PRIMARY INTRA-ORBITAL OPTIC NERVE SHEATH MENINGIOMA IN AN ADOLESCENT: A Case Report

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SUMMARY

Background: Primary optic nerve sheath meningioma is rare, but nearly always results in blindness; whether left alone or surgically treated.

Case report: A 19-year-old male student without antecedents presented to the ophthalmic service of the University of Ilorin Teaching Hospital, Ilorin, Nigeria, with a two-month history of progressive proptosis, pain, and a foul smelling discharge in the right eye. Upon clinical examination with a Hertel's exophthalmometer, the degree of axial proptosis of the right eye was found to be 20mm compared with 16mm in the left eye. Visual acuity was 6/18 in the right eye and 6/5 in the left eye, with a right relative afferent pupillary defect. An ocular ultrasound revealed intra-orbital soft tissue mass. Computed tomography (CT) scanning of the head was not done at this time due to financial constraints on the part of the patient. He thereafter defaulted from follow-up but re-presented a year later with more severe proptosis, conjuctival chemosis, infected exposure keratitis and total loss of vision in the right eye. A computed tomography of the head revealed an enhancing right retro-bulbar intra-orbital soft tissue mass. Enucleation of the right eye and complete extirpation of both the tumour and the intra-orbital optic nerve were carried out without complications. The histopathology revealed transitional meningioma of the optic nerve sheath. The patient is now socially rehabilitated, wears an artificial eye and has returned to his former occupation and society.

Conclusion: This case typically illustrates the complications that could arise if a treatable benign condition is left untreated because of limited affordable diagnostic facilities or the adoption of an unmonitored 'observational strategy' in the early phase of the disease.

Key words: proptosis, optic nerve sheath, meningioma

INTRODUCTION

Among space occupying intracranial lesions in the African indigene, meningiomas have consistently featured as a prominent histological group constituting 20% of all lesions.[1] In spite of this relatively common occurrence, optic nerve sheath meningiomas (ONSMs) are rare tumours in our experience and in other centres in Nigeria.[2] The present case is the first to be reported from this centre in its 5 years of existence as a neurosurgical service. We are reporting this case because of its rarity, the diagnostic and treatment challenges posed and the relatively young age of the patient.

CASE REPORT

A 19-year-old male student without antecedents presented to the ophthalmic service of the University Teaching Hospital, Ilorin, Nigeria in April 2003, with a 2-month history of progressive proptosis (fig. 1), associated with pain and a foul smelling discharge in the right eye. The degree of axial proptosis of the right eye, measured with Hertel's exophthalmometer was 20mm. This was significantly different from the value of 16 mm in the left eye. The proptosis was not reducible and there was no bruit over it. Visual acuity was 6/18 in the right eye and 6/5 in the left eye, with relative afferent pupillary defect in the right eye. There was full external ocular motility in both eyes, and the lens, vitreous, discs, macula and posterior poles were normal. Ocular ultrasound revealed an intra-orbital soft tissue shadow. A computed tomography scan was requested but this was not done because of financial constraints. The patient thereafter discontinued follow-up until he re-

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presented a year later with more severe proptosis, pain, and total loss of vision in the right eye. An examination revealed no perception of light, severe axial proptosis, severe conjunctival chemosis and infected exposure keratitis in the right eye. The left eye was essentially normal.

A cranial computed tomography (fig. 2) revealed a brilliantly enhancing right retro-bulbar intra-orbital soft tissue mass which was attached to the intra-orbital portion of the optic nerve, displacing the right eyeball antero-laterally and compressing its posterior wall.

Figure 1. Clinical photograph showing a teenage male patient with right intra-orbital primary optic nerve sheath meningioma, severe axial proptosis and exposure keratitis in the right eye.

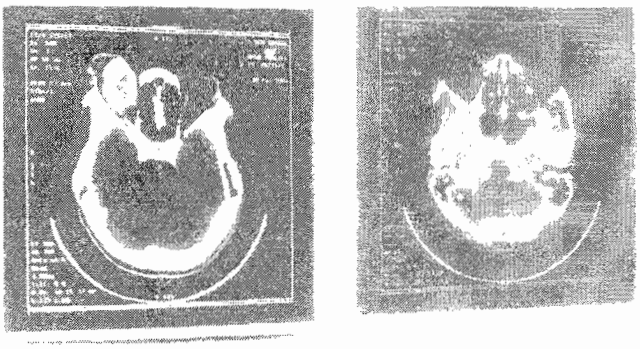

Figure 2. Computed tomography of the head, sinuses and orbit showing a right retrobulbar contrast enhancing soft tissue mass attached to the optic nerve and displacing the eyeball antero-laterally.

Other mass effects include compression of the ipsilateral maxillary antrum and widening of the orbital cavity. Enucleation of the right eye and complete extirpation of both the tumour and the intra-orbital portion of the optic nerve were carried out without complications via a trans-orbital approach. The trans-cranial approach was avoided in order to minimize post-operative morbidity. A histology of the surgical specimen revealed transitional meningioma of the optic nerve sheath. Examinations at intervals of two months for two years after discharge have shown no deterioration, suggesting that the tumour did not recur. The patient is now socially rehabilitated, no longer feels withdrawn and is back to his former society and previous occupation. He is fitted with an artificial eye of almost the same size and colour as the normal eye and uses sunglasses to protect the good eye.

