

Intraosseous Meningioma: Case Report

İntraosseöz Meningiyom

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ABSTRACT Meningiomas usually arise from arachnoid cap cells, however these tumors may also develop from extracranial areas such as the skull; they have been termed intraosseous meningiomas. We reported a 57-year-old female presenting with a scalp swelling of the left parietooccipital region. The patient was operated and polymethylmetacrylate (PMMA) was used to restore the bony defect after total tumor removal was achieved. The patient has been well with no evidence of recurrence one year after surgery. The pathological examination revealed intraosseous meningioma. In symptomatic primary intraosseous meningioma, we suggest total tumor removal with a wide surgical resection followed by cranial reconstruction because of its potential for malignant transformation.

Key Words: Meningioma; skull

ÖZET Meningiyomlar, genellikle araknoid "cap" (kasket) hücrelerinden kaynaklansalar da, kafatası gibi ekstrakraniyal alanlarda da görülebilirler. Bu tür meningiyomlara intraosseöz meningiyom denilmektedir. Biz de 57 yaşında bir kadında intraosseöz meningiyom olgusu sunduk. Sol parieto-oksipital bölgedeki kemikte yumuşama şikâyeti ile başvuran hasta opere edildi ve lezyon total olarak çıkartıldı; defekt polimetimetakrilat (PMMA) ile kapatıldı. Postoperatif dönemde herhangi bir şikâyeti olmayan hastanın patolojisi intraosseöz meningiyom olarak bildirildi. Bir yıllık izlemde nüks saptanmadı. Semptomatik intraosseöz meningiyomlarda, bu tür lezyonların malign transformasyon potansiyelleri nedeni ile geniş cerrahi rezeksiyonu takiben kraniyal rekonstrüksiyon yapılmasını öneriyoruz.

Anahtar Kelimeler: Meningiyom; kalvaryal

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Meningiomas usually arise from arachnoid cap cells which are located on the outer layer of the arachnoid membrane.¹ However these tumors may also arise from other areas such as the skull, and have been termed intraosseous, calvarial, diploic, or epidural meningiomas.²⁻⁵ Primary extradural meningiomas are rare lesions, accounting for less than 2% of all meningiomas. Intraosseous meningiomas are increasingly rare, occurring in 14% of cases of primary extradural meningiomas.⁶ Yamazaki et al reported 48 published cases up to 2001.⁷ On the other hand, osteoblastic or mixed osteoblastic-osteolytic lesions compose most intraosseous meningiomas with pure lytic lesions the least common. In 2004, Rosahl et al reported 16 cases of primary intraosseous osteolytic meningiomas witho-

ut evidence of soft-tissue invasion.⁸ Our case is the 17th osteoclastic case of the literature.

A 57-year-old female presented with a scalp swelling of the left parietooccipital region.

Computed tomography (CT) and magnetic resonance (MR) imaging of the patient showed an osteolytic intracalvarial lesion. Following wide surgical resection, the histological examination revealed an intraosseous fibrous meningioma. In this study, the clinical and radiological findings of primary osteolytic intraosseous meningioma were discussed and the relevant literature was reviewed.

CASE REPORT

A 57-year-old female presented with a scalp swelling of the left parietooccipital region, which has been present for the last 20 years and has gradually increased in size. The swelling has become painful for the last 1-2 years, and there was no history of trauma. Physical examination revealed an approximately 4 cm swelling of the left parietooccipital region, which was not adherent to the overlying skin.

The patient had no neurological deficit. The laboratory studies were unremarkable. Radiographs of the skull revealed a well-defined area of osteolysis in the left parietooccipital region (Figure 1A). CT revealed a pure intradiploic (intraosseous) mass expanding the calvaria and making remarkable bone destruction (Figure 1B, 1C). MR imaging showed a calvarial lesion, which was hypointense on T1-weighted and hyperintense on T2-weighted images. The T1-weighted imaging following gadolinium injection demonstrated the pro-

minent and homogeneous enhancement of the lesion. The patient was operated and PMMA was used to restore the bony defect after total tumor removal was achieved. Intraoperative assessment revealed that the tumor did not adhere to the scalp and had caused destruction of both the inner and outer tables of the skull. The tumor mainly occupied the intradiploic space and there was no evidence of dural invasion and no extension to the extracalvarial area. The tumor and the surrounding bone were removed, followed by cranioplasty reconstruction. Dural involvement was absent. Histological diagnosis was fibrous meningioma. The patient has been well after the operation with no evidence of recurrence one year after surgery (Figure 2A, B and C).

