INTRODUCTION

Meningiomas are the most common benign intracranial neoplasm commonly arising from arachnoid cap cells located in the external layer of the arachnoid membrane. Almost 1% to 2% of meningiomas are lesions described as extradural meningiomas without association with the surface of the arachnoid membrane. Primary intraosseous meningiomas are rare (1,2). Frontoparietal and orbital regions are the most common sites for intraosseous meningiomas (2). Here, we report a case of primary intraosseous meningioma arising in the orbit.

CASE REPORT

A 46-year-old woman presented with left exophthalmos. She first noticed it approximately two years ago. She had experienced occasional blurred vision without diplopia or pain. Neurological examination was normal. She had no history of antecedent trauma. Laboratory studies were unremarkable. Orbital magnetic resonance imaging (MRI) revealed an expansile bony lesion in the orbital lateral wall of the left sphenoid bone. The patient underwent craniotomy for excision of the bony lesion. Pathologic examination revealed an intraosseous meningioma.

Index terms
Intraosseous Meningioma Diplopia Orbit

Primary Intraosseous Meningioma in the Orbital Bony Wall: A Case Report and Review of the Literature Review

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Received June 6, 2014; Accepted November 10, 2014
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Primary intraosseous meningiomas are rare form of meningioma. However, they account for approximately 67% of all extradural menigiomas (2). Frontoparietal and orbital regions are the most common sites for intraosseous meningioma. In our case, this tumor was detected at the orbital lateral wall of the left sphenoid bone.

The etiology of intraosseous meningioma has not yet been clarified. Many different hypotheses exist for the origin of primary intraosseous meningioma. This tumor has been thought to arise from ectopic arachnoid cap cells trapped in cranial su-

DISCUSSION

Most meningiomas are considered primary intradural lesions located in the subdural space. In contrast, extradural meningiomas arise from the neck, skin, and nasopharynx. Extracranial meningiomas are rare, accounting for 1–2% of all meningiomas.
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tures during molding of the head at birth or from entrapped dura and arachnoid during a trauma with dural tear (3). Although this tumor was closely related to sphenozygomatic suture, only 8% of such tumors are found in the proximity of a cranial suture (3).

Lang et al. (4) have suggested to classify primary extradural meningiomas to type I (purely extracalvarial), type II (purely calvarial), and type III (calvarial lesions extending beyond the calvaria). Each type is further divided into subgroup of B (skull base) and C (convexity). In our case, the tumor was sphenoidal bony lesion extending beyond the calvaria. Therefore, it could be classified as type III B tumor.

Radiologically, primary intraosseous meningiomas can be classified to osteoblastic, osteolytic, or mixed type. Primary intraosseous meningiomas are mostly osteoblastic type (5). Conventional radiographs can detect abnormalities in 30% to 60% of patients with intraosseous meningiomas, including hyperostosis, atypical vascular marking, and irregular foci of calcification. However, conventional radiographs are usually limited in the diagnosis of extracranial meningiomas due to their superimposed bony structures. CT with bone window showing hyperdense lesions settled in the focal thickened bone is necessary for their detection. After contrast administration, a densely enhanced dural lesion can be found adjacent to the bony lesion. Less frequently, primary intraosseous meningiomas may present as the osteolytic type (6). The lesions appear hypodense on conventional radiographs. The lesions are hyperdense compared to brain tissue on non-enhanced CT scan with homogeneous enhancement after contrast administration. MR images allow better anatomic delineation in the evaluation of soft-tissue component and extradural extension of the lesion. On MR images, the findings of both osteoblastic and osteolytic subtypes of intraosseous meningiomas are similar to each other. Typically, these lesions are hypointense on T1-weighted images and hyperintense on T2-weighted images. After gadolinium administration, homogeneous enhancement can be found. In our case, T2-weighted MR images showed hypointensity accompanying the dural lesion. Thickened and enhanced dura adjacent to neoplasm may represent either reactive inflammation or tumoral invasion. Correctly recognizing dural invasion from reactive change has important surgical implications. Previous study has shown that the presence of pial enhancement, focal dural nodules, or dural thickening of more than 5 mm is highly accurate in predicting the presence of neoplastic dural invasion (7). Because dural thickening in our patient was more than 5 mm, it was more likely a tumor invasion than a reactive dural thickening.

