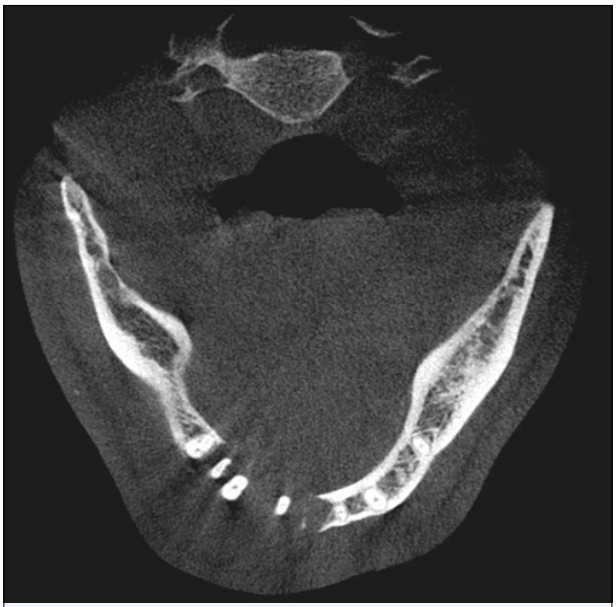

Fig. 2. Cone-beam computed tomography showed complete destruction of the cancellous bone in the buccal and lingual cortical bone surrounding the affected teeth and in the relevant region.

light microscopic examination, a tumoral lesion that started under the non-keratinized stratified squamous epithelium and caused partial ulceration in the mucosa was observed (Fig. 3a). The tumor consisted of oval-round eosinophilic cells with occasionally elongated nuclei and coarse chromatin. Tumor cells formed solid structures, focal prominent fascicles, and alveolar spaces (Fig. 3b). Many atypical mitoses and pleomorphic cells were also observed. While specific staining was not detected in the histochemical PAS staining applied for differential diagnosis, diffuse staining with vimentin, MyoD1 (Fig. 3c), desmin (Fig. 3d), Bcl-2, and focal staining with Pan CK and CD99 were observed immunohistochemically. Melan A, HMB-45, Sox-10, S-100, P40, CK18, Myogenin, ERG, Kaldesmon, SMA, NF, TLE-1, MUC-4, and CD57 were negative. In conjunction with this histological result, it resulted in the embryonal subtype of RMS being most supportive. The patient was referred to the oncology service and radiotherapy and chemotherapy treatment were started before the operation of the related tumor.

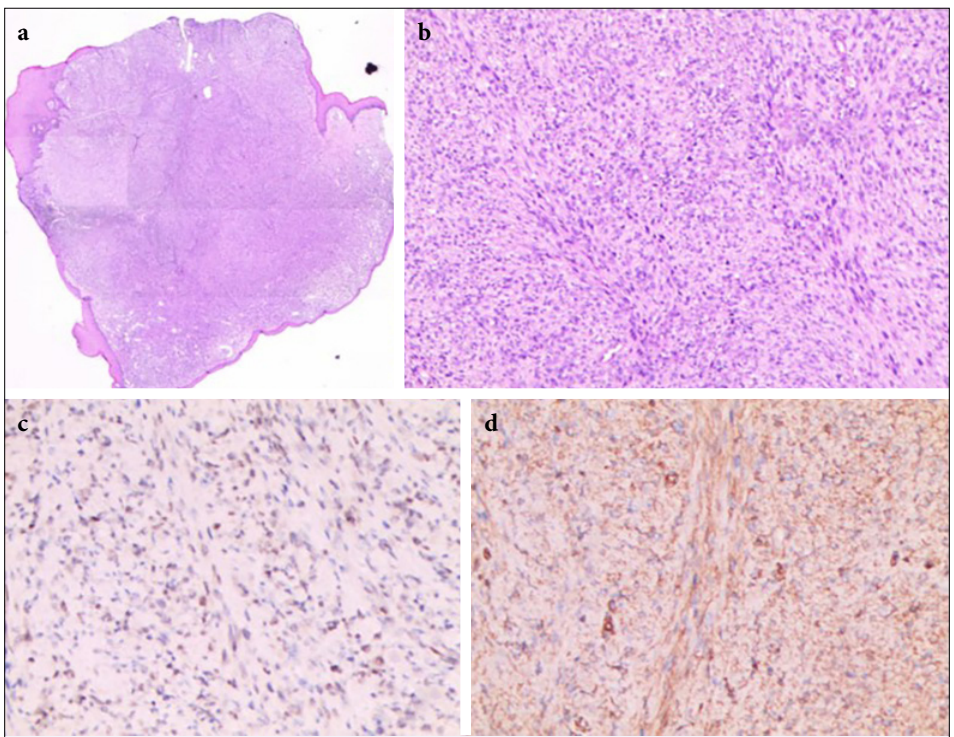

Fig. 3. (a) Light microscopic examination revealed a tumoral lesion that started under the non-keratinized stratified squamous epithelium and caused partial ulceration of the mucosa; (b) tumor cells formed solid structures, focal prominent fascicles, and alveolar spaces; (c) immunohistochemical staining of vimentin and MyoD1; and (d) immunohistochemical staining of desmin.

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It is always difficult to diagnose RMS due to its variable clinical presentation and histological diversity. It usually presents as a painless swelling in the early stages.[5] Other signs and symptoms of oral RMS are tooth mobility, pain, paresthesia, trismus, and cervical lymphadenopathy. Most of these symptoms were also observed in our case.

The classical embryonal variant has a 66% 5-year survival rate, while the botryoid and well-differentiated spindle variants have a 5-year survival rate of approximately 90%. Although embryonal prognosis is better at all ages compared to alveolar subtype, adults tend to be much worse than children regardless of subtype, as adult embryonal RMS still has a worse prognosis than childhood alveolar RMS.[6]

A PubMed search for adult RMS of the oral cavity revealed fewer than 10 case reports published in the English literature. A few cases of RMS affecting the oral tissues in adult patients have been presented in the literature.[3,7] This report adds to the literature by presenting a case of embryonal RMS affecting the gingiva, alveolar mucosa, and right anterior mandible.

RMS treatments include surgery, radiotherapy, and different chemotherapy regimens. Treatments are based on tumor stage and clinical manifestations.[8] Long-term treatment sequelae such as facial asymmetry, abnormalities in tooth formation, jaw hypoplasia, trismus, and hyposalivation/xerostomia may persist and significantly alter the patient's lifestyle.[9] Adult RMS should continue to be treated aggressively, but new and tailored treatment strategies are needed to improve long-term outcome.[10]

In conclusion, in multivariate analysis, age, histological subtype, primary site, stage, surgery, radiotherapy, and local control are the most important factors determining survival. Clinicians often encounter cases with facial swelling and pain. We believe that such swelling should be carefully examined in patients and the results of treatment should be followed up regularly. In such cases, high suspicion, early diagnosis, and multidisciplinary treatment approach are of great importance.

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