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International Journal of Current Research
Vol. 7, Issue, 03, pp.14038-14040, March, 2015

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

ISSN: 0975-833X

CASE REPORT

SPHENOID WING MENINGIOMA IN A 78 YEAR OLD URBAN WOMAN: A LATE PRESENTATION AND NEED FOR ADEQUATE DIAGNOSTIC FACILITIES IN OUR HOSPITALS

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ARTICLE INFO

Article History:
Received 19th December, 2014
Received in revised form
06th January, 2015
Accepted 28th February, 2015
Published online 31st March, 2015

Key words:

Sphenoid wing meningioma,
Orbital, ophthalmic,
Computerized tomography scan.

ABSTRACT

Meningiomas of the Sphenoidal wing occur most commonly in the 5th decade in males and in the 6th decade in females. The diagnosis is confirmed by neuro-imaging using Computerized Tomography and Magnetic Resonance Imaging with contrast, facilities that are uncommonly found in our hospitals. We herein present a case of Sphenoid Wing Meningioma diagnosed in a 78year old urban dwelling woman 5 years after initial presentation, highlighting the late presentation and the need for availability of adequate diagnostic facilities in our hospitals.

INTRODUCTION

Meningiomas are solid, lobulated, usually rounded masses with well-defined dural bases that project into and compress the brain tissue but are easily separated from it (Graham *et al*., 2008; Frosh *et al*., 2010). They usually arise within the cranial cavity, are dura-based and found in the area of the superior sagittal sinus or from the base of the skull mainly in the region of the olfactory groove or the Sphenoidal ridge (Frosch *et al*., 2010; Rosai 2004). They are mostly benign, slow growing tumours of the adults that present either with vague non-localized symptoms or with symptoms due to compression of the underlying brain tissue. Meningiomas appear in childhood or adolescence (Germano *et al*., 1994; Perry *et al*., 2001) but most are encountered in middle or later adult life (Longstreth *et al*., 1993; Karp *et al*., 1974). Meningiomas account for about 15% of brain tumours (Maroon *et al*., 1994). Meningiomas of the Sphenoidal ridge are thought to be less aggressive in behavior than those arising from the meninges of the optic nerve (Karp *et al*., 1974). Meningioma arising from the optic meninges generally cause some loss of vision, atrophy of the optic nerve and exopthalmus while those from the inner portion of the Sphenoidal ridge cause more severe compression of the optic nerve within the optic canal resulting in papilloedema or optic atrophy before proptosis (Rosai J, 2004; Longstreth *et al*., 1993). In a good number of patients, it is known that these spheno-orbital meningiomas cause minimal discomfort and symptoms, thus in the absence of risk factors, a "wait and see" policy is advised while for those with large tumours or with large soft tissue component at first visit or with fast growing tumours (i.e. >1cm^3/year), a follow-up examination every 6months is necessary (Saeed *et al*., 2010).

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Case report

Mrs E came to the eye clinic with a 5 year history of swelling of the right temple and the right eye. She also complained of aches and pains around the affected eye. She had been to various eye and medical practitioners concerning the eye over the years. On examination she was frail looking but quite alert and well oriented in time, person and place. There was a large mass on the right temporal region that was soft in consistency, non-tender and not inflamed (Figure 1). Ocular examination revealed acuities of No Perception of light [NPL] in the right eye and 6/36 in the left eye.

The right eye was markedly proptosed, with dessication and xerophthalmia, which had lead to perforation and extrusion of

the crystalline lens. The conjunctiva was thickened and lichenified, with marked inflammation and echemosis. The eyelids were markedly swollen and pigmented (Figure 2). No fundal details were discerned. In the left eye there was a nuclear sclerotic cataract grade 3. Fundal examination including the optic nerve was normal.

