CASE REPORT

Case Report: Rare site for intraoral meningioma [version 1; peer review: awaiting peer review]

Hatem Amer [1], Layla Hafed [2], Sally Ibrahim[3], Shady Shaker[1]

[1]Faculty of Dentistry, Cairo University, Cairo, Egypt
[2]Faculty of Dentistry, Ahram Canadian University, Cairo, Egypt
[3]Faculty of Dentistry, El-Fayoum University, Cairo, Egypt

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Abstract

Extracranial meningioma is very rare with few cases reported, especially in the oral cavity. Its diagnosis considered a challenge owing to the unusual site of occurrence. We report, to our knowledge, the first case of extra-cranial meningioma as a primary tumor in the palate with no detected intracranial extension. A 59-year-old ~~female Egyptian~~ patient presented with a 22-year history of a large painless swelling at the right side of the palate, which could not be seen on radiographs. An incisional biopsy was taken and, after assessment with a panel of immunohistochemical markers, the lesion was diagnosed as extracranical meningioma. The patient did not show up for surgical excision and follow-up was not performed because of ~~loose~~ of contact with the patient. Intraoral meningioma is a rare ~~unsuspected~~ tumor. Immuohistochemical markers are important ~~when confirming~~ this diagnosis.

Keywords

Intra-oral meningioma, Benign tumor, Ectopic meningioma, Palatal lesion

Open Peer Review

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Any reports and responses or comments on the article can be found at the end of the article.

Corresponding author: Layla Hafed (layla.hafed@gmail.com)

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Introduction

Meningioma is a benign neoplasm of meningothelial cells[1]. Meningioma may develop as a direct extension of a primary intra-cranial meningioma or as a true primary extra-cranial meningioma[2].

Extra-cranial (ectopic) tumors are mostly seen in the head and neck region with no connection intra-cranially[3]. The most common extra-cranial site is the orbits. Meningioma arising in the oral cavity is extremely rare[4]. To the ~~extent~~ of our knowledge, 18 cases have currently been reported in the oral cavity[2,4–19] and we are reporting the first case in the palate.

Case report

A 59-year-old female patient presented to the outpatient clinic in the Oral and Maxillofacial Surgery Department, Cairo University in January 2019 complaining of a large painless swelling in the palate (Figure 1). The patient reported that the swelling had been present in her oral cavity for 22 years. The patient's medical and familial histories were unremarkable. Upon clinical examination the day of admission, a large palatal swelling (3 cm × 3 cm) was evident on the right side of the hard palate. The swelling was covered by normal mucosa and showed a slight bluish tinge. The lesion was not visible on radiographs.

An incisional biopsy of the lesion was performed. Hematoxylin and eosin stained sections revealed meningothelial cells arranged in lobules. The cells exhibited round to oval nuclei (Figure 2). Psammoma bodies were also present (Figure 3). No mitotic activity and no cellular atypia were found. Immunohistochemical staining for tumor-associated markers was performed to confirm ~~a~~ diagnosis of meningioma ~~diagnosis~~ and to exclude other mimic tumors as metastiatic carcinomas, schwannoma, neurofibroma, paraganglioma and perineurioma. Cells were positively stained using primary antibodies for epithelial membrane antigen (EMA) and vimentin (Figure 4a, b), but were not stained when using primary antibodies for S100, pancytokeratin, p63, chromogranin and renal cell carcinoma glycoprotein (Figure 5a–e).

No therapy was administered to the patient during her admission. Unfortunately, the patient did not show up for surgical excision and follow-up.

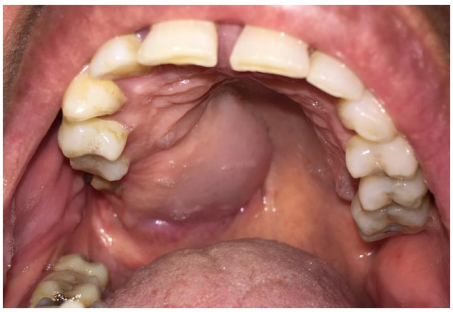

Figure 1. Preoperative clinical picture showing 3 × 3cm swelling in the palate.

