A Case of Recurrence of Benign Convexity Primary Intraosseous Meningioma

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Abstract

Meningiomas originating within the bones of the skull are rare and have been reported as primary intraosseous meningiomas (PIOM). Moreover, PIOMs with a skull base location or malignant pathology are predisposed to recurrence; however, recurrence is quite rare among PIOMs characterized by a convexity location and benign pathology. Here, we present a case of extensive recurrence of a convex intraosseous meningioma with benign pathology. A 72-year-old woman presented with a headache to our hospital. Gd contrast-enhanced magnetic resonance imaging revealed an enhanced tumor in the left frontal to the parietal region extending through the calvarial bone and invading the subdural space and subcutaneous tissue. Skull radiograph and computed tomography identified a remarkable osteolytic change in the lesion. Macroscopic complete resection (MCR) of the tumor and the surrounding tissues was performed. The tumor was histopathologically diagnosed as a transitional meningioma (World Health Organization grade 1). Seven years after the surgery, the patient presented with dysarthria, and the recurrence of the tumor was identified as massive lesions extending through the calvarial bone to the orbital bone, partially protruding into the brain and scalp. MCR was performed again, with the reconstruction of the skull for an extensive calvarial area using a titanium plate. This case is unique due to the extensiveness of the recurrent tumor and its rarity. Here, we report the details of the clinical course and discuss the characteristics of this case.

Keywords: meningioma, intraosseous, recurrence

Introduction

Meningiomas are the most common among all primary intracranial tumors, comprising about 30% of all recorded cases.[1] Most meningiomas are located in the subdural space; however, those arising outside the intracranial compartment are rare and reported as primary extradural meningiomas (PEMs).[2] Primary intraosseous meningioma (PIOM) is a subtype of PEMs that arises in bones.[2,3] PEMs account for 1%-2% of all meningiomas,[3,4] whereas PIOMs account for two-thirds of all PEMs.[2] Regarding the incidence of PEMs, some reports exhibit two peaks in the age of predilection, with one peak between 50 and 70 years of age and another in the second decade of life. PEMs usually occur in both sexes with approximately the same frequency.[2,5] Moreover, any location in the skull can be affected by PIOMs, with the orbital cavity and frontoparietal site being the most common locations.[6,7] A painless scalp mass is the most common presentation in convexity intraosseous meningioma.[6,8] In skull base intraosseous meningioma, more neurological symptoms are expected because of cranial nerve defects.[6] Headache is reported to be the second most common symptom in both types of meningioma.[8]

