Case Report

Meningioma Mimicking Craniofacial Fibrous Dysplasia

Niphon Chirapapaisan MD*, Komgrid Charngkaew MD**,
Wanicha Chuenkongkaew MD*, Theerapol Witthiwej MD***

** Department of Ophthalmology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand*
*** Department of Pathology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand*
**** Division of Neurosurgery, Department of Surgery, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand*

Background: *A 48 year-old woman with nasal mass and an 8-year history of bilateral hearing loss, developed loss of vision on her right eye for 1 year. Her eye examination showed best corrected visual acuity of 6/12, 6/7.5, and exophthalmos with a deficit in adduction and abduction, a relative afferent pupillary defect and swollen optic disc on her right eye. Her brained CT showed characteristics of craniofacial fibrous dysplasia which were expansile with bilateral sclerotic ground glass and cystic lesion, predominantly at sphenoid and temporal bones. The mass extended to infratemporal fossa, surrounding pterygoid bone at right nasopharynx and at frontal region.*
Objective: *To report a case with meningioma mimicking craniofacial fibrous dysplasia.*
Material and Method: *A right craniotomy for tumor resection was performed.*
Results: *Histopathology revealed a meningioma.*
Conclusion: *This is a rare case of meningioma with an initial manifestation as well as a brained CT finding showing features of craniofacial fibrous dysplasia.*

Keywords: *Meningioma, Craniofacial, Fibrous dysplasia*

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Meningioma is a benign tumor deriving from the arachnoid membrane in the central nervous system. It accounts for 15-20% of primary intracranial tumor and is more frequently in adult woman[1]. It extends extracranially to surrounding orbit, paranasal sinus or nasal cavities with an incidence of 20% of primary intracranial meningioma[2]. Proptosis and loss of vision are clinically dominant in spheno-orbital meningioma. To visualize bony invasion in meningioma, computerized tomography (CT) scan is more effective which typically shows homogeneous, sclerotic or feathered appearance in adjacent bone than MRI which is used to detect soft tissue extension and mostly shows rapid and intense contrast enhancing area[3]. Complete surgical resection of meningioma is preferable procedure due to its high recurrence rate especially with the remaining of inaccessible tissue or bony invasion.

However, a clear radiographic distinction between meningioma and other craniofacial conditions particularly with bone involvement may be difficult for a proper diagnosis which probably misleads to a different surgical approach.

We describe a rare case of meningioma in an adult woman with the initial imaging high suspicion of craniofacial fibrous dysplasia (CFD).

Correspondence to:
Chuenkongkaew W, Department of Ophthalmology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand.
Phone: +66-2-4198037, Fax: +66-2-4111906
E-mail: wim.wanicha@gmail.com

Case Report

A 48-year-old woman with nasal mass presented with progressive loss of vision on her right eye for 1 year. She had a hearing loss both ears since 40 years old.

Eye examination showed best corrected visual acuity of 6/12, 6/7.5. The right eye had exophthalmos with a deficit in adduction and abduction, a relative afferent pupillary defect, and swollen optic disc (Fig. 1).

Cranial CT showed characteristics of expansile with bilateral sclerotic ground glass and cystic lesion, predominantly at sphenoid and temporal bones, the mass extends to infratemporal fossa, surrounding pterygoid bone at right nasopharynx and at frontal region (Fig. 2). CFD was initially in consideration of the diagnosis, and a right craniotomy demonstrated mass,

on histopathology, a piece of flat irregular gray white tissue which is compatible with meningioma (Fig. 3).

Discussion

This was an adult woman who had unilateral exophthalmos and progressive visual loss with swollen optic disc and hearing loss. Her CT brain scan showed bilateral sclerotic ground glass mass with an extension to adjoining bone.

A combination of her clinical manifestations as well as radiographic studies are compatible with CFD, which is a rare benign developmental bone disorder consisting of the progressive replacement of normal bone by abnormal fibrous and osteoid and more commonly affects skull and facial bone in 10 to 25% and 50% of patients with monostotic and polyostotic respectively[3-6]. It usually commences in children and early adolescents with exceptional adulthood in some reports compared to mostly adulthood in meningioma with a woman predilection in both diseases[5].