DISCUSSION

Meningiomas developing in locations outside the dural compartment have been called ectopic, extradural (epidural), calvarial, cutaneous, extracranial, extra neuraxial, or intraosseous meningiomas.⁹ Most intraosseous meningiomas are associated with normal or thickened underlying dura mater.^{2,10} Ectopic meningiomas arise from arachnoidal cell rests that persist in non-dural locations.⁷ Ectopic meningiomas in extradural location account for only 0.8-0.9% of all meningiomas and have occurred in many locations in the head and neck including the subcutaneous tissues of skin, orbit, paranasal sinuses, intraosseous, salivary glands, and along the perineural sheaths of cranial nerves.^{11,12,13} To avoid the confusion in nomenclature, Lang et al proposed a single term, "primary extradural meningioma" for

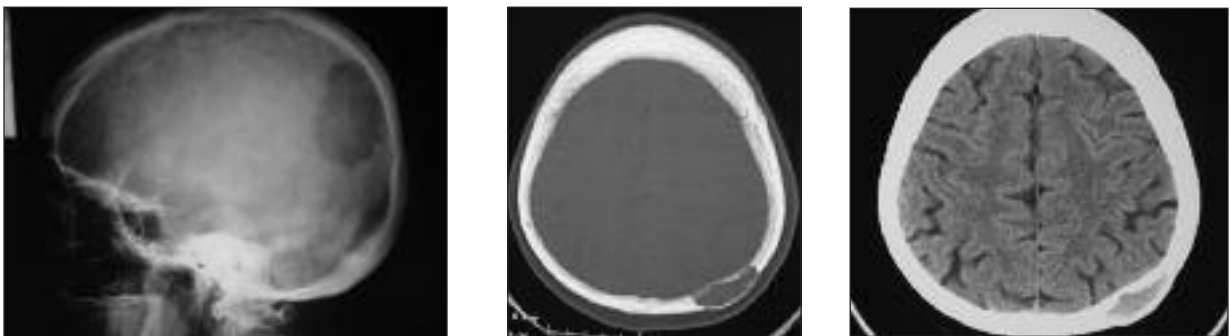


FIGURE 1: Lytic lesion is shown in plane graphic (A), CT image of the same lesion on left parietal region, in bone window (B) and in paraneural window (C), preoperatively.

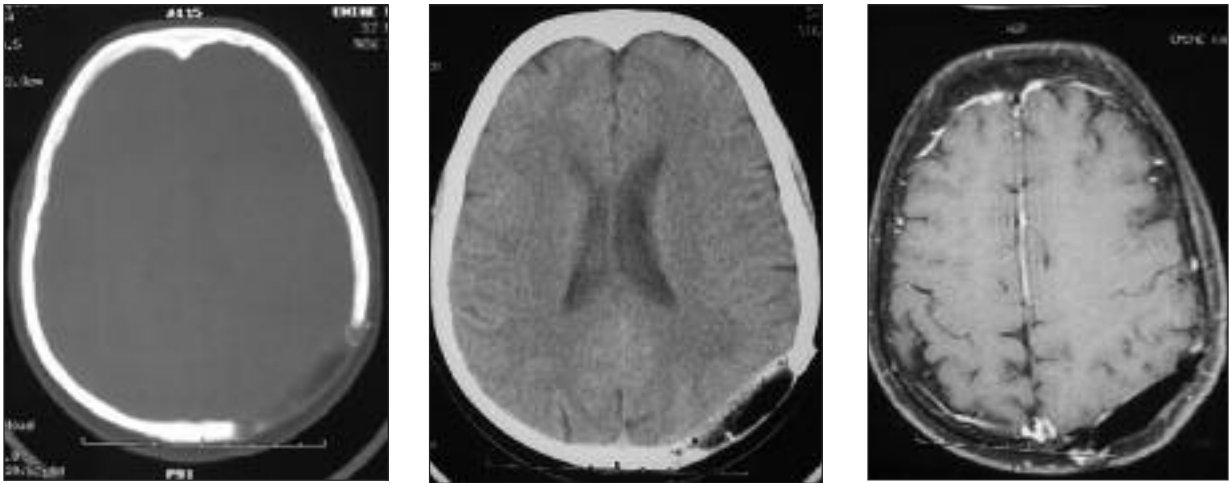


FIGURE 2: Postoperative CT images are shown in both bone window (A) and in paranchyme window (B), and control MRI of the patient one year after surgery (C)

such lesions.¹⁴ Primary extradural meningiomas are classified as purely extracalvarial (type I), purely calvarial (type II), or calvarial with extracalvarial extension (type III). Type II and type III tumors were further categorized as convexity (C) or skull base (B) lesions. True primary intraosseous meningioma is defined as a lesion that does not involve the underlying dura.⁴ However, intraosseous meningiomas can be classified as primary and secondary, and the dura may become involved later in the course of the tumor growth even in primary intraosseous meningioma. Secondary intraosseous meningioma is due to the extension of an intracranial meningioma into the calvarium.²