Extradural meningiomas are indistinguishable from intradural meningioma histologically or immunophenotypically. Histopathologic examination often reveals findings pathognomonic of intradural meningiomas, including uniform spindle-shaped cells arranged in whorls, and interconnecting fascicles. Nuclei are oval and regular. Nuclear pseudoinclusions can be found. Almost all meningiomas should react positively with EMA (stains cytoplasmic membranes) and vimentin (stains cytoplasm) (8).

Differential diagnoses from the osteoblastic type of primary intraosseous meningioma include fibrous dysplasia, meningioma en plaque, osteoma, osteosarcoma, and Paget disease (8). The osteoblastic type of primary intraosseous meningioma ap-

Fig. 3. Histopathological feature of intraosseous hemangioma.
A. Microscopic examination reveals a set of neoplastic cell lobules between bony trabecula (hematoxylin & eosin stain, × 100).
B. Within a high-power field, the tumor cells have indistinct cell borders and show a whorled appearance (hematoxylin & eosin stain, × 400).
C. Immunohistochemical staining for vimentin shows strong cytoplasmic reactivity (x 400).
pears as a focal hyperdense skull lesion on CT. Fibrous dysplasia is a developmental disease. It commonly occurs at young age. It stops growing after bone maturation, whereas intraosseous meningioma appears after puberty and continues to grow slowly. In addition, fibrous dysplasia does not have tumor blush on angiography that is typical of meningiomas. Fibrous dysplasia expands the medullary space and widens the bone. An intact rim of cortical bone is often seen over the margins of the involved bone on CT. MR images usually show low to intermediate signals on all imaging sequences with variable enhancement on T1-weighted contrast-enhancing images (9). Malignant transformation is very rare (in less than 1% of cases). Osteomas are the most common benign tumor in calvaria. On CT, osteomas are dense, smooth, and well-demarcated lesions. However, osteomas show void signal without soft-tissue components on MR images. Osteosarcomas usually show poorly defined margins and heterogeneous signals. The intramedullary components of tumors are often associated with the zone of cortical destruction. Imaging features of osteosarcomas are more aggressive. Their prognoses are worse than those of meningiomas. Paget disease of bone shows variable imaging patterns that are related to pathologic stages. Typically, MR images of Paget disease involving calvarium are heterogeneous and hypointense on T1-weighted images due to marrow replacement by fibrous tissue and the presence of nonehancing lesions (10). Serum alkaline phosphatase is typically elevated in patients with Paget disease (2).

Primary intraosseous meningiomas have been observed to be benign and slowly growing. Recent studies indicate that intraosseous meningiomas have a tendency to be more malignant than their intradural counterparts (9). These malignant features may be indicated by microscopic tumor invasion of underlying dura or overlying soft tissue structures. In addition, the potential to transform into malignant is higher in the osteolytic type than in the osteoblastic type (2).

Total tumor removal with wide surgical resection followed by cranial reconstruction is the treatment of choice. When total resection is not feasible because of the involvement of critical structures within the orbit, paranasal sinuses, or skull base, residual tumors should be radiologically followed. Adjuvant therapy must be indicated in patients with malignancy as well as non-resectable tumors. Adjuvant therapies include gamma knife, chemotherapy, and disphosphonate therapy (2, 4).

In conclusion, primary intraosseous meningiomas are rare. They are mostly benign. However, intraosseous meningiomas are more likely to be malignant than their intradural counterparts. Therefore, preoperative radiologic evaluation of the size, location, characteristics, and type of tumors is important. In particular, MR imaging may be valuable in the diagnosis and evaluation of soft tissue component and extradural extension.

REFERENCES

안와에 발생한 일차성골내수막종: 증례 보고 및 문헌고찰

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수막종 중에서 두개골 밖에서 기원하는 종양을 경질외수막종으로 부르고 있다. 경질외수막종은 모든 수막종의 약 2% 보다 적게 발생하는 드문 질환이다. 일차성골내수막종은 바로부터 기원하는 경질외수막종을 부르는 용어이다. 본 증례는 46세 여자 환자가 왼쪽 안구돌출증을 주소로 내원하였다. 시행한 CT와 MR 영상에서 왼족 나비뼈의 바깥쪽 복에 평창성의 병변이 발견되었다. 머리뼈절개 수술을 시행하였고, 병리학적 검사상 골내수막종으로 확인되었다.

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