

CASE REPORT

Ectopic meningioma of the mandible in a 20-year-old woman: a case report and literature review

Niklas Rommel*[†], Oliver Bissinger[†], Andrea Rau, and Thomas Muecke

Department of Oral and Maxillofacial Surgery, Technische Universität München, Klinikum rechts der Isar, Munich, Germany

*Correspondence address. Department of Oral and Maxillofacial Surgery, Technische Universität München, Klinikum rechts der Isar, Ismaninger Str. 22, D-81675 Munich, Germany. Tel: +49-4140-9744; Fax: +49-89-4140-2934; E-mail: niklasrommel@tum.de

Abstract

Ectopic meningiomas are a very rare tumor entity. We present a case of a meningioma arising in the mandible of a young woman and initially supposed to be a radicular cyst. Histopathological and immunohistochemical evaluation showed typical cell characteristics of a meningioma. Only six cases of ectopic meningiomas in the mandible have been described in the literature until now, mainly in women at an advanced age and with surgical removal of all tumors. For the first time, no surgical excision has been performed in this case and follow-up control after 12 months showed no significant progression or increasing clinical complaints. Hence, surgical removal seems non-urgent. In conclusion, unclear lesions of the jaws, even if they seem to be clear following diagnostics, should be evaluated by incisional biopsy and histopathological evaluation.

INTRODUCTION

Meningiomas are one of the most common neoplasms arising from cellular elements of the meninges, in particular, from arachnoid villi structures. Commonly, meningiomas are attached to the dura and grow in the cranial cavity or intraspinal region. However, ectopic locations of meningiomas cannot be excluded. In this report, we present a rare case of this special tumor entity in the mandibular bone of a young woman.

CASE REPORT

A 20-year-old woman was referred to our Department with the suspicion of a radicular cyst resulting from a periapical infection of tooth 36 in the mandibular bone. The patient reported slight local pain in the left mandible over a long period. On radiographic examination, panoramic X-ray and computed tomography (CT) imaging showed a 2 × 1.8-cm radiolucent lesion of



Figure 1: Radiologic diagnostics in two planes with panoramic X-ray. The panoramic radiograph showed a 2 × 1.8-cm radiolucent lesion of the left mandible. The lesion involved both apices of the first molar and the mesial apex of the second molar and extended to the caudal border of the mandibular bone.

[†]These authors contributed equally to the manuscript.

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Figure 2: Axial, sagittal and coronar CT showing a 2 × 1 × 1.8-cm lesion in the left posterior mandible, its expansive and destructive character, and its penetration of the medial corticalis.

the left posterior mandible. The lesion involved both apical roots of the first molar and the mesial apical root of the second molar. It showed an expansive character and penetrated the medial corticalis of the mandibular bone (Figs 1 and 2).

After completion of a discussion of the results, the patient was treated under general anesthesia by intraoral subtotal incisional biopsy by using Piezosurgery (Mectron, Cologne, Germany). Microscopic examination revealed a mesenchymal tumor with fragments of lamellar bone including resorption signs. The tumor had a high cell density including fascicular, storiform and spindle-shaped patterns. The cells were ordered into cell cords, but also into whorls. Additionally, collagenous and hard matter areas were present evoking psammom bodies, and the cells clearly formed reticulin fibers. No mitotic activity and no cell atypia were found (Fig. 3A). Using avidin–biotin complex immunostaining, we found a strong positive reaction with antibodies for vimentin, epithelial membrane antigen (EMA) and somatostatin and also a positive reaction for desmoplakin in a smaller amount of tumor cells (<20%) (Fig. 3B–D). The final histopathological results of the biopsy revealed a mesenchymal tumor classified as an ectopic meningioma WHO grade I.

DISCUSSION

Meningiomas are one of the most common tumor entities in the central nervous system, are generally benign and have their origin in the arachnoid villoid structures of the meningocytes. However, in rare cases, ectopic forms of this tumor entity can appear extracranially and extraosseously in the head and neck region. With respect to the jaws, we have found only eight cases including two meningiomas of the maxilla [1, 2] and six meningiomas of the mandible [3–7] in the current literature. We have found the seventh case of an extracranial meningioma of the mandible in a young woman who presented with a cystoid-like lesion in the left mandibular bone and no specific clinical symptoms. Because of the absence of typical radiographic features, no clear diagnosis was possible either with a panoramic X-ray or 3D imaging. However, the role of CT is seen significantly to assess the relationship between the tumor and the bony surfaces and to exclude potential malignancy [8].

Several hypotheses have been proposed for the occurrence of extracranial meningiomas, including extradural enclosing of arachnoid cell nests during embryogenesis, ectopic migration, and the development of arachnoid cells in combination with the peripheral nerves, or metaplasia of the mature peripheral nerve sheath cells or progenitor cells [8]. However, ectopic meningiomas have also been postulated to be mesenchymal tumors that arise from multipotential mesenchymal cells, particularly if no associations to the cranial nerves are apparent [9]. In the head and neck region, this tumor entity is often

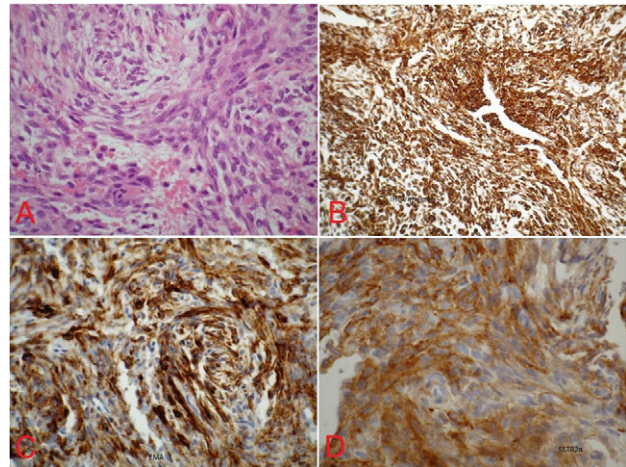


Figure 3: (A) Spindle-shaped cells arranged in cell cord and in whorls with no mitotic figures (hematoxylin–eosin, original magnification ×400). Immunohistochemical stains with immunopositive reaction for (B) vimentin (×100), (C) EMA (×400) and (D) somatostatin (×400).



