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A Primary Intraosseous Rhabdomyosarcoma of Mandible: Report of a Rare Case and Review of Literature

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A B S T R A C T

Rhabdomyosarcoma is a rare soft tissue malignant neoplasm with striated muscle differentiation often found in the head and neck region. In our case a 33-year-old man presented with a bulging mass near his right mandibular second molar tooth, identified as a unilocular destructive lesion on an X-ray. An incisional biopsy confirmed rhabdomyosarcoma, leading to a hemi-mandibulectomy. Histopathologic findings showed spindle and rhabdoid cells with positive immunoreactivity to Desmin, myogenin and myoD1. In conclusion, it is crucial to differentiate rhabdomyosarcoma from other more common intraosseous malignancies, and using Desmin staining as part of the initial immunohistochemical panel can aid in the diagnosis process.

Keywords: Rhabdomyosarcoma; Mandible; Neoplasm

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INTRODUCTION

Rhabdomyosarcoma is a rare malignant soft tissue neoplasm that originates from mesenchymal cells with striated muscle cell differentiation [1]. It accounts for only 1% of all malignancies, with 35% of cases occurring in the head and neck region [2,3] Oral cavity rhabdomyosarcoma is even rarer, representing 10-12% of cases of this type of sarcoma [4]. It is commonly found in the tongue, soft palate, and hard palate and typically affects younger individuals [2,4]. Here, we present a case of an adult patient with a mandibular mass diagnosed as primary intraosseous rhabdomyosarcoma.

CASE REPORT

A 33-year-old man presented with a swollen and ulcerated area in his oral cavity in October 2020. His right mandibular second molar tooth was extracted, and he remained symptom-free until January 2021 when a bulge developed in his right mandibular region. An incisional biopsy revealed a spindle cell neoplasm with positive immunoreactivity for Vimentin and Desmin stains and negative reactions for CD31, CD34, CD56, CD99 and

S100. The patient was referred to the Cancer Institute Hospital, Tehran, Iran for further evaluation. Imaging studies showed a destructive unilocular radiolucent lesion extending from the alveolar ridge to the inferior border of the mandibular body involving both buccal and lingual plates with perforation and soft tissue involvement. The lesion was in close proximity to the inferior alveolar nerve (Fig 1).

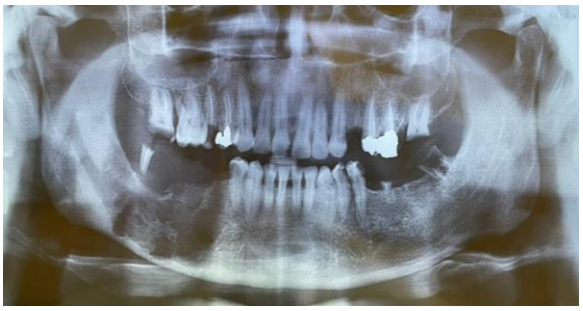

Fig 1. Panoramic view of the patient. The destructive lesion is seen at right mandibular region.

An excisional surgery was performed, and the specimen revealed a well-circumscribed mass measuring 4x2.5x2.5cm on the superior aspect of the mandiblecomposed of small round rhabdoid cells and spindle cells with hyperchromatic nuclei and prominent nucleoli with few mitotic figures (Fig 2). Immunohistochemical staining confirmed the primary diagnosis of intraosseous rhabdomyosarcoma, spindle cell variant. The neoplastic cells were negative for S100, SOX10, and CD34 but positive for Desmin, Myogenin, and Myo-D1 (Fig 3). The proliferation marker Ki-67 showed a proliferative activity of 20% in neoplastic cells.

DISCUSSION

Rhabdomyosarcoma is rarely seen in adults, and its etiology remains unclear [5]. It presents clinically with a wide range of symptoms, from a cutaneous facial nodule to a progressed painless facial swelling [6]. Histologically, four subtypes of this soft tissue tumor are described: alveolar, embryonal (the most common), botryoid and spindle cell. The spindle cell variant is the least common but has a better prognosis [7]. It is believed to originate from primitive mesenchymal cells, despite showing skeletal muscle differentiation [8]. Rhabdomyosarcoma is unlikely to originate from skeletal muscle fibers, as they are long-lasting cells that do not divide after birth. The interosseous origin of this neoplasm in adults has been rarely reported (Table 1). Our case is the fifth reported case of an adult with primary interosseous rhabdomyosarcoma and the third reported case involving the mandible. The differential diagnosis for our case includes locally aggressive odontogenic tumors, odontogenic carcinomas, locally aggressive non-odontogenic tumors, primary interosseous non-odontogenic carcinomas and malignant bone and cartilage forming tumors. Microscopic examination ruled out malignancies with neural differentiation, vascular-origin neoplasms, and sarcomas. Thus, SOX10 and S100 staining were negative, and CD34 staining ruled out vascular differentiation. Positive staining for Desmin, Myogenin, and MyoD1 in the neoplastic cells confirmed the diagnosis of rhabdomyosarcoma.