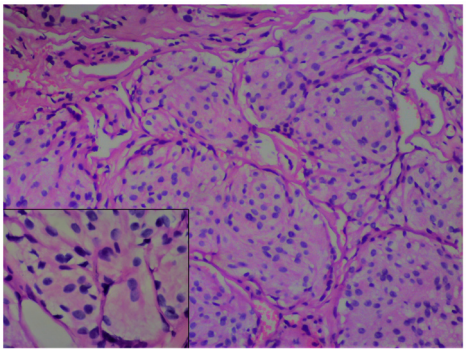

Figure 2. Hematoxylin and eosin-stained sections showing epithelioid cells forming meningiothelial whorls (magnification, ×100). Indistinct cell membranes with uniform nuclei and no mitotic figures (inset; magnification, ×200).

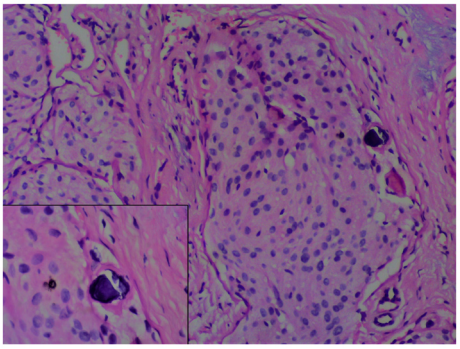

Figure 3. Hematoxylin and eosin-stained sections showing syncytial cells (magnification, ×100). Psammoma bodies seen between meningiothelial cells (inset), (×200).

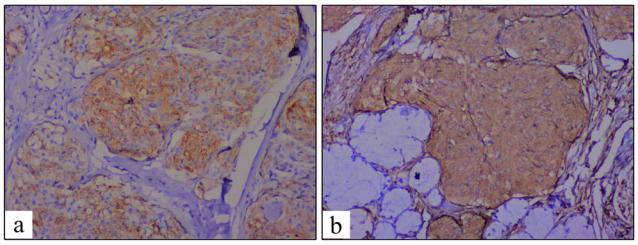

Figure 4. Meningioma tumor cells showing a positive cytoplasmic immunohistochemical reaction for (**a**) epithelial membrane antigen and (**b**) Vimentin (magnification, ×200).

Discussion

Primary extra-cranial meningioma is an unusual tumor, especially in the oral cavity[4]. The first intraoral meningioma reported was by Brown *et al.* in 1976, which presented as a periapical radiolucency anterior maxillary region[5].

To the ~~extent~~ of our knowledge, 18 cases of primary meningioma in the oral cavity have been reported. Of these, 13 were in female patients, which is also true ~~of~~ the present case. However, the age range was ~~large~~ in the reported cases – between

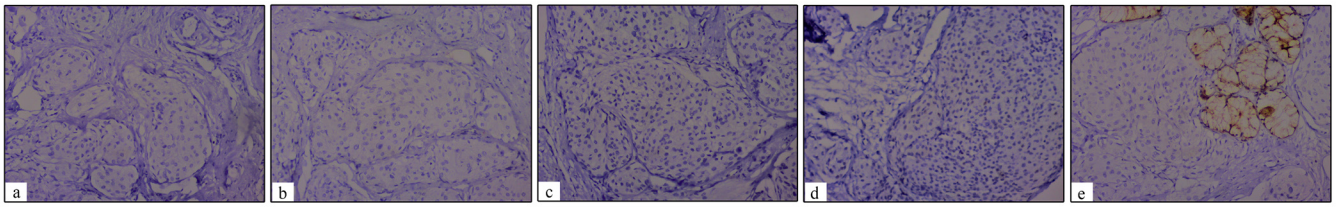

Figure 5. Meningioma tumor cells react negatively following immunohistochemical staining for (**a**) renal cell carcinoma glycoprotein, (**b**) S100, (**c**) chromoginin, (**d**) p63, (**e**) PanCK (magnification, ×100).

Table 1. Clinicopathological and radiographic data of the documented cases of extracranial meningioma.

