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Primary extracranial meningioma of the mandible

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Abstract

Meningiomas are benign tumors of mesodermal origin that arise from arachnoid cell clusters that penetrate the dura to form arachnoid villi. These neoplasms represent one of the most common neoplasms developing within the central nervous system and are usually located at points of entry of vessels and nerves through the dura. Extracranial meningiomas (EM) comprise only 2% of all meningiomas, and only six cases of primary EM of the jawbones have been described to date. They may arise as an extension of intracranial meningiomas or as primary tumors and may be clinically indistinguishable from other benign tumours of the jaws, as they usually present as a well-delineated unencapsulated tumors. In this article a case of primary intramandibular primary EM that appeared as a well-defined osteolytic radiolucent lesion of the jaw is reported. The salient clinico-pathological features of this case is compared to those previously reported in the literature and differential diagnosis and therapeutic considerations are discussed.

Key words: *Meningioma, jaws, extra-axial meningioma.*

Introduction

Meningiomas are benign tumors of mesodermal origin that represent one of the most common neoplasms developing within the central nervous system. They arise from meningeal arachnoid cells that normally line the inner and outer aspects of arachnoid membrane. The occurrence of extracranial meningiomas (EM) is very rare, and they comprise 2% of all meningiomas. These lesions may occur as a direct extension from a primary intracranial meningioma (IM) or as true primary EM, which possibly develop from ectopic arachnoid cells (1). The aim of this article is to present a case of primary intramandibular EM, as well as to review the literature on these extremely rare lesions.

Case Reports

A 53 year-old female was admitted to the hospital with the main complaint of volume increase in the right mandibular region. A radiological study (orthopantomography) revealed a 4 cm osteolytic lesion with well defined borders, located in the molar area of the right mandibular body (Fig.1). An incisional biopsy was performed in which the surgeon found a solid tumor composed of soft tissue covered by a very thin bone cortex. The pathological diagnosis of primary mandibular meningioma was established and the patient was submitted to local resection, with no evidence of tumor recurrence after six months. Biopsy specimen consisted in two irregular fragments of pink-brown soft tissue, measuring 1.5 cm in their greatest dimension. Microscopically, the tumor was composed of a homogeneous population of polygonal to spindled cells arranged in solid sheets and trabeculae surrounded by septae of connective tissue showing abundant thick-walled blood vessels (Fig.2). The cells disclosed ample cytoplasm with a fibrillary appearance and faint cell borders. The nuclei were ovoid, with uniform size and shape. Nuclear chromatin was bland with inconspicuous nucleoli; many cells disclosed large cytoplasmic pseudo-inclusions (Fig.3), and no mitotic

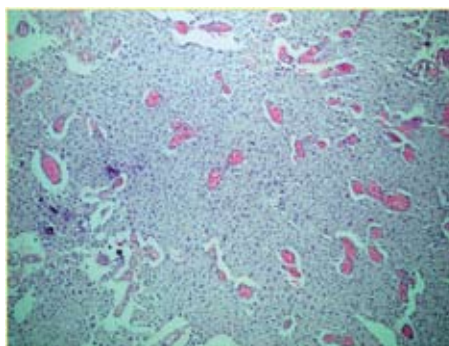


Fig. 2. Histological appearance of the tumor. The polygonal cells are arranged in solid sheaths surrounded by septae with prominent vasculature and some Psammoma bodies are identified.

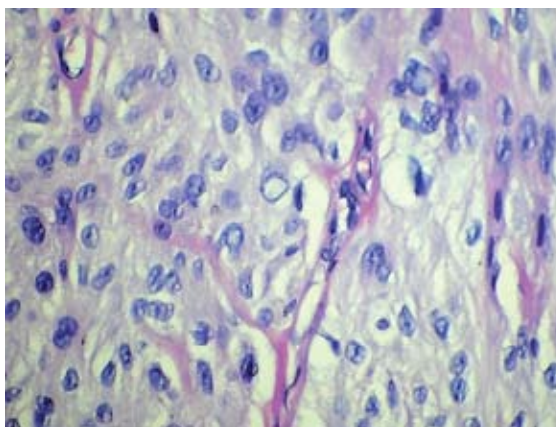


Fig. 3. The cells have faint borders and regular nuclei with frequent cytoplasmic pseudo-inclusions.