Table 1. Literature review

Author	Year	Patient's age	Symptoms	location	Histology subtype
Kimario, O.M [9]	2021	25	Facial swelling	Right maxilla	Alveolar type
Rodrigues [10]	2021	33	Asymptomatic gingival mass	Maxilla	Epithelioid
Joy [11]	2018	52	Facial swelling	Left mandible	Spindle cell
Sadhasivam [12]	2018	25	Facial swelling	Mandible	Spindle cell

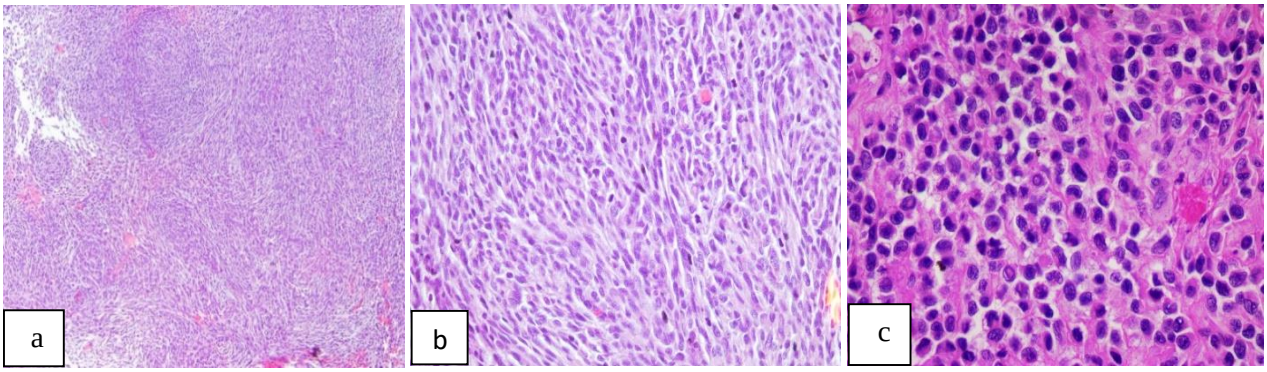

Fig 2. Hematoxylin and eosin staining reveals a hypercellular round to spindle cell neoplastic lesion with cellular atypia and scattered mitotic figures. a. x40, b. x100, c. x400.

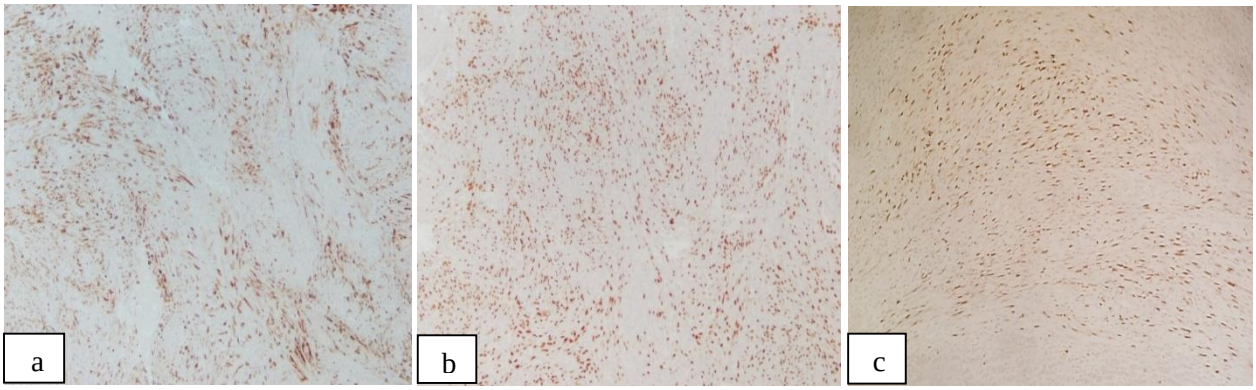

Fig 3. Immunohistochemical staining shows the expression of skeletal muscle antigen in neoplastic cells. a. Desmin X100, b. MyoD1 X100, c. Myogenin X100.

CONCLUSION

Primary intraosseous rhabdomyosarcoma is an extremely rare neoplasm that needs to be distinguished from a long-list of differential diagnoses. It may be advisable to use a Desmin stain as part of the initial immunohistochemical panel as a screening test to confirm or rule out rhabdomyosarcoma.

CONFLICT OF INTEREST STATEMENT

None declared.

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