Omofoye et al. reported a recurrence of PIOMs in 12.6% of cases, accounting for 14 of 111 cases in their systematic review.[8] The recurrence rate depends on multiple factors, including tumor grade, tumor location, and extent of surgical excision.[2,8] In particular, the skull base location is reported to demonstrate a 26% recurrence rate, whereas the convexity location displays 0% among pathologically benign tumors, according to the systemic review by Lang et
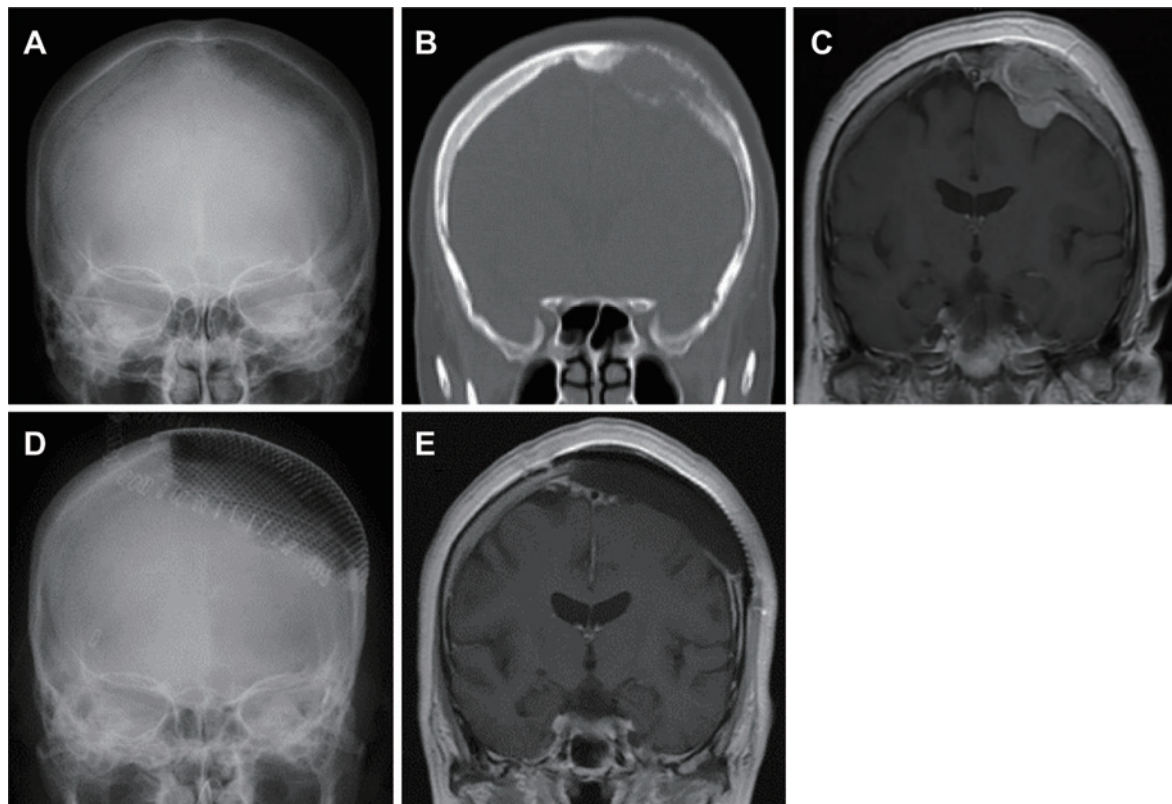
Fig. 1 Preoperative and postoperative radiographic findings. A, B: Preoperative skull radiograph and CT scan with a bone window showing an osteolytic change through the left frontal to the parietal bone. C: Preoperative coronal Gd contrast-enhanced MRI depicting a large, heterogeneously contrasted mass within the left parietal bone, with extensive invasion of the skull, intradural and intracranial epidural spaces, and subcutaneous tissue. D: Postoperative skull radiograph displaying cranioplasty with a custom-made titanium mesh after the lesion resection. E: Gd contrast-enhanced MRI demonstrating total resection of the lesion. CT; computed tomography, MRI; magnetic resonance imaging

al.[2] They also reported that recurrence rates in patients with atypical/malignant tumors were higher than that in patients with benign tumors (38% compared with 21%).[2] Therefore, the recurrence of PIOMs with convexity location and benign pathology is rare.

Here, we present a case of recurring convex PIOM pathologically diagnosed as transitional meningioma, World Health Organization (WHO) grade 1, affecting the patient extensively 7 years after complete resection. The recurrence pattern was aggressive regarding the extent of the cranial area involved, a unique feature of this case. Here, we describe the details of the clinical course and discuss the characteristics of this case.

Case Report

A 72-year-old woman with a history of rectal cancer surgery 13 years before presented to our hospital with a headache. The patient did not exhibit any neurological symptoms. Contrast-enhanced magnetic resonance imaging (MRI) revealed a massive tumor inside the calvarial bone, extending laterally and medially through the bone, expanding inward and outward from the bone, and partially invading the subdural space (Fig. 1). Skull radiography and computed tomography (CT) scan of the head displayed remarkable osteolytic changes of the lesion from the left frontal to the parietal bone, with the osteoid element covering the outer and inner surfaces of the lesion. Whole-body CT revealed no malignant lesions. Angiography displayed that feeding arteries to the tumor were developed from the middle meningeal artery (MMA) and superior temporal artery (STA). However, for the osteolytic lesion, the preoperative differential diagnosis included hemangioma, chondroma, chondrosarcoma, dermoid, epidermoid tumor, brown tumor, multiple myeloma, plasmacytoma, giant cell tumor, aneurysmal bone cyst, eosinophilic granuloma, and metastatic cancer.[6,7,9]