A predominant facial bone and skull base involvement in CFD causes progressive visual loss, facial disfigure, slowly globe displacement and auditory disturbance as the common manifestations of CFD[5]. Due to more common orbital involvement in CFD than in meningioma, anterior globe displacement which leads to exophthalmos without any orbital decompensation elsewhere in this case does not explain the concurrence diagnosis of CFD, but is more likely found in meningioma. Unilateral swollen optic disc caused by a compressive orbital lesion is more prominent in particularly primary optic nerve sheath meningioma, with a rare occurrence in both sphenoid meningioma and CFD[7,8].

Although a clinical correlation in this case was not completely consistent with CFD. However, it can be radiologically distinguished from meningioma though not a quite clear destination in some cases. In CFD, a typical CT imaging shows either an expansile lesion with a ground-glass appearance or a mixed area of lucency and increased density as "pagetoid" whereas a MRI reveals less distinctive characteristic features of CFD[3,4]. In contrast to CFD, a characteristic imaging in meningioma shows a homogeneous, sclerotic appearance with hyperostosis in an adjacent bone. Further including the consideration of meningioma is particularly an adjacent soft tissue involvement with rapid contrast-enhanced MRI[3].

According to her clinical and radiographic findings, she was initially diagnosed as CFD. Alternatively as the biopsy-proven meningioma by histological demonstration of whorls of meningothelial cells instead of spindle cells with immature woven bone

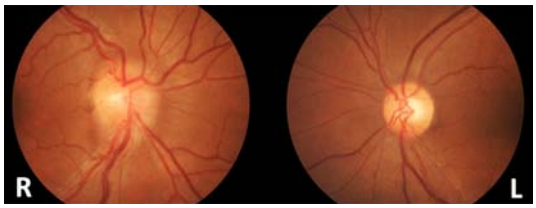

Fig. 1 Swollen optic disc of the right eye (R).

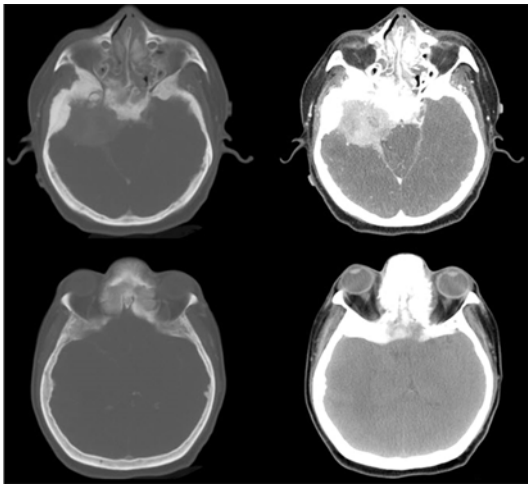

Fig. 2 Contrast-enhanced cranial CT shows characteristics of expansile with bilateral sclerotic ground glass and cystic lesion, predominantly at sphenoid and temporal bones, the mass extends to the infratemporal fossa.

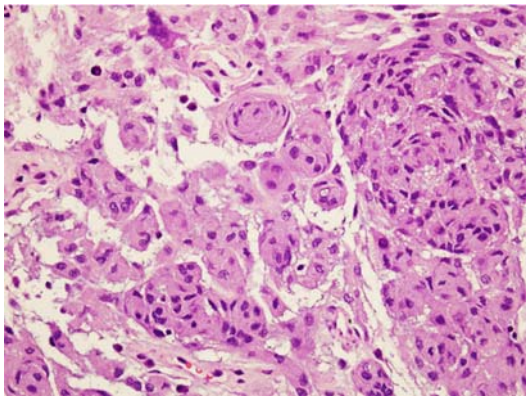

Fig. 3 The histopathology demonstrates whorls of meningothelial cells with occasional intranuclear pseudoinclusions, consistent with meningioma, WHO grade I (H&E, original magnification, 200x).

trabeculae, she then finally was indicated the reversal diagnosis to meningioma. Surgical intervention is exclusively indicated for both meningioma and CFD with inevitable and reluctant progressive visual dysfunction due to optic nerve compression. Although a favorable prognosis with low rate of malignant transformation on CFD, a tremendous narrowing of encroached-optic canal or aesthetic facial damage is an additional indication for surgical treatment[9-11].