Fig. 1. Osteolytic lesion located in the molar area of the right mandibular body.

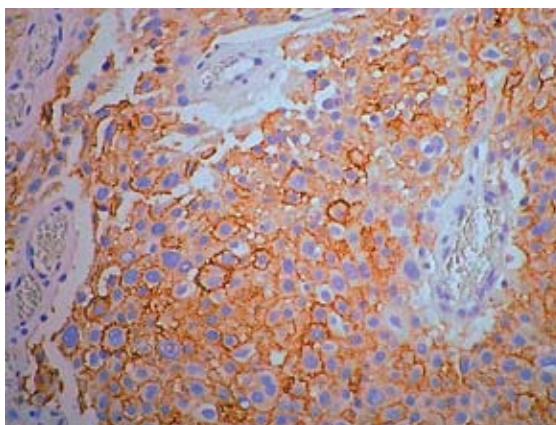


Fig. 4. The cell membranes are intensely positive for EMA.

activity was detected. Psammoma bodies were occasionally identified within the solid sheets of tumoral cells (Fig.2). The cells were positive for vimentin and epithelial membrane antigen (EMA) (Fig.4), and they were negative for AE1/AE3 cytokeratin, CK 18, S-100 protein and CD34. In addition, a very low expression index for ki-67 was found (<1%).

Discussion

Primary EM developing within the jaw bones are extremely rare neoplasms. In addition to the present example, only six cases have been reported in the English language literature (1-5). All but one case have occurred in women, whose ages ranged from 26 to 74 years, with a mean of 52.4 years. Five cases developed centrally within the mandible (four in the molar area and one in the anterior region) and two were located in the maxilla. Table 1 shows the salient clinical and radiological findings of the cases reported to date. It is worth noting that the vast majority of the reported cases of mandibular meningioma, including the present one, have occurred in women, which is in accordance with the fact that meningiomas in general have a marked predilection for female gender, and that it has been postulated that hormones play a significant role in the development of these neoplasms, since they commonly express progesterone receptor. In fact, meningiomas are particularly frequent in women with breast carcinoma, and they tend to enlarge during pregnancy and during the luteal phase of the menstrual cycle (6); however, to date there is no satisfactory explanation for this female predominance.

It is interesting to note the variable radiographic pattern that have been described in the cases of EM of the jaws, which range from well-defined radiolucent lesions to ill-defined mixed radiolucent and radiopaque defects

(1-5). Although this variable pattern makes necessary to include in the differential diagnosis a large number of lesions, the slow growth observed in most cases reduces the likelihood of being a malignant or an inflammatory condition, and therefore a benign odontogenic tumor or a fibro-osseous lesion would be the favored presumptive diagnosis. The radiopaque material identified in some cases may be the consequence of psammoma bodies and/or other foci of mineralization located within the mass; however, because of the rarity of EM involving the mandible and the high frequency of tumors that may produce mineralized substance in this bone, the internal calcification may not be a good indicator of the radiographic diagnosis (5).

Although fine needle aspiration cytology has proved to be a useful method for diagnosis of EM located in other sites of the head and neck (7), no such procedure has been performed for intra-osseous lesions of the jawbones; therefore, definitive diagnosis in cases located centrally within the jaws had come through suitable biopsy specimens.

Meningiomas are usually divided into different histologic subtypes, such as meningotheliomatous, transitional, fibroblastic, psammomatous and angioblastic, according to the predominant cellular morphology (8). Excluding the angioblastic meningioma, which is a relatively aggressive variety, no significant biologic or prognostic differences have been found among the other histologic subtypes (9).

