Primary intraosseous osteolytic meningioma: a case report and review of the literature

Sae Min Kwon, Yong Ko and Seong Sik Bang

Abstract

Background: Primary intraosseous meningioma is a subset of extradural meningioma that arises in the bone, and only a few cases have been reported to date.

Case presentation: An 80-year-old man presented with decreased hearing on the right side accompanied by a disturbance of balance 10 months prior to admission. Magnetic resonance imaging revealed an 8 × 7 cm osteolytic mass in the right posterior fossa related to the petrous bone, with extension to the cervical region. During surgery, the tumor was found to be located extradurally, with no invasion of the dura. The tumor was removed entirely, apart from a small portion around the jugular foramen to avoid lower cranial nerve injury.

Conclusion: The final diagnosis was primary intraosseous osteolytic meningioma with atypical pathology. Here, we report a rare case of an osteolytic skull lesion in the skull base not invading the dura and with extensive bone destruction.

Keywords: Intraosseous, Meningioma, Osteolysis

Background

Meningiomas are common intradural lesions that arise from the arachnoid cap cells of the arachnoid layer. In contrast, primary extradural meningioma is a relatively rare entity, accounting for less than 2% of all meningiomas [1, 2]. They may arise from other locations, such as the skin, orbit, nasopharynx, and neck [3–5]. Primary intraosseous meningioma, which arises in the bone, is a subset of primary extradural meningioma, and only a few cases have been reported [1, 3]. Here, we report a recent case of primary intraosseous osteolytic meningioma with extension to the cervical region which was successfully removed.

Case presentation

An 80-year-old man presented with a progressive decrease in hearing on the right side accompanied by dizziness and disturbance of balance 10 months prior to admission. The neurological examination revealed right hypoglossal nerve palsy. Audiometry documented complete sensorineural hearing loss on the right side.

Skull x-ray and cranial computed tomography (CT) scans showed a large osteolytic lesion with bone destruction, including the temporal bone, occipital bone, clivus, jugular foramen, and hypoglossal canal (Fig. 1a and b). Magnetic resonance imaging (MRI) revealed an 8 × 7 cm homogeneous enhancing mass in the right posterior fossa related to the petrous part of the temporal bone, with extension to the cervical region (Fig. 1c). The cerebellum was displaced, and definite brain invasion was not seen. The preoperative diagnosis was a temporal bone origin malignancy such as squamous cell carcinoma or meningioma with invasion of the petrous bone.

The patient underwent surgery to obtain a pathological diagnosis and for complete removal of the mass. A C-shaped postauricular skin incision was made that extended to the neck. The scalp was reflected anteriorly, and the mass infiltrating the subcutaneous tissue was exposed. The lesion appeared as a firm gray mass that had destroyed the temporal and occipital bones. The dura was intact with no invasion, and the lesion...
was easily peeled off. For the cervical part of the tumor, the major vessels were secured inferiorly, and the mass was removed up to the skull base. The tumor was removed entirely, except for a small portion around the jugular foramen to avoid lower cranial nerve injury. Finally, the large empty space was filled with a sternocleidomastoid muscle flap (Fig. 2). There were no neurological deficits after surgery.

Histopathological studies confirmed a WHO (World Health Organization) grade II atypical meningioma with up to 6 mitoses per 10 high-power fields (Fig. 3). The Ki-67 proliferation index was 15%. The results of immunohistochemical staining are provided in Additional file 1: Figure S1.

**Discussion and conclusions**

Primary intraosseous meningiomas are rare cranial lesions that arise from the bone, and they represent the most common type of primary extradural meningiomas [1–3]. The majority of intraosseous meningiomas are osteoblastic and cause hyperostosis, which may mimic fibrous dysplasia. In contrast, and more rarely, they may present as an osteolytic skull lesion [6, 7]. Reportedly, less than 20% of intraosseous meningiomas are osteolytic [8].

Primary extradural meningiomas are most commonly found in skull convexities, the paranasal sinus, and the middle ear but rarely in the skull base. Liu et al. reported 170 cases of extradural meningiomas in the head, and only 5.8% were located in the skull base [9]. Notably, there are few reports of osteolytic intraosseous meningiomas in the skull base. To date, 50 cases of osteolytic subtypes (including the present case) have been reported in the English literature (Table 1). Of these, only seven were located in the skull base, and all but two originated from the petrous bone.
The exact origin of extradural meningiomas is unclear, but several theories have been proposed. Their unusual locations are assumed to be the result of the aberrant differentiation or misplacement of undifferentiated mesenchymal stem cells [51]. Alternatively, extradural meningiomas may arise from differentiated arachnoid cap cells associated with blood vessels or nerves traversing the skull [52, 53]. Another theory proposes that they originate from arachnoid cap cells that get trapped in the cranial sutures during embryogenesis or molding of the cranium at birth [20, 21, 54]. Trauma with skull fracture has also been proposed as a causative factor of some extradural meningiomas, suggesting direct dural entrapment within bone fragments at the time of trauma [55].

The osteolytic subtype of intraosseous meningiomas is often misdiagnosed as a primary or secondary bone tumor due to its radiological appearance. The differential diagnosis of a solitary osteolytic skull lesion includes hemangioma, chondroma, chondrosarcoma, eosinophilic granuloma, epidermoid cyst, giant cell tumor, myeloma, and metastatic skull tumor [6, 23, 33].

