

RECURRENT LEFT SPHENOIDAL RIDGE MENINGIOMA WITH OCULAR MANIFESTATIONS: A Case Report and Literature Review

NO MAGULIKE, BMBCH, FMCO, FWACS, FRCOphth

Department of Ophthalmology, University of Nigeria Teaching Hospital, Enugu, Nigeria

email: zevymagulike@yahoo.com

SUMMARY

A 55-year-old male presented 17 years ago with symptoms of defective vision in the left eye which had lasted for 3 years. This was associated with left-sided headache that was relieved with the use of paracetamol. He also complained of nausea, vomiting and convulsions.

Visual acuity was 6/5 in the right eye and perception of light (PL) in the left eye, unaided. The main findings were disc swelling in the right eye, left optic atrophy, and left afferent pupillary defect. There was also loss of smell in the left nostril. An x-ray of the skull showed a supratentorial hyperdense lesion.

Surgery was suggested to the patient but he decided to travel overseas for treatment and there he had resection of a left sphenoid ridge meningioma. There was a marked improvement of symptoms.

After the patient came back, he was followed up for 2 years, after which he failed to keep further appointments. Twelve years later, he presented again, this time with left proptosis, left ophthalmoplegia, loss of sensation in the left (V₂ and V₃) 5th cranial nerve areas of his face. He had left lagophthalmos with keratinization of the lower conjunctiva and enlargement of the left side of his face.

A CT scan showed a recurrence of the left sphenoid ridge meningioma, spreading into the left orbit, left ethmoid and sphenoid sinuses, middle cranial fossa and pterygopalatine fossa. There was also hyperostosis of the left sphenoid bone. He had a second surgery and the tumour was histologically shown to be a meningioma.

These complications could have been prevented if the patient had kept his appointments. This aim of this case report is to show the importance of regular follow up and early detection of any complication that could be treated immediately. There was a poor follow-up period between the first and second presentations.

Patients must be adequately warned that the consequences of failure to keep appointments after surgery could be very grave.

Key words: recurrent meningioma, poor follow-up, and ocular complications

INTRODUCTION

In 1614, Felix Plater first described meningiomas at an autopsy, and in 1938 Harvey Cushing introduced them as a separate category of extraparenchymal tumours.¹ Meningiomas are believed to arise from arachnoid cap cells, usually attached to the dura. These tumours may arise from any location where the meninges exist, e.g. the nasal cavity, paranasal sinuses, middle ear and mediastinum.²

Meningiomas account for 15% of intracranial tumours. Among these, 90% are intracranial, and commonly occur in the fourth through the sixth decades of life. They are more common in females and rare in children.² Studies in Africa, however, show that meningiomas occur more in males than in females and there is an affinity between intracranial tumours and the youth in Nigeria, with 50% of the cases found in patients within the first two decades of life.^{3,4,5}

Ocular manifestations of intracranial meningiomas, apart from those related to intracranial mass lesions, depend largely on the involvement of the visual pathway. Meningiomas of the sphenoid ridge are traditionally divided into three types: outer, middle and medial.⁶ Outer sphenoid ridge meningiomas are usually accompanied by epilepsy, focal weakness and problem with language functions when present on the left side. Those of the inner sphenoid ridge usually compress the optic nerve and present with early unilateral visual loss, and this may involve the cavernous sinus causing double vision and numbness of the face.⁶

The clinical course of a meningioma characteristically spans a period of years and the tumour is known to recur following surgical intervention.²

Ocular manifestations could get worse following recurrence and spread into the orbit, cavernous sinus,

visual pathway and include bony involvement, leading to hyperostosis.

This case report presents the early and late ocular changes following the recurrence of a left sphenoid ridge meningioma after the initial surgery and stresses the importance of adequate follow-up in order to detect any complications early.

CASE REPORT

A 55-year-old male was first seen 17 years ago at the University of Nigeria Teaching Hospital (UNTH) eye clinic. He complained of defective vision in the left eye of 3 years duration. This was associated with a headache mainly in the left temporal area which was relieved with the use of paracetamol. There were no aggravating factors. He also complained of nausea, vomiting and convulsions.

On examination, visual acuity was 6/5 in the right eye and perception of light (PL) in the left eye, unaided. The main findings were moderate optic disc swelling in the right eye. In the left eye, the pupil showed an afferent pupillary defect; there was optic nerve atrophy. The intraocular pressure was 15.0 mmHg in each eye. There was also loss of smell in the left nostril.