Preoperatively, transcatheter arterial embolization was performed on MMA. In the initial surgery, after the subcutaneous mass was exposed by meticulous dissection, the intraosseous mass was resected through craniotomy around the affected area. Additionally, the bone surrounding the craniotomy window was removed using rongeurs to secure a sufficient safety margin extending beyond the
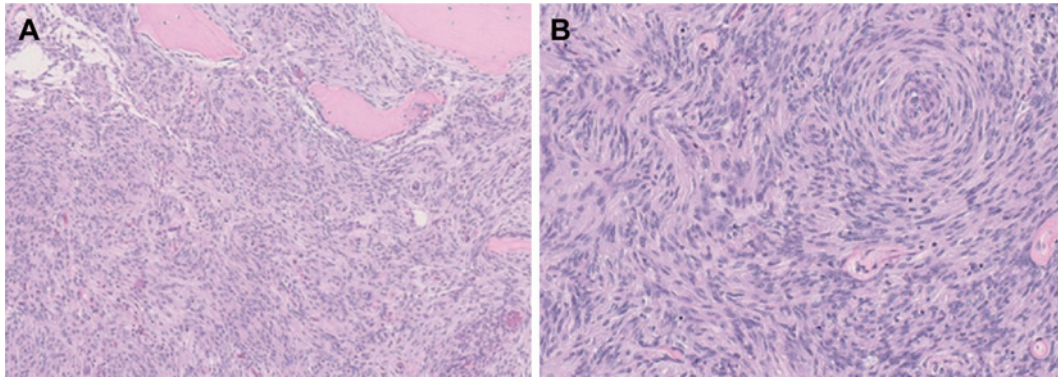
Fig. 2   **Pathologic findings. A: The tumor's bone invasion. (H&E original magnification ×100) B: Cellular whorl formation and thick collagen fibrosis without malignant changes displaying transitional meningioma, WHO grade 1 (H&E original magnification ×200).**
H&E; hematoxylin and eosin, WHO; World Health Organization

superior sagittal sinus (SSS). The subcutaneous tissue where the tumor was partially adhered was resected, and the non-adherent dura mater attached to the tumor was excised. Around SSS, the dura has not adhered, and no residual tumor was seen macroscopically. Therefore, the tumor and the surrounding tissues were completely removed macroscopically. The dural space was closed with prosthetic dura, and cranioplasty was performed using a custom-made titanium mesh plate (Fig. 1). Postoperative MRIs indicated complete tumor resection, although slight enhancement was seen around SSS (Fig. 1).

Histopathological examination of the excised specimen revealed tumor growth within the bone marrow of the diseased skull, in the subcutaneous tissue, and inside and outside the dura mater. Irregularly convoluted elongated cells with whorl formation and thick collagen fibrosis were observed in some areas, and transitional meningioma was diagnosed (WHO Health Organization grade 1). The Ki-67 proliferation index was 3%-5% (Fig. 2).

Periodic MRIs demonstrated no signs of recurrence, including the area around SSS, until two years later, when the follow-up of the patient ended. Seven years after surgery, she visited our hospital with a chief complaint of dysarthria. Neurologically, she exhibited mild weakness in the right upper and lower extremities. She had a huge bulge on the right parietal skin, which she noticed was growing gradually, without any concerning symptoms. Furthermore, CT and MRI of the head revealed a recurrent tumor over an extensive area. First, the lesion in the right frontoparietal area centered on the bone was massively protruding into the skin and extending to the middle part of the brain, where a massive invasion of the intracranial space was observed. Second, the lesion in the left calvarium with osteolytic changes was thickened, compressing the brain. Third, the lesion in the left orbital bone was enlarged into the intraorbital space to cause exophthalmos (Fig. 3). Angiography revealed developed feeding arteries

from MMA and STA as initial tumors.

During the second operation after TAE, the protruding mass lesion on the right parietal scalp was exposed subcutaneously by meticulous dissection and removed until the bone edge was observed. After removing the titanium plate used in the previous surgery, the tumor on the left aspect of the brain was removed as it continuously extended into the left orbital bone. As the tumor invaded the diploic space beyond the margin of the resected bone, additional bone resection was performed until normal bone structure was observed. Most of the tumors were located outside the dura mater; however, in the median region, the tumor had extended into the interhemispheric fissure through the dural defect in the previous surgery, where adhesions to the brain were not strong. The dura mater in the operative field did not adhere to the tumor and could be separated as a whole; a thinned inner plate of the cranial bone was observed partly on the dura mater. Thus, the complete removal was accomplished, including duraplasty using an artificial substitute, filling the dead space of the bone defect with fat from the abdomen. Moreover, cranioplasty with a new custom-made titanium mesh was performed.

Histopathological examination revealed a dense proliferation of elongated cells with irregularly convoluted bundle patterns and whorl formations, scattered psammoma bodies, and a transitional meningioma was diagnosed, as in the previous specimen. The Ki-67 proliferation index was 5%-10% at the highest site, and there was some size disparity in the nuclei of the tumor cells; however, no noticeable mitotic figures were observed, confirming that the tumor was WHO grade 1 (Fig. 4). Postoperatively, the patient's symptoms improved, and she displayed no evidence of recurrence 1 year after the second surgery.