DISCUSSION

The incidence of meningioma increases with advancing age and peaks in the seventh decade of life in women and in the eighth decade in men.[3] Meningiomas have a 2:1 female to male ratio in Caucasians[3] though this female preponderance may not be as pronounced in black Africans.[4] Primary ONSMs arise from the cap cells of the arachnoids surrounding the intra-orbital or, less frequently, the intra-canalicular optic nerve,[3] and occur predominantly in middle-aged women.[5] Meningiomas are therefore rare in the age and sex group of the patient presented. When they occur, they tend to be associated with background neurofibromatosis, with cranial meningioma occurring in 4.2-16% of younger patients with neurofibromatosis type-1. In addition, meningiomas in this age group tend to be more aggressive in terms of growth rate, tumour size, propensity to undergo malignant changes and recurrent rate.[3] Aggressive tumour growth could be responsible for the rapid visual deterioration experienced by this patient within one year of initial presentation when a 'wait-and-see' policy would have appeared appropriate as a management strategy. However, an observational policy without proper monitoring would also have resulted in a worse visual prognosis.

The types of symptoms of ONSMs that manifest depend on the precise location of the tumour along the course of the optic nerve. However, the most characteristic profile is a painless chronic compressive optic neuropathy that may or may not be accompanied by proptosis.[3] The presence of a progressive proptosis as the main feature at presentation in our patient suggests an exophytic tumour around the intra-orbital portion of the nerve. Intra-orbital meningiomas give rise to the three classical symptoms of orbital tumour, including proptosis, impairment of mobility of the globe and visual deterioration.[2] When visual failure is significantly disturbing, as it is in many cases, the patient often presents early in the hospital before proptosis develops.[2] Most of such cases have intra-canalicular meningioma and usually present with visual failure without proptosis.[2] The most common features of intra-cranial ONSMs are atrophy of the optic nerve and a slowly progressive loss of vision in one eye if the pre-chiasmal portion of the nerve is affected.[2] When the chiasma is involved, these signs may become bilateral, and visual field examination then shows bitemporal or homonymous hemianopia or total blindness in one eye and hemianopia in the other.[2] However, the classic triad

of ONSMs irrespective of the site is visual loss, optic atrophy and optociliary shunt vessel. Visual loss is usually very slow and is associated with preservation of the central visual field for years.[3] When an optociliary venous shunt in the disc coexists with visual loss or disc pallor, it is suggestive but not pathognomonic of optic nerve sheath meningioma. The vessels involved in the venous shunting are the retino-choroid venous collaterals arising from chronic compression of the central retinal vein.[3]

The computed tomography (CT) scan with contrast enhancement is an excellent imaging technique for the evaluation of ONSMs, and was the diagnostic modality employed for this patient.[3] However, a CT scan should be done early during the evolution of the disease in order to facilitate early detection, prompt treatment and better outcome before onset of irreversible visual morbidity as it occurred in this case. Magnetic resonance imaging (MRI) with gadolinium contrast enhancement is however superior to CT, but it is not readily available in most African countries. In addition, the cost of an investigation is highly prohibitive. Furthermore, MRI with selective partial inversion recovery/fluid attenuated inversion recovery (SPIR/FLAIR) sequences, has been shown to offer significant advantages over currently used fat-suppressed MRI sequences for the investigation of orbital disease.[6]

Though the treatment of ONSM remains controversial[7] and several treatment options now exist, which include observation, radiation alone, surgical excision alone, and combined radiation and surgery,[8] hormone therapy and more recently hydroxyurea, the most reasonable option for this patient was an en-bloc resection of the tumour and the intra-orbital optic nerve together with enucleation of the blind, ulcerated and infected right eye. He was considered inappropriate for 'wait-and-see' strategy or radiation therapy as primary options. This was because he presented with total blindness, large tumour size, marked proptosis and severe exposure keratitis. Some authors would recommend leaving the nerve and the eyeball behind since there is no intra-cranial extension, while others opine that whenever surgery is indicated it should be an en-bloc excision [9] but the nerve and eyeball could not be spared for the same reasons and the improvement in quality of life following resection and rehabilitation gives credibility to this option.

Primary optic nerve sheath meningioma is a rare but almost invariably blinding tumour when its natural history is observed in a 'wait and see' strategy.[10] Surgery has thus been advocated only in cases of progressive disease involving intra-cranial structures, as it leads to iatrogenic blindness in the overwhelming majority of cases. Therefore, treatment options bearing less risk of functional deterioration are highly desirable; both in cases of intra-cranial involvement as well as during earlier phases of the disease, which are currently generally left untreated.

Radiotherapy has now become a major advance and has proved successful in preventing the blindness that could follow surgery[8] in many studies involving large series of patients. Stereotactic fractionated conformal radiotherapy has recently been used extensively to halt the spread of ONSM,[10] though this facility is not available in our setting. However, radiotherapy could also be administered as conventional type or as interstitial brachytherapy. In order to minimize side effects, most authors in the developed world now prefer fractionated stereotactic external radiation which is also associated with better visual outcome.[10, 11] Anticancer chemotherapy as an option is better reserved for patients with unresectable, recurrent or previously irradiated meningiomas.[3] It has recently been observed that chemotherapeutic treatment with hydroxyurea, 20mg/kg body weight/day orally for several months has proven to be clinically effective in the management of ONSM. This leads to improvement in vision though without detectable change in tumour size.[12] If this patient had presented and was diagnosed earlier, he would have benefited from initial observation followed by conventional radiotherapy, but if he could have obtained financial assistance a referral elsewhere outside Nigeria for a better vision preserving stereotactic fractionated conformal radiotherapy would have been preferred.

The visual morbidity accompanying an otherwise curable benign condition as it occurred in this case clearly identifies the need for improvement in affordable diagnostic facilities in Nigeria and other developing countries of Africa.

CONCLUSION

This case typically illustrates the kind of complications that could arise from a treatable benign condition because of limited affordable diagnostic facilities or where an unmonitored observational strategy is adopted in the early phase of the disease.

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