A skull x-ray showed a supratentorial hyperdense defect. Surgery was advised, but the patient opted to travel to Britain, where he had a subtotal resection of a left sphenoidal ridge meningioma, and came back with marked improvement of symptoms. He was followed up for 2 years and then failed to keep further appointments.

Twelve years later, the patient presented at the same clinic with a reoccurrence of symptoms. He complained of discomfort in the left eye with drooping of the left upper lid, bulging and redness of the left eye, and swelling of the left side of his face. He had become hypertensive and was on medication.

Visual acuity was still 6/5 in the right eye with no perception of light (NPL) in the left eye. The main changes were in the left eye with marked ptosis and proptosis. The conjunctiva was congested with keratinization of the lower region. The pupil was mid-dilated and fixed. There was lagophthalmos. Ocular movement showed left ophthalmoplegia. There was total loss of the sense of smell in the left nostril with loss of sensation in the left V2 and V3 areas of the 5th cranial nerve. The left side of the face was enlarged (figures 1a, 1b, 1c). Informed consent was obtained from the patient to take photographs and the case report was sanctioned by the ethics committee of the hospital. The only abnormality in the right eye was a raised intraocular pressure (IOP) of 26.0mmHg with a cup-disc ratio of 0.7. The central visual field using Humphrey's 640 visual field analyses was normal, using Central 24-2 Threshold Test. There was a mean deviation of -5.45DB P (0.5%), and a PSD of 2.84DB P (10%). Systemic examination was normal.

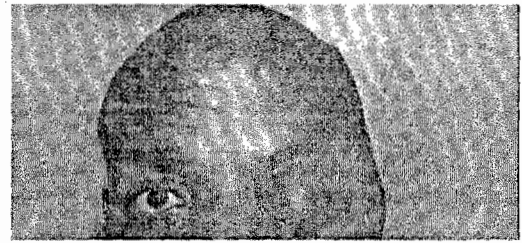


Figure 1a. Front view of patient showing marked ptosis and enlargement of the left side of the face.

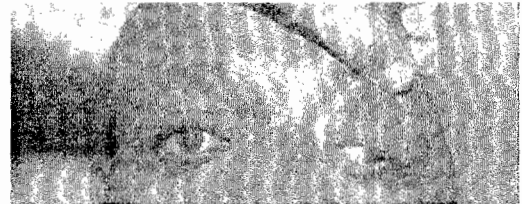


Figure 1b. Front view of patient showing the displacement of the eyeball down and laterally.



Figure 1c. Side view of patient showing the protrusion of the left eye and ptosis.

The patient was started on G. Timoptol 0.5% BD to the right eye and G. Xalatan was added to reduce the IOP to 12.0mmHg. For the left eye, he was placed on artificial tears and chloramphenicol eye ointment nocte and eventually had a lateral tarsorrhaphy.

He was referred immediately to the neurosurgeon for further evaluation. A CT scan was done without and after contrast (ultravist 300 X 60cc). This showed a large (7.5 X 5.3cm) enhanced tumour in the region of the left sphenoid ridge. The tumour extended into the left orbit (producing proptosis), the left ethmoid and sphenoid sinuses, middle cranial fossa and pterygopalatine fossa. There was surrounding oedema of the left temporal lobe and hyperostosis of the sphenoid bone was evident. A full blood count, electrolytes, urea and creatinine were normal.

A diagnosis of recurrent left sphenoidal ridge meningioma was made. The patient had a left fronto-temporal craniotomy and orbital roof craniectomy with excision of the meningioma including the orbital extension. Histology confirmed the diagnosis of a meningioma. The patient is currently being followed up.

DISCUSSION

Visual outcome in brain tumour is mainly determined by the duration of symptoms and tumour size, especially in a situation where the optic nerve or the visual pathway is compressed.⁸ This could be compounded by a recurrence which is not detected early.

In this case, there were initially, a three-year history of symptoms and signs such as defective vision in the left eye, left optic atrophy, right disc swelling, loss of smell in the left nostril – all of which are typical of the Foster Kennedy syndrome. This probably led to a diagnosis of a possible intracranial tumour in the region of the left olfactory lobe, or inner sphenoid ridge.

The patient went to Britain, where he had a resection of a left sphenoidal ridge meningioma with marked improvement of the symptoms. He was however lost to follow up after two years. Twelve year later, the patient re-presented at the clinic with a relapse of symptoms, which were worse. These could have been noticed earlier if he had kept his appointments for follow up.