Discussion

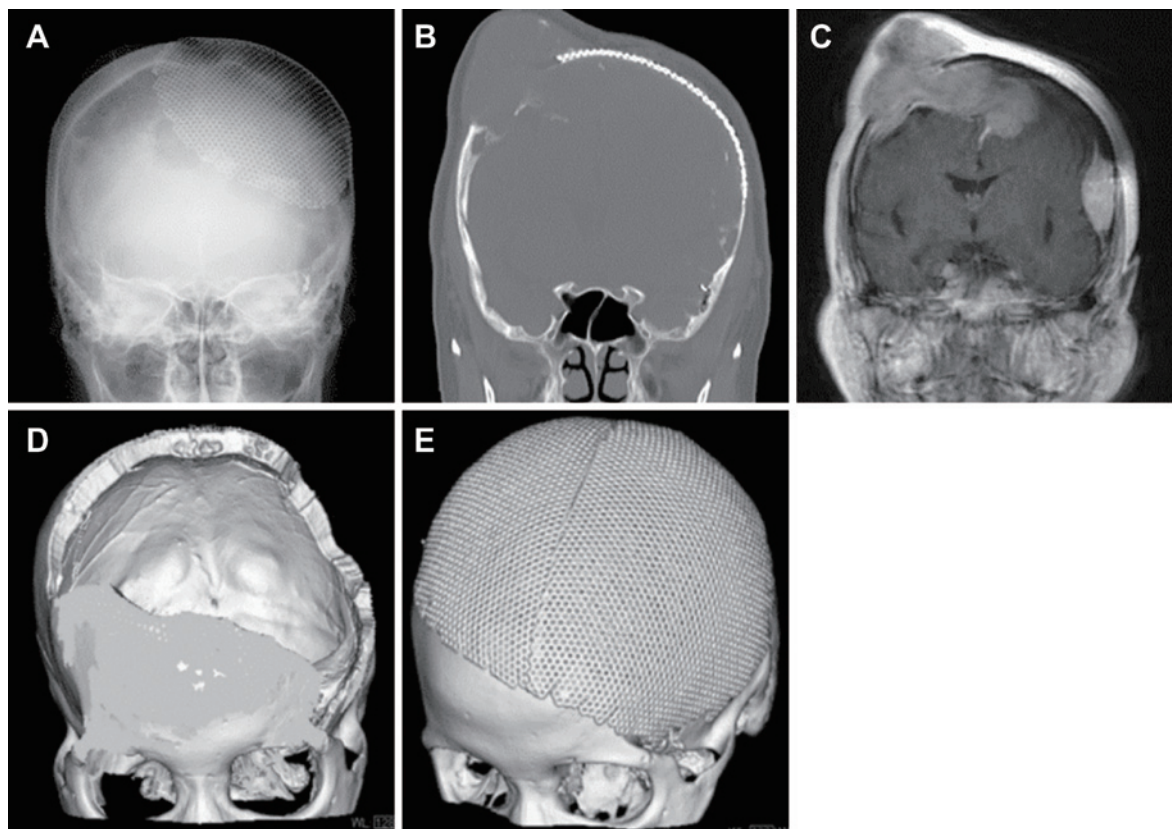Meningiomas, typically located intracranially, are known