Study	Age, years	Gender	Site	Tumor size	Radiographic findings	Treatment	Follow-up
Brown *et al.*[5]	69	M	Maxilla	NA	ML RL	Not completed	8 years
Simpson and Sneddon[6]	63	F	Maxillary alveolus	4.5 × 2.7 × 2.7 cm	Well-defined mixed RL RO	Surgical excision.	Under review
Landini and Kitano[7]	48	F	Mandible	NA	Well-defined RL	Block resection	2 years
Reddi *et al.*[2]	26	F	Maxilla	3 cm	Ill-defined RL	Surgical excision	2 years
Pfeifer *et al.*[8]	77	F	Maxilla (temporal fossa)	NA	Dense soft tissue mass	Surgical resection	NS
Jones and Freedman[9]	41	F	Mandible	4 × 2 cm	Well defined RL	Excisional biopsy	NS
Jones and Freedman[9]	74	F	Mandible	4 × 3 cm	Well-defined RL	Excisional biopsy	NS
Kubotaa *et al.*[10]	10	M	Mandible	NA	Well-defined RL	Enucleated	4 years
Mussak *et al.*[11]	62	M	Mandible	7 × 3 cm	Well-defined RL	Segmental mandibulectomy	NS
Lell *et al.*[12]	40	F	Mandible	NA	Well-defined RL	NS	NS
Mosquede-Taylor *et al.*[13]	53	F	Mandible	4 cm	Ill-defined mixed RO RL	Surgical excision	6 months
Rushing *et al.*[14]	NA		Mandible	NA			
Simsek and Komerik[4]	51	F	Maxilla	2 × 2 cm	Ill-defined mixed RL-RO	Surgical excision	5 years
Pinting *et al.*[15]	59	M	Maxilla	NA	Well-defined RL	Surgical excision and radiotherapy	NS
Maeng *et al.*[16]	66	F	Buccal mucosa	2 cm	Heterogenously enhanced mass	Surgical excision	Year and half
Nair *et al.*[17]	60	F	Buccal mucosa	4 × 3 cm	Mass of heterogeneous density	Surgical resection	One year
Rege *et al.*[18]	35	M	Mandible	NA	Ill-defined ML RL	Partial resection	5 years
Rommel *et al.*[19]	20	F	Mandible	2 × 1.8 cm	Well defined RL	No surgical intervention.	One year

M, male; F, female; RL, radiolucent; RO, radioopaque; UL, unilocular; ML, multilocular; NA, not available; NS, not stated.

10 and 77 years old[2,4–19]; in the present case, the patient was 59 years old. Regarding the reported cases of intraoral primary meningioma, 6 of the 18 were in the maxilla[2,4–6,8,10,15], 10 were in the mandible[7,9–14,18] and 2 in the buccal mucosa[16,17]. To our knowledge, we report the first case in the palate.

The histopathological criteria of extracranial meningiomas are similar to those of their intracranial counterparts. All documented cases shared the same characteristics: whorls of spindle cells or epithelioid cell proliferation and psammoma bodies. In our case, diagnosis was challenging because of the tumor's

similarity with other tumor entities of peripheral nerve origin, as well as the uncommon location of the tumor. An immunohistochemical panel of tumor-associated markers ~~were~~ used to confirm the diagnosis and to avoid unnecessary aggressive treatment. Most of the 18 cases reported in the literature ~~achieved their diagnosis~~ using immunohistochemical markers. All reported cases that used immunohistochemistry techniques to diagnose meningioma[4,8–10,12,13,15,16,18,19] observed that the tumor cells stained positive for monoclonal antibodies against EMA and vimentin, with no immunoreactivity for S-100 protein, which was similar to our findings.

Unfortunately, our patient did not show up for surgical excision and follow-up was not done because of ~~loose~~ of contact with the patient. However, most of the documented cases ~~was~~ treated successfully without recurrence by surgical excision. Some of studies, such as that by Rommel *et al.*[19], preferred only to follow-up ~~with~~ the patient rather than conduct surgical intervention. However, others preferred to perform aggressive treatment, such as ~~as~~ segmental mandibulectomy or ~~segmented~~ resection[7,11]

In conclusion, meningioma is a rare intraoral benign neoplasm. Immunohistochemical markers are an important tool to achieve a final diagnosis, especially for the differentiation from histological mimic entities of peripheral nerve origin, such as perineurioma and neurothekeoma. Vimentin and EMA are the two important markers to confirm extra-cranial meningioma diagnosis ~~from other lesions~~.

Data availability

All data underlying the results are available as part of the article and no additional source data are required.

Consent

Written informed consent for publication of their clinical details and clinical images was obtained from the patient.

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