Meningiomas are known to recur.² In a series by Bonnal et al.⁹ and a case report by Hanakita et al.,¹⁰ recurrence occurred in 2 to 9 years and more. In the case presented, there was a period of 12 years between the first and second resections, giving enough time for recurrence and invasion. Sphenoid ridge meningioma could spread into the orbit with proptosis, pterygomaxillary fossa, cavernous sinus with ophthalmoplegia, ethmoidal and sphenoid sinuses and swelling of the face from bony involvements.^{10,11,12} Similar changes occurred in primary meningioma diagnosed late.¹³ Ophthalmoplegia will indicate involvement of the posterior part of the orbit as the ocular motor and sensory nerves pass through the superior and inferior orbital fissures or when the cavernous sinus is involved. In this case there was no spread into the cavernous sinus.

Patients should be followed up regularly with the aim of early detection of any recurrence.¹⁴ The author has noted that patients fail to keep appointments when they feel better and strong enough to pursue their daily business. In addition, they resent a second intracranial or eye surgery because of probable negative results. It took time to convince this patient to undergo a second surgery; he yielded because he was made to understand that it was a matter of life and death.

CONCLUSION

Recurrence of an intracranial mass that gradually grows unnoticed or not attended to in time, will certainly aggravate symptoms and signs by invading the surrounding areas (intracranial and extracranial), causing further damage to very important tissues compacted into a rigid cranium. This case report shows the consequences of late intervention, caused by poor follow up and consequently late detection of complications from recurrence of the meningioma.

Medical practitioners should make all effort to convince their patients of the importance of keeping their appointments in order to avert serious consequences. This is very important after undergoing any form of surgery. A prolonged follow up should always be the case for lesions that could go on for long or could recur after treatment.

Acknowledgement

I am very grateful to Professor SC Ohaegbulam for his advice and for granting me access to the information about this patient in Memfys Neurosurgery Hospital, Enugu.

References

1. Sall B. Zachariah, Meningioma, sphenoid Wing. e Medicine Specialties >Ophthalmology, Neurologic Disorders. Updated June 2,2006.
2. New York-Presbyterian Hospital – Olfactory Groove and Sphenoid Wing Meningioma, <http://wopub2.med.cornell.edu/cgi-bin/WebObjects/PublicA.woa/4/wa/viewHContent?web...> (1-2-2007).
3. Ohaegbulam SC, Saddeqi N, Ikerionwu S. Intracranial tumours in Enugu, Nigeria. *Cancer* 1980; **46**(10): 2322-2324.
4. Adeloje A. Neoplasms of the brain in the African. *Surg Neurol* 1979; **2**: 247-255.
5. Ohaegbulam SC. Large sphenoid ridge meningioma in a child. *Surg Neurol* 1977; **7**: 143-144.
6. UCLA Neurosurgery: Brain Tumour Diseases and Disorders (Overview and Directory) http://www.neurosurgery.ucla.edu/Diagnoses/BrainTumor/BrainTumorDis_15.html (1-2-2007).
7. Olfactory Groove and Sphenoid Wing Meningiomas New York Presbyterian Hospital. <http://www.nyp.org/health/olfactory-groove-sphenoid-wing-meningiomas.html> (1-28-2007)
8. Sakai N, Nakatani K, Sakai H, Shirakami S, Yoshimura S, Ito T, et al. Visual impairment complicated with meningioma. *No To Shinkei* 1991; **43**(a): 851-6.
9. Bonnal J., Thibaut A., Brotchi J., Born J.: Invading meningiomas of the sphenoid ridge. *J Neurosurg* 1980; **53**(5): 587-99.
10. Hanakita J, Kinuta Y, Takeuchi J, Handa H, Makimoto K. Meningioma of the sphenoid ridge recurring into the pterygomaxillary fossa following craniotomy. A case report and surgical approach. *No Shinkei; Geka.* 1983; **11**(8): 857-64.
11. Ishikura A, Tachibana O, Hunaki N, Watanabe K. Extracranial recurrence of sphenoid ridge meningioma – A case report. *Gan No Rinsho* 1986; **32**(7): 787-91.
12. Pompili A, Derome PJ, Visot A, Guiot G. Hyperostosing meningiomas of the sphenoid ridge – clinical features, surgical therapy, and long-term observations: Review of 49 cases. *Surg Neurol* 1982; **17**(6): 411-6.
13. Shuangshoti S, Sirianugkul S, Suwanwale N. Primary meningioma intimately related to skull: case report and review of Literature. *Surg Neurol* 1994; **42**(6): 476-80.
14. Guo HH, Zhou LF. Further experience in the diagnosis and treatment of sphenoid ridge meningioma. *Zhongna Wai Ke Za Zhi* 1994; **32**(12): 740-2.