Fig. 3 Radiographic findings at recurrence. A–C: Preoperative skull radiograph and CT scan displaying osteolytic lesions bilaterally at the site of the bone resection. Gd contrast-enhanced MRI displaying a large, heterogeneously contrasted mass with the extensive invasion of the craniotomy site, intradural and intracranial epidural spaces, and subcutaneous tissue. D, E: Postoperative CT scan exhibiting extensive removal of cranial bone and cranioplasty by titanium plate.
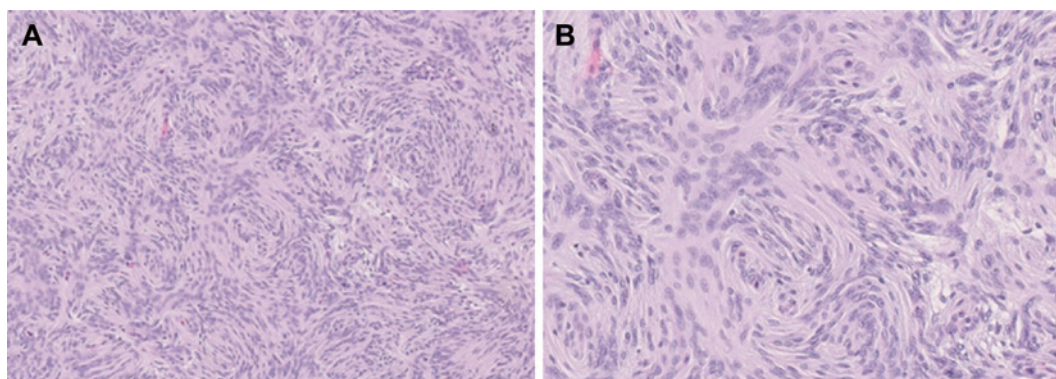**CT; computed tomography, MRI; magnetic resonance imaging**

Fig. 4 Pathologic findings of the tumor specimens at the second surgery. A, B: Histopathological specimen displaying transitional meningioma, with some size disparity in the nuclei, no noticeable mitotic figures or necrosis, confirming the tumor as WHO grade 1 (H&E original magnification A: ×100, B: ×200).
H&E; hematoxylin and eosin, WHO; World Health Organization

to occur in the calvaria, scalp, orbit, paranasal sinuses, skin, neck, mediastinum, and lungs. As meningiomas have a non-dural origin, they are referred to as PEMs.[2] In 1932, "Primary intraosseous meningioma" was a term reported for tumors originating in bones and could be categorized as a subset of PEMs.[3,10] Theoretically, PEMs do not have any connection to the dura mater or any intracranial structures, and a spared dura would be a reliable sign of

ectopic origin.[2,8)] However, PEMs can spread to the dura mater and invade the intracranial space.[11)] Additionally, PEMs can be confused with typical convexity meningioma extending outside the dura. The dural tail sign, typical in convexity meningiomas, can also be present in PEMs and is not a determinant of dural origin.[2)] In such cases, the presumed site of origin can be assessed using the center of the tumor expansion and the direction of bone remodeling.[12,13)] In our case, the initial tumor invaded the subdural space and had a dural tail-like sign, suggesting the possibility of convexity meningioma. However, radiological findings show that the initial tumor was centered in the skull, with the inner and outer plates of the bone partially visible, suggesting that the tumor originated in the bone. Among PIOMs, another rare category representing tumors originating within the diploic space is called 'primary intradiploic meningioma' and is reported to be localized in the frontoparietal and orbital regions.[14)] Considering the tumor location, recurrence pattern, and invasion through the calvarial bone, our case might fit in primary 'intradiploic meningioma.' Moreover, PIOMs can be subclassified into osteoblastic (hyperdense changes), osteolytic (bone destructive changes), and mixed types based on their appearance in radiographs or CT scans.[15)] On a CT scan, the osteoblastic type demonstrates focal hyperdense lesions with prominent expansion of the skull, whereas thinning of the bone is predominant with similar hyperdense lesions in the osteolytic type.[6)] The osteoblastic subtype is reported to be common in PIOMs, with osteolytic appearance rates ranging from rare to 35%.[7,10,16)] Moreover, PIOMs can also be subclassified by their location into convexity and skull base lesions.[2)] Clearly, the tumor in our case is among the subgroups of osteolytic type and convexity location.

The pathogenesis of PIOM is still unexplained and controversial. Several authors propose that the origin of intraosseous meningioma is related to the intracranial suture line, where a portion of the dura mater or arachnoid cap cells is trapped.[9,17)] Moreover, they assume that the entrapment of the portion of the dura mater or arachnoid cells into the suture line could result from trauma, embryonic development, or head molding at birth.[18-20)]

Lang reported that only 14 (8%) of the 168 cases of PEM were associated with cranial suture.[2)] Meanwhile, Crawford reported that only 14% had a history of trauma.[7)] Referring to our case, which was located around the coronal suture line with no particular history of trauma, tumorigenesis remained unclear. Shangshoti proposed a mechanism of intracranial remnants of meningocytes differentiated by multipotent mesenchymal tissues,[21)] which might be a multifactorial causality, as suggested by Lang.[2)]