According to the microscopic descriptions of the reported cases and the published photomicrographs, four cases can be classified as transitional, two meningotheliomatous and one fibroblastic. These neoplasms are unencapsulated but usually well circumscribed tumors, composed of uniform spindle-shaped or polygonal cells

Table 1. Salient clinical and radiological findings of primary extracranial meningiomas.

Case	(Ref)	Age	Gender	Location	Rx image	Histologic subtype	Treatment	Follow-up
1	Reddi et al ¹	26	F	Right anterior maxilla	Ill-defined mixed RL-RO	Meningotheliomatous	Surgical excision	2 yr. NED
2	Simpson and Sneddon ²	63	F	Left posterior maxilla	Well defined mixed RL-RL	Transitional	Surgical excision	NA
3	Landini and Kitano ³	48	F	Left posterior mandible	Well-defined RL	Fibroblastic	Block resection	2 yr. NED
4	Jones and Freedman ⁴	74	F	Right anterior mandible	Well-defined RL	Transitional	Surgical excision	NA
5	Jones and Freedman ⁴	41	F	Right posterior mandible	Well-defined RL	Transitional	Surgical excision	NA
6	Mussak et al ⁵	62	M	Left posterior mandible	Well-defined RL	Transitional	Segmental resection	NA
7	Present case	53	F	Right posterior mandible	Ill-defined mixed RL-RO	Meningotheliomatous	Surgical excision	6 mo. NED

with pale eosinophilic cytoplasm and oval-shaped nuclei, which tend to be arranged in whorls, rosettes and interconnected fascicles. The tumors may have a predominantly spindled or predominantly epithelioid morphology or a combination of both. There may be also nuclear palisades resembling peripheral nerve tumors. Neither mitotic figures or atypia were seen in the tumor cells, and the presence and amount of psammoma bodies varies from case to case.

Immunohistochemical analysis of the reported cases of EM of the jaws are similar to their intracranial counterpart, as there has been reported intense cytoplasmic positivity of the tumor cells for vimentin and membrane positivity for EMA (10). Expression of S-100 protein and anti neuron-specific enolase were demonstrated in some cases in adjacent nerve axons, while anti-cytokeratin and anti-smooth muscle actin were negative (1,3,4).

There are several possible mechanisms to explain the occurrence of meningiomas outside the central nervous system. Most cases represent direct extension from intracranial or intraspinal tumors. Other cases may result from proliferation of perineural cells of cranial or spinal nerves. Since the perineurium is a direct extension of the arachnoid membrane, it may produce a meningioma extending along any of these pathways (7). Also, these neoplasms may be the result of proliferation of ectopic arachnoid tissue along the course of cranial nerves (11,12). Other authors suggest that the tumor may arise from misplaced embryonic rests of arachnoid cells, or from multipotential mesenchymal cells (13-16).

Regarding the origin of primary mandibular EM, it has been suggested that these tumors may develop from the ectopic arachnoid villi or from perineural cells of the mandibular nerve (3). As three of the five cases of EM of the jaws reported to date have occurred in edentulous areas, some authors (3,4) consider that nerve injury during the extraction of teeth, in addition to chronic inflammation of the surrounding tissues might be responsible for the stimulation and proliferation of ectopic arachnoid tissue that lead to the development of an extracranial meningioma.

There are, however, two cases that have developed in tooth-bearing areas with no evidence of traumatic antecedents which makes difficult to accept that hypothesis as the only one to explain this unusual location for EM (1,4).

Treatment of choice for extracranial meningiomas is surgical excision. The prognosis is good after complete resection. All the reported cases have been treated in this way, and given the long evolution and the lack of recurrence in all of them, it may be concluded that this lesion behaves in a non aggressive fashion. However, due to the limited number of cases found within the jawbones, long-term follow-up is recommended to confirm this observation.

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