The pathogenesis of the so-called primary “intraosseous” meningiomas remains obscure. Many different hypotheses exist regarding the origin of primary calvarial meningiomas. In general, ectopic meningiomas of the calvarium are believed to derive from arachnoid cells that have been incorporated into the skull during development. Another hypothesis is that they arise from ectopic meningiocytes or arachnoid cap cells trapped in the cranial sutures during molding of the head at birth and subsequently develop into a meningioma since such meningiomas usually occur along the skull sutures.² Trauma is also thought to be responsible for some primary intraosseous meningiomas; arachnoid cap cells caught in the fracture line during head trauma may be responsible for intraosseous meningiomas.¹⁵ Misplace-

ment and entrapment of meningotheial cells into suture or fracture lines as a result of trauma may be the possible mechanism of calvarial meningiomas.¹⁵ Mesenchymal precursors have multipotential ability to differentiate into various tissues and meninges are mesenchymal in origin; meningiomas may develop from multipotential mesenchymal cell precursors.¹⁶ The latter hypothesis is supported by the occasional observation that these tissue types develop via metaplasia within meningiomas.⁷

Meningiomas occurring outside the cranium can be classified in four groups: 1- Direct extension from a primary intracranial meningioma via the foramina of the skull base; 2- extracranial growth of arachnoid cells into the sheaths of cranial nerves; 3- extracranial growth of embryonic arachnoid remnants with no evident connection to the foramina of the skull base or cranial nerves; and 4- distant metastases from intracranial meningiomas.¹⁷ Meningiomas originating in the subcutaneous, intraosseous, or paranasal sinuses are classified in the third group, as in our case.

According to the literature, 68% of the primary extradural meningiomas involved the calvaria.¹⁴ Frontoparietal and orbital regions are the most common locations. Only 35.5% of reported cases were associated with trauma; on the other hand, 70.8% of cases had an association with any of the sutures, especially with coronal suture. In histological examination, 62.5% of all cases were

meningotheliamatous menengioma.⁷ In our case, there was no definite history of trauma or radiographic manifestation of an old skull fracture.

Unlike primary intradural meningiomas, primary extradural meningiomas occur at approximately the same frequency for each sex.^{3,14} Both primary intradural meningiomas and primary extradural meningiomas occur predominantly during the 5th or 6th decades of life, but primary extradural meningiomas have a second peak incidence in younger patients during the second decade.¹⁴ Most of these lesions may be detected incidentally.¹⁸ In general, calvarial meningiomas are benign lesions and have slow growing patterns. On the other hand, calvarial meningiomas are more prone to progress into malignant changes (11%) compared with intracranial meningiomas (2%).^{3,14} Meningiomas presenting with scalp swelling, osteolytic skull lesions, and extracranial soft-tissue masses are thought to be more aggressive than others.¹⁹

In our case, the tumor purely occupied the intradiploic space and there was no evidence of dural invasion and no extension to the extra calvarial area; thus, it was classified as primary extradural menengioma type IIC.

CONCLUSION

Osteolytic lesions of the skull may have several causes such as metastatic malignant tumors, histiocytosis X, epidermoid, hemangioma, myeloma, plasmacytoma, giant cell tumor, hemangioma, epidermoid cyst, osteogenic sarcoma, eosinophilic granuloma, aneurysmal bone cyst, or fibrous dysplasia etc. We suggest that osteolytic lesions should be extirpated as soon as possible, before the tumor invades the subdural space.

In symptomatic primary intraosseous meningioma, we propose total tumor removal with a wide surgical resection followed by cranial reconstruction because of its potential for malignant transformation. If only subtotal resection is possible due to the anatomic location of the tumor, the residual tumor should be monitored radiologically. In patients whose residual lesions are symptomatic and/or show evidence of progression, adjuvant radiation therapy is recommended.¹¹

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