Despite a low potential to turn to be malignancy, meningioma tends to grow very fast and may spread beyond the brain. It usually recurs if incomplete removal while partial surgical excision for CFD without any presence of progression is an adequate treatment[12]. According to this increasingly important perspective, meningioma should be entirely resected.

Conclusion

Meningioma and fibrous dysplasia especially craniofacial bone involvement are difficult in an explicit clinical and radiological distinction. However, physicians should have high index of suspicion and be carefully aware of a differentiation between these two entities in making correct diagnosis for more precise, prompt and proper surgical intervention.

What is already known from this topic?

Meningioma is a common entity of brain tumor which not only involving the underlying brain, but also adjacent craniofacial bone.

What this study adds?

Meningioma with hyperostosis of craniofacial bone can mimic some craniofacial conditions, such as fibrous dysplasia. Surgery is the mainstay treatment to relieve mass effect, obtain pathological diagnosis, and improve function as well as cosmetic.

Potential conflicts of interest

None.

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เนื้องอกเยื่อหุ้มสมองที่มีลักษณะคล้ายคลึง fibrous dysplasia ที่กะโหลกศีรษะและใบหน้า

นิพนธ์ จิรภาไพศาล, คมกริช จ่างแก้ว, วณิชา ชื่นกองแก้ว, ธีรพล วิทธิเวช

ภูมิหลัง: *รายงานผู้ป่วยหญิงไทยอายุ 48 ปีมาด้วยอาการสำคัญ คือ ตาขวาม้วมา 1 ปีเคยมีเนื้องอกในโพรงจมูกมาก่อนและเมื่อ 8 ปีก่อนมีอาการหูดับทั้งสองข้าง วัดระดับสายตาข้างขวา 6/12 ข้างซ้าย 6/7.5 ตรวจตาพบ ตาขวาโปน ตาขวากลอกเข้าด้านในและออกด้านนอกไม่ได้ การทำงานของรูม่านตาขวาผิดปกติ และพบขั้วประสาทตาขวาบวม ผลการตรวจวินิจฉัยเพิ่มเติมโดยการเอกซเรย์คอมพิวเตอร์สมองพบลักษณะของ fibrous dysplasia ของกะโหลกศีรษะร่วมกับลักษณะ bilateral sclerotic ground glass และถุงน้ำ โดยเฉพาะที่บริเวณกระดูก sphenoid และกระดูก temporal โดยที่เนื้องอกลุกลามไปยังบริเวณ infratemporal fossa และรอบ ๆ กระดูก pterygoid ที่บริเวณโพรงจมูกด้านขวาและบริเวณกระดูก frontal ผู้ป่วยรายนี้ได้รับการผ่าตัดสมองตัดเนื้องอกออกและผลชิ้นเนื้อทางพยาธิวิทยา พบเป็นเนื้องอกเยื่อหุ้มสมอง*

วัตถุประสงค์: *รายงานผู้ป่วยหนึ่งรายที่เป็นเนื้องอกเยื่อหุ้มสมองซึ่งมีลักษณะคล้ายคลึง fibrous dysplasia ของกะโหลกศีรษะและใบหน้า*

วัสดุและวิธีการ: *ผู้ป่วยได้รับการผ่าตัดเปิดกะโหลกศีรษะข้างขวาเพื่อรักษาเนื้องอก*

ผลการศึกษา: *ผลทางพยาธิวิทยาของเนื้องอกพบว่าเป็นเนื้องอกเยื่อหุ้มสมอง*

สรุป: *โดยสรุปรายงานผู้ป่วยรายนี้เป็นผู้ป่วยโรคเนื้องอกเยื่อหุ้มสมองที่มีลักษณะทางคลินิกและภาพทางรังสีคล้ายคลึงภาวะผิดปกติชนิด Craniofacial fibrous dysplasia ซึ่งพบได้น้อยมาก*