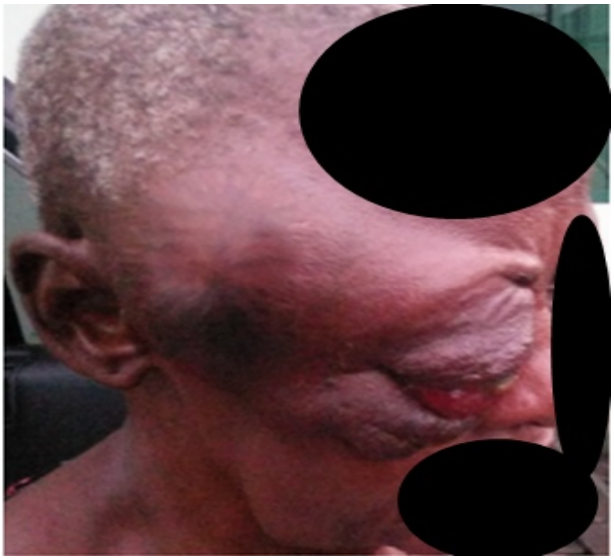

Figure 1. Right temporal region showing a large mass

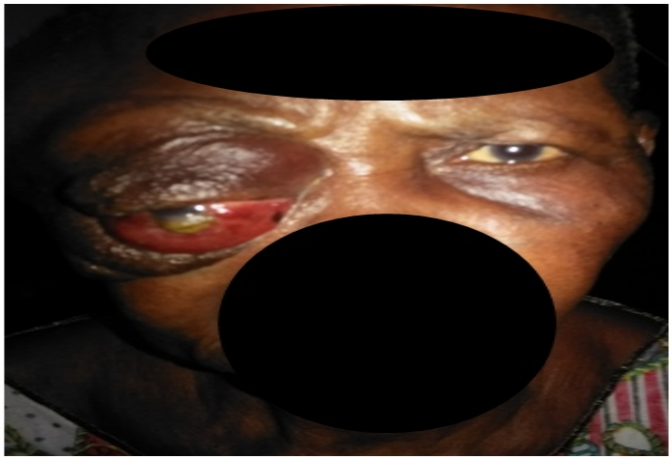

Figure 2. Right eye showing marked proptosis with extrusion of the crystalline lens, thickened and lichenified conjunctiva with marked inflammation and echemosis. The eyelids were markedly swollen and pigmented

A Computerized Tomography Scan revealed the presence of a localized lesion in the sphenoid region of the base of the skull. A full blood count was normal, so were the Urea and Creatinine levels. The random glucose and glycosylated hemoglobin levels were also normal. Given these findings, and the poor prognosis for visual outcome, the options were discussed with the patient and her family. It was decided to enucleate the eye and debulk the tumour. Following surgery, the histopathology report revealed tumour disposed in lobular microachitecture and composed of cells having delicate round or oval nuclei with inconspicuous nucleoli and indistinct cytoplasm (Figures 3 & 4). There were also psammoma bodies with mild infiltration by foamy macrophages and a diagnosis of sphenoid wing meningioma was confirmed. She was referred to the regional neurosurgical centre after discharge. Permission to report this case was sought from the patient and she consented to publishing it anonymously.

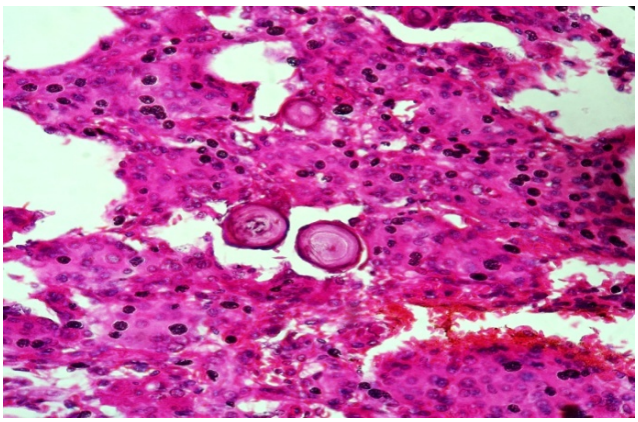

Figure 3. Sphenoid Wing Meningioma showing tumour cells with delicate round or oval nuclei, inconspicuous nucleoli and indistinct cytoplasm with some psammoma bodies (H&E X400)

