Intraorbital meningioma in a Young African Female: A case report

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Abstract

Background: Meningiomas are tumours that arise from arachnoid cap cells of the meningeal arachnoid villi. Intra-orbital meningiomas account for a small percentage of all intra-orbital tumours and can either arise primarily from within the orbit or secondarily invade the orbit from the intracranial cavity.

Presenting Complaint/Investigations: We present a case of a 30 year old woman who presented with a 1 year history of gradual protrusion of the left eye, deteriorating vision and restriction of eye ball motility. A CT scan showed a soft tissue mass in the left lower orbit with a smooth outline, displacing the eyeball antero-superiorly and distinct from surrounding structures.

Diagnosis/therapeutic intervention/outcome: Surgery was done with the removal of a well circumscribed mass from the lower quadrant of the orbit via a transverse incision in the inferior conjunctival fornix. Pathological examination revealed a well circumscribed encapsulated mass with grayish white cut surfaces. Histological examination revealed a meningothelial meningioma WHO grade 1. Patient did well postoperatively with marked improvement of vision and restoration of orbital anatomy.

Conclusion: The diversity of subtypes and grades of meningioma encountered within central nervous system is not usually encountered in primary intra-orbital meningiomas, most cases of intra-orbital meningioma are benign. Surgical removal of intra-orbital meningiomas is difficult and recurrence is common if surgical excision is incomplete. The index case appears to be completely excised.

Keywords: Intra-orbital Meningioma, Meningothelial, Ectopic

Date received: 11 December 2020; accepted: 22 January 2021

Highland Med Res J 2020;20(2):62-65

Introduction

Meningiomas can occur in intracranial, intra-spinal and orbital locations with rare examples reported outside the neural axis.[1] These tumours arise from arachnoid cap cells of the meningeal arachnoid villi.[2] Meningiomas account for between 3-10% of all intra-orbital neoplasms.[3,4] Orbital meningiomas are of either primary or secondary origin, with primary lesions arising from within the orbit and secondary lesions extending into the orbit from adjacent structures.[2,3,5] Primary intra-orbital meningiomas mostly arise from the arachnoid around the optic canal or the posterior pole of the eyeball.[5, 6] Approximately 70% of orbital meningiomas are secondary and commonly occur as invasion of the orbit by tumours of the sphenoid wing, clinoid, cavernous sinus and tuberculum sella.[2,3]

There is scanty published literature and very few cases of intra-orbital meningioma documented in Nigeria. The percentage of intra-orbital meningioma amongst orbito-ocular tumours ranged from 1-4% in reports from Kano, Benin and Zaria. [7,8,9]These reports however did not provide details such as presentation, treatment, and outcome on the cases of orbital meningioma seen in their reports. Odebode et al reported a case of intra-orbital meningioma arising from the optic nerve sheath of a 19 year old male at the University of Ilorin teaching hospital in 2006.[10] The patient presented late with complications (severe proptosis, infected exposure keratitis and total blindness). Enucleation was carried out, the tumour was of the transitional meningioma variant and there was no recurrence at 24 months of follow up after surgery.[10] Intra-orbital meningiomas arising from ectopic arachnoid tissue are very rare and some authors have disputed the existence of these "ectopic orbital meningiomas".[2] Huang et al reported a series of six ectopic orbital meningiomas which were selected from 162 cases of orbital meningioma diagnosed over an 18-year period of review.[2] These patients had an average age of 33.2 years (range 7-56 years), with a male to female ratio of 2:1, The six patients presented with exophthalmos, eyelid edema and eyeball motility impairment. Five of the six cases were of the meningothelial variant and one was of the psamommatous type. There was no further deterioration of vision after surgery and non re-occurred after surgery.[2] We present this case of ectopic orbital meningioma because of its rarity.

THE CASE

History and symptoms

A 30 year old negroid female patient presented with a 1

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year history of gradual protrusion of the left eye and deteriorating vision. She also had double vision, blurred vision, tearing, redness, pain on movement of the eye, swelling around the orbit and inability to completely close the left eyelids. There was no complaint in the right eye; no history of swellings in other parts of the body and no history of loss of smell.