Recurrence in our case was unanticipated, considering that the supposedly benign tumor and the surrounding tissues achieved a successful MCR. In particular, convexity location coupled with benign pathology seems to be the most favorable for treatment.[2)] However, other possible factors should also be considered for the recurrence. First, the osteolytic type may be a significant indicator of recurrence. Osteolytic intraosseous meningiomas are known to be biologically active, especially in cases of rapid proliferation, and they are more likely to present with anaplastic or malignant histopathology.[6,10)] Therefore, patients with osteolytic appearance in PIOMs need a prompt histologic diagnosis due to a higher likelihood of malignancy than osteoblastic intraosseous meningiomas.[3)] However, Chen reported that 11% of histologically unresectable benign intraosseous meningiomas might display evidence of malignant changes, so the risk of recurrence must be considered even if the histopathology was benign.[3)] In addition, the importance of the genetic profile of the tumors, such as mutation, promoter methylation, or loss of chromosomes, has been reported to predict recurrence and prognosis.[22,23)] Although we did not have a chance to perform genetic analysis, such investigation could provide more predictable information for PIOMs.

Second, invasion into surrounding tissues, such as the dura mater or soft tissue, could significantly impact the recurrence, and malignant features may be indicated by microscopic tumor invasion of underlying dura or overlying soft tissue structures.[24)] Additionally, scalp swelling and extracranial soft tissue mass indicate malignant meningiomas.[4)] The rate of dural infiltration of PIOMs is reported as 26%-87.5% histopathologically and 60.0%-68.8% radiographically,[24,25)] suggesting that PIOMs are prone to infiltrate the dura. In particular, the tumor in our case was situated near SSS, and a slight enhancement was observed postoperatively, indicating a possible dural infiltration around SSS. We assumed that the dura around SSS was intact based on the observation that SSS had not adhered to the tumor intraoperatively, and slight enhancement did not change until two years after surgery. However, since special care should always be taken for meningiomas around SSS, we should highlight that close monitoring should have been forced for a long period for our patient. Third, the presence of residuals or tumor seeding in the diploic space of the bone after resection may also be a source of recurrence. Chen reported that even if PIOMs are histologically benign, they may encroach on or involve certain parts of the skull that might hinder the complete resection of the lesion.[8)] Given our case's marked osteolytic features and possible dural infiltration, it can be established that the tumor could recur. In particular, the recurrence pattern through an extremely extensive area suggested that the tumor had a strong tendency to disseminate through the diploic space, which we speculate was also responsible for the recurrence.

As an operative strategy for PIOMs, maximizing the extent of bone resection is the first key step for the aforementioned reasons. Since confirming the boundary for the bony tumors is impossible, macroscopically or pathologically, a navigation system with meticulous evaluation using

MRI will help determine the cutting line of the affected cranial bone. Further removal through cutting and shaving away the bone along the margin could help eradicate residuals within the diploic space. Extensive bone defects of the skull can be tolerated, as our case demonstrated, and custom-made three-dimensional cranial prostheses are aesthetically and functionally favorable with few complications.[26,27] Therefore, expanding the extent of bone removal should be done without hesitation, and it should be considered that the titanium plate would be the most suitable for calvarium surgery and should be designed for a larger area than the expected resection site so that the plate would be applicable even when the extent of resection is increased. Furthermore, thorough resection of contiguous tissues, such as the dura mater or subcutaneous tissues, is critical. In particular, total resection of the dura at the craniotomy site should be considered for every case, as PEMs and PIOMs could recur even if intraoperative findings indicate no dural invasion.[24] Adjuvant therapy, including radiotherapy, chemotherapy, or bisphosphonate therapy, can be the treatment option if the tumor recurs or the histopathology exhibits atypical or anaplastic meningioma.[6,28] Finally, we must highlight that the patient did not visit our hospital until she exhibited dysarthria despite being aware of the growing scalp mass. The recurrence led to the involvement of an extensive area. Therefore, it should be emphasized that careful long-term follow-up is essential after surgery for intraosseous meningiomas.

Conclusion

We encountered a rare intraosseous meningioma displaying osteolytic changes and recurrence involving an extensive area. Intraosseous meningiomas, especially those with osteolytic changes or invasion of the surrounding tissues, such as the dura mater or bone, are at a high risk of recurrence and should be excised with a sufficient margin. Regardless of the pathological findings or degree of gross removal, careful long-term imaging follow-up is required.

Informed Consent

Informed consent has been obtained from the patient in this case report.

Conflicts of Interest Disclosure

All authors declare no conflict of interest.

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