Primary Intraosseous Meningioma

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Meningioma is usually known to occur stuck to the dura mater, but extradural meningioma occurs rarely. Most of the extradural meningiomas are located in the head and neck and we report a case of the primary intraosseous meningioma in the orbit. A 50-year-old woman presented with the left eye hyperemia and exophthalmos. Neuroimaging modalities showed hyperostosis at the left sphenoid and orbital wall. On microscopic view, spindle cells and psammoma bodies between the woven bones were observed. The origin of the intraosseous meningioma is explained with various potential possibilities and differential diagnosis thoroughly explored.

KEY WORDS: Meningioma · Extradural · Primary intraosseous.

Introduction

Meningioma constitutes 15~20% of primary brain tumors. It is a tumor originating from the arachnoid cap cells and mostly dura-based slow growing one. Rarely extradural meningiomas have been reported in less than 2% of the meningiomas located in the scalp, neck, paranasal sinus, skin, lung, and even finger and so on. Here we introduce one case of the intraosseous meningioma at the orbit and discuss more about that.

Case Report

A 50-year-old female had presented with severe left eye hyperemia and tears for 6 months. Recently left exophthalmos developed. On ophthalmologic & neurologic examination, there was no deficit except the left eye injection and exophthalmos. Orbit computed tomography (CT) revealed that a hyperostotic lesion was located at the left sphenoid bone, part of orbital roof and lateral orbital wall. Fine radiating spicules looked bursting perpendicularly to the bone surface. Brain magnetic resonance image (MRI) revealed hypo-intensity on T1 and T2 weighted images. Gadolinium enhanced MRI showed enhancement of the adjacent soft tissue and dura, and no enhancement of the main bone lesion (Fig. 1). On Technesium-99m methylene diphosphonate (Te-99m MDP) scan, the uptake at the lesion increased (Fig. 2).

The operation was done with navigation system. Left periorbital scalp incision was followed by the fronto-temporal craniotomy, after which sphenoid bone, part of orbital roof and orbit lateral wall was drilled out. Optic nerve was decompressed. The main mass consisted of rather fragile abnormal bone, resembling that of fibrous dysplasia. The fronto-temporal bone flap was repositioned using fixation devices.

On microscopic views, oval shaped and spindle shaped cells formed lobules between the woven bones. Nuclei frequently had central clearing and psammoma bodies/whorls were viewed. Cytoplasm and surrounding collagens were positive in immunohistochemical studies of epithelial membrane antigen (EMA), vimentin, and Ki-67 was negative (Fig. 3).

After operation, exophthalmos and left eye hyperemia disappeared. On follow-up MRI and CT, compressed orbital cavity was fully decompressed but small amount of mass remained (Fig. 4).

Discussion

The extradural meningiomas are uncommon. Many locations such as calvaria, scalp, orbit, paranasal sinus, neck, skin, lung, mediastinum, finger, et al were reported and more than 90% of extradural meningiomas were located in the head and neck7. As the nomenclature has yet to be definitely defined, we have named the extradural meningioma at the skull as the primary intraosseous meningioma (PIM). The definition of PIM versus intradural meningioma depends on the dura invasion. Hoye, et al defined the PIM as no involvement of the dura5, whereas Oka and colleagues reported that some calvarial meningiomas infiltrated dura, but it was not primarily intradural.
origin because the mass displaced the dura away from the inner table. Lang, et al explained that as the calvarial mass grew and eroded the inner table of the skull, there would be sure to be reaction with the dura. So dural invasion was not exclusive criteria and main mass location, growth direction and dural displacement pattern were of importance. In our case, intraoperative dural specimen was not biopsied but main mass was located in the bone and growth direction was outward expanding pattern without definite dural thickening and intradural mass.

Lang, et al reviewed the 142 patients' literatures and concluded the differences between extradural meningioma(EM) and intradural meningioma(IM). Female and male incidence ratio is 2:1 in the IM, 1:1 in the EM. IM occurs during the later decades, but EM has two peaks during early and late decades. EM is more prone to be malignant than IM; 11% in the EM, 2% in the IM. It is well known that hyperostosis and skull invasion of the intradural meningioma is not related with the malignancy. High malignant transformation rate(11% in EM, 2% in IM) is remarkable. Although data is scanty and confirmed knowledge is rare, active treatment may be advocated.

The several hypothesis were given about the possible origin of the primary intraosseous meningiomas. Azar-Kia suggested that PIM arose from the arachnoid cap cells trapped in the cranial suture during birth. Eight percent in the Lang, et al's report, 64% in the Crawford, et al's report were suture related on T2-weighted image and T1-weighted enhancing image, and iso- or low signal intensity on T1-weighted image. CT findings have osteolytic features. Common involving area and imaging features are different from intraosseous meningioma. Fibrous dysplasia is a developmental disease so that it commonly occurs in young ages and nearly stops to grow after bone maturation. It has a ground-glass appearance in CT; characteristically ballooning and thinned but intact cortical bone. It shows low signal on T1-weighted MR image, low to isosignal intensity on T2-weighted images and moderate to strong enhancement on T1-weighted contrast enhancing image. Tc-99m MDP scan commonly shows increased uptake in the lesion. Age and lesion detection time in our case is later than that in the fibrous dysplasia and en-masses. Second explanation is that PIM arose from the entrapped dura and arachnoid by previous trauma. But Shuangshoti found that only 0.2% of 504 extradural meningiomas were related with trauma. The third explanation is that extradural meningioma arose from the multipotentential mesenchymal cells. This hypothesis may explain the mass located far from the head and neck.

Differential diagnosis of the skull tumor includes chondroma, fibrous dysplasia, osteoma, osteosarcoma, plasma cell tumor, and so on. Chondomas originate from the embryonic remnants of the notochord, arising at the sphenopatinal area of the skull; clivus, middle fossa, parasella. MRI shows inhomogeneous signal intensity

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**Fig. 1.** Pre-operative brain magnetic resonance(MR) image and computed tomography(CT). A, B: Orbit CT showing a hyperostotic lesion located at the sphenoid bone and lateral orbital wall with the line radiating spicules perpendicular to the bone surface and marginal blurring. C, D: T2 and T1-weighted MR images showing low signal intensity at the lesion. E, F: Gdidinium enhanced T1-weighted MR images showing enhancement of the adjacent soft tissue and dura, but no enhancement of the main bone lesion.

**Fig. 2.** Technesium-99m methylene diposphonate scan showing the increased tracer uptake at the left orbit lateral area and sphenoid bone.
Fig. 3. Pathology. A: Woven bones and lobules of cells (×40, H & E). B: Oval and spindle shaped cells with psammoma bodies (×400, H & E). C: Epithelial membrane antigen positive finding (×400).

Fig. 4. Post-operative computed tomographs showing that not most of the bone mass was removed.

Enhancing patterns in MRI are different. Plasma cell tumor in skull is rare and characteristic of osteolytic feature on simple X-ray, high signal on T1/T2-weighted MR image, homogeneous enhancement on enhancing MR image. It occurs mainly in cranial base rather than in orbit. Osteoma is most common skull tumor and usually grows outward. Osteoma grows more slowly than meningioma and spicules on the surface of the osteoma are not seen on simple X-ray or CT. Osteosarcoma exhibits malignant features of ill-defined margin and cortical destruction, so imaging features are more aggressive and the prognosis is worse than meningioma.

Conclusion

Primary intracranial meningioma is rare one. When skull lesion is found, it should be carefully evaluated and should be taken into account the possibility of the intracranial meningioma.

References