Pre-operation findings

Examination of the left eye revealed a visual acuity of 1/60. There was periorbital fullness and lagophthalmos, with severe chemosis and dilated engorged conjunctival vessels inferiorly. There was non-axial proptosis of 8mm (25mm anterior displacement as compared to 17mm on the right with Hertels exophthalmometer) with the globe displaced superomedially by 9mm and associated resistance to retropulsion. The cornea was clear not staining with fluorescein dye and anterior chamber was of normal depth and calm, the pupil was round and reacting briskly to light reflex. Fundoscopy revealed a pink round disc with vertical cup disc ratio of 0.7, diffuse retinal edema and choroidal folds. Intraocular pressure was 12mmhg in the right and 17mmhg in the left eye with an applanation tonometer. There was restriction of ocular motility in downward and outward gaze.
The right eye was essentially normal.

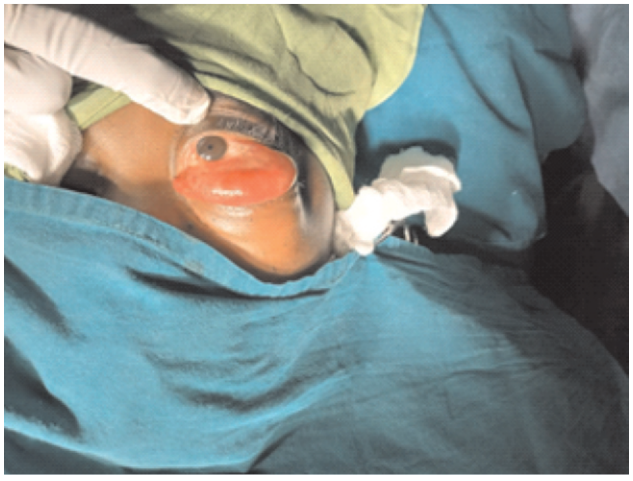

Figure 1. Upward and outward displacement of the eyeball with conjunctival chemosis.

Investigations done

CT scan showed a soft tissue mass that measured 3.9x3.2cm in the left lower orbit, with a smooth outline and displacing the eyeball anterior-superiorly. The mass was avidly enhancing and was seen distinct from, but close to the lateral rectus. The left eyeball appeared normal in morphology and density. The remaining intra-orbital contents were normal. No bony involvement was seen. The right eyeball and orbit appeared normal in morphology and density.

An impression of an orbital mass? cause was made.

Intra-operative findings

Surgery was done under general anesthesia after obtaining an informed consent and after the anesthetist had reviewed and the patient confirmed fit for surgery. A transverse incision was made in the inferior conjunctival fornix to access the orbit. Blunt dissection was done to expose a well circumscribed intra-orbital mass in the lower outer quadrant of the orbit. This was totally removed while applying traction with the aid of a cryo probe. The left eye ball was intact. Conjunctival incision was closed with 3 interrupted sutures using 6/0 vicryl suture. Haemostasis was achieved with pressure packing.

Pathology of specimen

On gross examination the tissue consisted of an encapsulated grayish-brown spherical mass which measured about 3.5cm in its widest diameter. Cut sections revealed uniform grayish white solid surfaces.

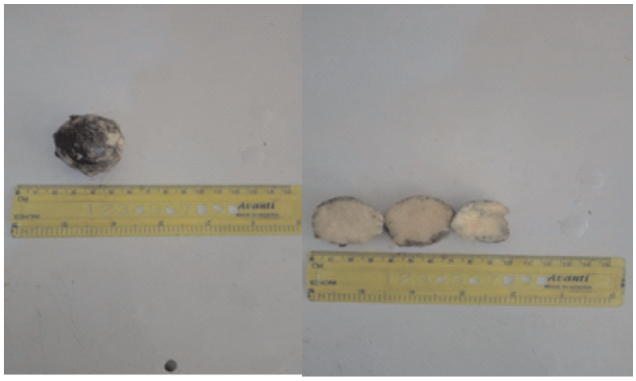

Figure 2. Roughly spherical encapsulated mass with a grayish white cut surface

Histological examination of the mass showed a fibrous capsule overlying lobules of epithelioid meningothelial cells in syncytium having multiple areas of whorling. These cells have round to ovoid nuclei with occasional intra-nuclear pseudoinclusions. Occasional Psammoma bodies and vascular channels were seen within the tumour. Also seen are areas of myxoid change and collagen deposition. A histological diagnosis of Meningothelial meningioma WHO grade I was made.

