ECTOPIC MENINGIOMA OF THE MANDIBLE: A RARE SITE OF PRESENTATION FOR A COMMON TUMOUR

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ABSTRACT

Meningiomas are leptomeningeal tumours that commonly occur intracranially. The other favoured sites include intraspinal and orbital regions. According to the WHO, case reports of meningioma occurrence have been published in almost all organs, albeit rarely.⁽¹⁾ These are known as ectopic meningiomas. To date only 5 cases of meningioma of mandible have been reported in the English literature, one of those having been reported in a 10-year-old child with basal cell nevus syndrome.^(2,3,4,5) We are hereby presenting the case of a 41-year-old female patient with a long-standing swelling in the right submandibular region. On imaging her mandible showed a depressed eroded area and the salivary gland was visualized separately. The tumour was excised and reported histopathologically as ectopic meningioma of the mandible. The diagnosis was later confirmed with immunohistochemistry.

KEYWORDS

Meningioma, Ectopic, Mandible.

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INTRODUCTION

Meningioma is a common leptomeningeal tumour usually restricted to the intracranial, intraspinal and orbital regions. Rarely these tumours can occur at other sites.⁽¹⁾ Such meningiomas are termed as ectopic meningiomas. Mandible is an extremely rare site of presentation for these tumours. To date, only five such cases have been published in the English literature.⁽²⁻⁵⁾ We present one such case of ectopic meningioma occurring in the mandible of a 40-year-old female patient. We have supported our histopathological observations with immunohistochemical studies, which showed positive staining for vimentin and EMA in the tumour. The Ki-67 index was around 2%, insufficient for a diagnosis of atypical (Grade II) meningioma.⁽¹⁾

CASE REPORT

A 41-year-old female patient presented with a history of an insidious onset, gradually progressive swelling of 10 years' duration located in the right submandibular region. Imaging studies showed that the tumour was not connected to the submandibular salivary gland. The tumour was attached to the mandible. At the site of attachment, the mandible showed a depressed eroded region.

The surgeon did an excision biopsy of the tumour and we received a greyish-white soft mass measuring 2.5x1.5x1.5 centimetres in size. It had a greyish white fibrous appearing cut surface.

Fig. 1: CT of Mandible showing Focus of Erosion

Fig. 2: Cross-Section of Tumour Mass

Histologically, the tumour was composed of epithelioid cells in syncytial sheets as well as spindled cells arranged in whorls and short fascicles. Nuclei were round to ovoid with open chromatin. There were not any features suggestive of atypical or anaplastic meningioma such as nuclear

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pleomorphism, increased mitosis, atypical mitosis or necrosis. Morphologically, the tumour was reported as extracranial meningioma. It was confirmed by a positive immunostaining for vimentin and EMA by most of the tumour cells. S100 showed positive reaction focally.

Fig. 3: Spindle Cells in Fascicles. H&E; x 100

Fig. 4: Epithelioid Cells in Whorls. H&E; x 100

Fig. 5: Elongated Plump Nuclei with Open Chromatin. H&E; x 450

Fig. 6: Vimentin Positivity in Almost all Tumour Cells

Fig. 7: EMA Positivity in Most of the Tumour Cells

Fig. 8: S 100 Positivity in Very Few Tumour Cells

DISCUSSION

Meningiomas are common in intracranial, intraspinal and orbital regions where leptomeninges cover the brain, spinal cord and the orbital nerve, respectively.⁽¹⁾ Ectopic meningiomas are rare. The sites where ectopic meningiomas have been reported with a higher frequency include those where neural tube closure problems during intrauterine life could have led to the development of meningeal rests. These are classified as type 1 ectopic meningiomas.

These sites are skin and soft tissue of the scalp and along the vertebral axis. Type 2 sites include regions near sense organs and along cranial and spinal nerve pathways. Ectopic meningiomas are extremely rare where meningeal rests are non-existent, such as the case in mandibular meningioma.⁽⁶⁾ Ackerman proposes that the cell of origin at such sites could be perineurial cells.⁽⁷⁾

To date only four cases of meningioma of mandible have been reported in the English literature. The first ever case was reported in 1992, in the Journal, Cancer. The authors supported their diagnosis with immunohistochemical as well as ultrastructural studies.⁽²⁾ The next report came out in 2001 of 2 female patients presenting with mandibular lesions reported as meningiomas by histopathology and immunohistochemical studies.⁽³⁾ The last case reported in 2007 was in a 62-year-old man. This was the first ever radiological description of a Mandibular meningioma in published literature.⁽⁴⁾ One other case has been reported in a 10-year-old boy with basal cell nevus syndrome.⁽⁵⁾

The main differential diagnosis at this site is perineurioma. The cells in perineurioma are longer and thinner with elongated spindled nuclei as opposed to the plump nuclei of meningioma cells. The cells in perineurioma tend to form perivascular whorls and in intraneural cases they form pseudo-onion bulbs around residual nerve fibres which stain positive for S100.⁽⁸⁾

At other sites such as the nasopharynx, an ectopic meningioma may mimic angiofibroma or olfactory neuroblastoma clinically.

Histology and immunohistochemistry can easily differentiate these tumours from meningioma. The

psammomatous variant of meningioma at ectopic sites near bone will need to be differentiated from aggressive psammomatoid ossifying fibroma. These tumours occur at a younger age and show the presence of osteoclasts and osteoblasts.⁽⁸⁾

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