Primary extradural meningiomas were practically classified according to their location by Lang and colleagues (Table 2) [3]. Therefore, intraosseous meningiomas could be considered Type II or Type III extradural meningiomas. Based on this classification, the present case falls into the type IIIIB category due to the presence of extracalvarial extension. This classification is helpful in predicting the risk of tumor recurrence. The IIC and IIIC subtypes rarely recur after complete resection, whereas the IIB and IIIB subtypes have a reported lifetime risk of recurrence of 26% [3].

Histopathological features are also important factors affecting tumor recurrence and prognosis. Recurrence was noted in 22% of benign intraosseous meningiomas in the literature, while it was reportedly 33% in cases of tumors with atypical or malignant pathology. In addition, aggressive atypical or malignant meningiomas had a significantly higher mortality of 29% compared to tumors with benign features (4.8%) [3]. Osteolytic meningiomas may have a higher incidence of atypical or malignant features [6]. In previous reports, benign features were reported in 87–89% of all extradural meningiomas, whereas in our literature review of osteolytic intraosseous meningiomas, 26% of cases were WHO grade II or III [3, 8].

Fig. 3 Histopathologic findings of atypical meningioma. The fragmented specimen (a) is seen as grayish-white solid masses. On microscopic examination, the tumor infiltrated the adjacent soft tissue (b, hematoxylin and eosin [H&E] stain, × 20, scale bar = 200 μm) and showed a whorled appearance and multifocal necrosis (c, H&E stain, × 100, scale bar = 100 μm). The tumor cells are composed of spindle cells with prominent nucleoli and ill-defined cytoplasm. Many mitoses are visible (d, H&E stain, × 400, scale bar = 20 μm).
Wide surgical excision is the main treatment for extradural meningiomas, and it is potentially curative if complete resection is achieved [6, 7]. In the present case, a small portion of the tumor near the jugular foramen could not be removed due to the possibility of cranial nerve injury. In the case of skull base lesions that cannot be totally resected, decompression of vital neural structures is performed.

In conclusion, we performed surgical treatment for a rare case of primary osteolytic intraosseous meningioma in the skull base with extension to the cervical area. The histopathologic diagnosis was atypical meningioma. If possible, complete resection is the treatment of choice, and serial follow-up should be done to confirm recurrence or progression.

### Table 1 Reports of primary intraosseous osteolytic meningiomas (Continued)

| Reference, year | Sex/age | Location | Type     | Pathology       |
|-----------------|---------|----------|----------|-----------------|
| Kim et al., 2012 [39] | M/68 | Parietal | IIC     | Atypical        |
| Kim et al., 2014 [43] | M/73 | Occipital | N/A     | Anaplastic      |
| Akhdaddar and Ennouali, 2014 [40] | F/74 | Frontal | IIC     | Papillary       |
| Tang et al., 2014 [41] | F/82 | Frontal | IIC     | Meningothelial   |
| Yun and Lee, 2014 [42] | F/65 | Frontal | IIC     | Atypical        |
| Kim et al., 2015 [45] | M/69 | Parietal | IIC     | Meningothelial   |
| Hong et al., 2015 [46] | M/61 | Frontoparietal | IIC | Benign          |
| Ben Nsir et al., 2016 [47] | M/42 | Petrous | IIIB    | Clear cell      |
| Bohara et al., 2016 [48] | M/38 | Parietal | IIC     | Atypical        |
| Mouri et al., 2017 [49] | F/76 | Frontal | IIC     | Transitional    |
| Richardson et al., 2017 [50] | M/23 | Frontal | IIC | Benign          |
| Present case | M/80 | Petrous | IIIB    | Atypical        |

### Table 2 Primary extradural meningioma classification by Lang et al. 2000 [3]

| Type | Description | Subtype |
|------|-------------|---------|
| I    | Purely extracalvarial with no bony attachment |  |
| II   | Purely calvarial | B (skull base) |
|      |             | C (convexity) |
| III  | Calvarial with extracalvarial extension | B (skull base) |
|      |             | C (convexity) |
Additional file

Additional file 1: Figure S1. Immunohistochemical staining results. The tumor showed a wild-type p53 pattern (A, × 200) and exhibited strong cytoplasmic expression of β-catenin (B, × 200). Some tumor cells exhibited weak membranous expression of EGF (C, × 200). The tumor was negative for Bcl-2 (D, × 200). The tumor shows membrane and cytoplasmic immunoreactivity for EMA (E, × 200) and negative for S-100 protein (F, × 200). Vimentin is diffusely expressed in the cytoplasm of tumor cells (G, × 200). The Ki-67 proliferation index is estimated to be approximately 15% (H, × 200). Scale bar = 100 μm. (DOCX 8214 kb)

Abbreviations
CT: Computed tomography; MRI: Magnetic resonance imaging; WHO: World Health Organization.

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Authors’ contributions
SMK collected data and drafted the manuscript. YK critically revised the manuscript and approved the final version. All authors revised the manuscript and approved the final version.

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Ethics approval and consent to participate
This study was approved by the institutional review board of Hanyang University Medical Center.

Consent for publication
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Competing interests
The authors declare that they have no competing interests.

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