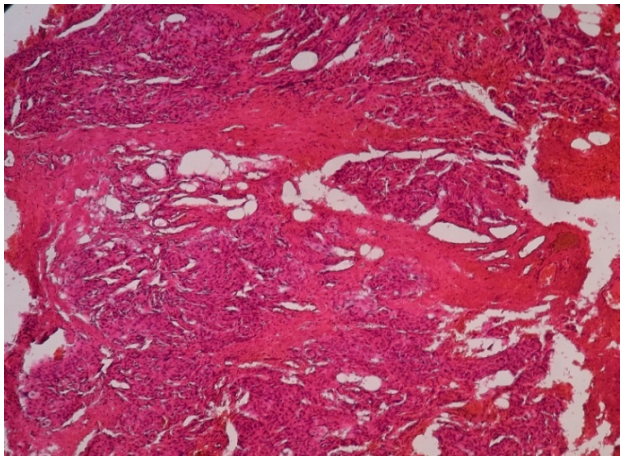

Figure 4. Meningioma of the sphenoid wing showing whorls of tumour cells interspersed by fibrous tissues (H&E X100)

DISCUSSION

The sphenoid bone is situated at the base of the skull in front of the temporalis and basilar part of the occipital bone. It is divided into a medial portion or body, two great wings, two small wings and two pterygoid processes which project from it below (Gray, 1918). The greater wing of the sphenoid forms the posterior part of the lateral wall of the orbit with the frontal process of the zygoma forming the anterior wall. A part of the body of the sphenoid bone contributes to the lateral wall of the orbit along with the ethmoid and frontal process of the maxilla. The roof of the orbit is formed by the frontal bone and the lesser wing of the sphenoid near the apex (Gray, 1918). The optic canal is located between the roots of the lesser wing of the sphenoid bone and transmits the optic nerve and ophthalmic artery (Turvey and Golden, 2012). From these anatomical descriptions, it is evident that pathological processes involving the sphenoid bone have a direct effect on adjacent orbital contents and indeed the eyeball. Meningiomas are the most

common tumour of the sphenoid wing with most originating from the lesser wing (Maroon *et al*., 1994). They are considered to be benign tumours however they may be invasive, spreading to the dura of the frontal, temporal, orbital and sphenoid regions (Samii *et al*., 1996). Sphenoidal wing meningiomas may involve the walls of the cavernous sinus medially, spread to the temporal bone laterally or invade the orbit anteriorly. Tumours found in the external third of the sphenoid bone are of two types: en-plague and globoid meningiomas (Wikipedia, the free encyclopedia, 2015). En plague meningiomas present as slowly increasing proptosis with downward dystopia largely due to reactive orbital hyperostosis (Frosch *et al*., 2010). Diplopia commonly results when the tumour invades the orbit. On the other hand, those with globoid meningiomas present with signs and symptoms raised intracranial pressure like headache and papilloedema (Wikipedia, the free encyclopedia, 2015). Fohanno and Bitar (1986) further classified these tumours into medial globular arising from medial third, lateral globular arising from the lateral two-thirds, en plague and invasive meningiomas.

Meningiomas are more common in females (Frosh *et al*., 2010) as is our finding with the index case. Similarly Mezue *et al* (2012) reported a slight female preponderance (M:F = 1:1.08) among the 74 patients diagnosed with intracranial meningiomas in Enugu, South Eastern Nigeria. The incidence increases with age (Graham *et al*., 2008), peaks in males at the 5th decade while in females, peak age incidence is in the 6th decade. Our case however presented lately in the 8th decade of life. Also notably she was diagnosed with the late signs of the tumour including marked proptosis with perforation and extrusion of the crystalline lens, acuities of NPL and a large mass on the right temporal region (bony invasion). Had the tumour been diagnosed with adequate facility earlier, these late effects of the tumour would have been averted. Although morbidity is generally high, the mortality rate from the benign tumour is low. Therefore management from ophthalmic point of view is underpinned by preservation of vision where possible especially where the lesion is diagnosed early. Diagnosis is confirmed by homogenous enhancement on urgent neuro-imaging: Computerized Tomography and Magnetic Resonance Imaging with contrast. These are facilities that are rarely available in hospitals in our environment. Based on the level of involvement, the tumour is best managed in a multi-disciplinary fashion, involving the ophthalmic orbital surgeon and the neurosurgical team. In their series, Simas *et al* (2013) found that total surgical resection was difficult and had a high recurrence rate in patients with supraorbital fissure involvement and they recommended radiation therapy in such residual tumour locations.

Conclusion

Meningiomas of the Sphenoidal wing can present quite late after the known peak incidence age. Early and adequate diagnosis at presentation is based on neuro-imaging using Computerized Tomography and Magnetic Resonance Imaging with contrast, underscoring the need for these diagnostic facilities in our hospitals.

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