Figure 4: Follow-up panoramic X-ray imaging at 12 months after initial diagnosis showing no significant progression of the tumor lesion.

associated with cranial nerves and, therefore, is considered to be derived from ectopic arachnoid tissue present around these nerves [3]. Therefore, the origin of the tumor arising in our patient seems to be the perineural cells of the mandibular nerve. However, ectopic arachnoid cells within the mandibular bone as a source of tumor growth cannot be excluded.

The histopathologic and immunohistochemical features of ectopic meningiomas are seen similar to those of their more frequent intracranial counterparts [4, 8]. The general histologic diagnosis of meningiomas describes spindle-shaped cells that



Figure 5: Likewise to panoramic X-ray, follow-up CT after 12 months showing no significant progression of the lesion.

can be arranged in whorls, rosettes and interconnecting fascicles. Mitotic figures or atypia is generally rare, and psammom bodies might be present [4]. Because of the differential diagnosis from other tumor entities of peripheral nerve origin, an immunohistochemical analysis is useful or even necessary. In meningiomas, the positive expression of EMA, desmoplakin, somatostatin and vimentin is a characteristic feature [3, 4, 10] and demonstrates the epithelial and mesenchymal patterns of cells within this tumor.

Epidemiologically, ectopic meningiomas are slightly more frequent in females with a ratio of 1:1.2, i.e. ~55% [8]. The average age of patients is 43.4 years, whereby females are older (48.7 years) than males (36.9 years) [8]. On closer consideration of extracranial meningiomas of the mandibular bone, all previously described cases in the current literature were women with an average age of 45.7 years. In our case, the gender was the same, but the age of the patient was, at 20 years, considerably younger than the average.

The therapy of choice for extracranial meningiomas in the jaw bones is surgical excision [3, 4, 8]. The prognosis of extracranial meningiomas after complete surgical tumor resection is good with disease-free rates of 82 and 78% at 5 and 10 years, respectively. In cases of subtotal resection or aggressive histopathologic features such as mitotic figures or foci of necrosis, close follow-up is necessary. Despite the strong recommendation for surgical treatment in the current literature, we decided on close follow-up examinations without any therapeutic interventions at the time of diagnosis, considering potential tooth loss or surgical damage of the inferior alveolar nerve as a potential complication of surgery. The follow-up examinations at 12 months including panoramic X-ray (Fig. 4) and CT imaging (Fig. 5) showed no significant progression of the tumor lesion and no new clinical symptoms were apparent. These observations indicated that immediate surgical treatment was not mandatory. This case remains under review and is the first case of an ectopic mandibular meningioma that has not initially been surgically removed after diagnosis.

The conclusion of this case is that unclear lesions of the jaws, even if they seem to be clear following diagnostics, should be evaluated by incisional biopsy and histopathological evaluation.

CONFLICT OF INTEREST STATEMENT

None declared.

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REFERENCES

1. Simpson MT, Sneddon KJ. Extracranial meningioma of the oral cavity. *Br J Oral Maxillofac Surg* 1987;25:520–5.
2. Reddi SP, Strauss SI, Strauss JE, Blanchaert RH Jr. Anterior maxillary lesion. *J Oral Maxillofac Surg* 1999;57:1234–8.
3. Landini G, Kitano M. Meningioma of the mandible. *Cancer* 1992;69:2917–20.
4. Jones AC, Freedman PD. Primary extracranial meningioma of the mandible: a report of 2 cases and a review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2001; 91:338–41.
5. Mosqueda-Taylor A, Dominguez-Malagon H, Cano-Valdez AM, Montiel-Hernandez AM. Primary extracranial meningioma of the mandible. *Med Oral Patol Oral Cir Bucal* 2009;14: E167–70.
6. Lell M, Tudor C, Aigner T, Kessler P. Primary intraosseous meningioma of the mandible: CT and MR imaging features. *AJNR Am J Neuroradiol* 2007;28:129–31.
7. Deshmukh SD, Rokade VV, Pathak GS, Nemade SV, Ashturkar AV. Primary extra-cranial meningioma in the right submandibular region of an 18-year-old woman: a case report. *J Med Case Rep* 2011;5:271.
8. Iaconetta G, Santella A, Friscia M, Abbate V, Califano L. Extracranial primary and secondary meningiomas. *Int J Oral Maxillofac Surg* 2012;41:211–7.
9. Shuangshoti S, Panyathanya R. Ectopic meningiomas. *Arch Otolaryngol* 1973;98:102–5.
10. Golemi A, Ambrosini A, Cecchi P, Ruii A, Chondrogiannis S, Farsad M, et al. (68)Ga-DOTANOC PET/CT detection of multiple extracranial localizations in a patient with anaplastic meningioma. *Rev Esp Med Nucl Imagen Mol* 2015;34: 258–60.