Post-operative care

Pressure packing was left in place for 48 hours. The patient was placed on oral antibiotics (Augmentin 625mg twice daily for 7 days), analgesics (Diclofenac 50mg twice daily for 5 days), chymotrypsin and Vitamin C tablets. She was discharged on the 3rd post-operative day.

Post-operative follow up

Four weeks post operatively, the patient was in stable condition with an improved visual acuity of 2/60, there was mild conjunctival injection with intact sutures in the inferior fornix. The cornea was clear, anterior chamber was calm, pupils reactive, lens transparent, fundus was pink with normal vessels and macula with no restriction in extraocular muscles movement.

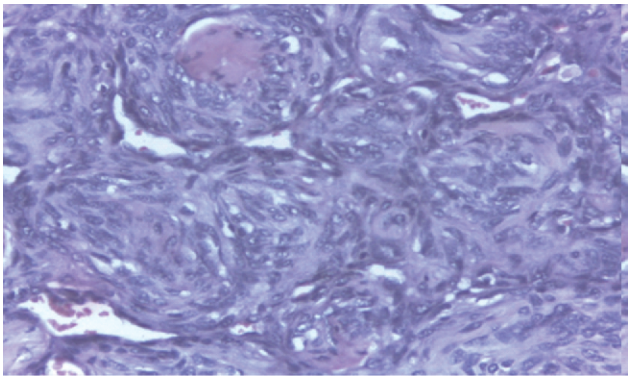

Figure 3. Photomicrograph (haematoxylin and eosin x 400 magnification) showing meningothelial cells in whorls, with nuclear pseudo inclusions

Visual acuity had been on a steady improvement since surgery and as at last visit 6 months post operatively her VA was 6/9 in the left eye with good extraocular motility and restoration of normal anatomy. The patient was however lost to follow up at about 6 months post surgery.

Discussion

This is the first documented case of intra-orbital meningioma recorded at the Jos University Teaching Hospital (JUTH) since reliable data documentation of tumours began in the 1980s. Published literature suggests this tumour is rare in Nigeria and the West African subregion.[7,8,11] Orbital meningiomas accounted for 0.5 percent of ophthalmic tumours in Kano state Northern Nigeria, with 2 cases diagnosed over a 12 year period of review.[7] In Benin, southern Nigeria one case of orbital meningioma was recorded in a 10 year study of orbito-ocular tumours.[8] A similar study at the Korle-bu Teaching Hospital, Accra, Ghana did not document any case of orbital meningioma over a 4 year period of review in which 190 orbital and adnexal masses were biopsied.[11] In contrast meningiomas account for between 3-10% of orbital tumours in studies from other parts of the world.[3,4]

Meningioma is the most common intracranial tumour with most cases being benign.[1,4] They represent between 18 – 20% of all intracranial tumours.[3,12] Majority of meningiomas occur in intracranial, intraspinal and intra-orbital locations, with rare cases outside the neural axis such as the glabella and lungs.[1,6] Secondary orbital meningiomas arise intracranially and extend into the orbit, with the sphenoid wing being the most common point of origin.[2,3,5] Meningiomas of the clinoid, tuberculum sella, olfactory grove, cavernous sinus and planum sphenoidale can also secondarily invade the orbit.[2,3,5] Meningiomas arising primarily within the orbital cavity account for approximately 30% of all intra-orbital meningiomas.[2] Primary tumours can arise from the arachnoid around the optic canal and posterior pole of the eye or less frequently from ectopic arachnoid in the dura, periosteum and orbit.[2,4,6] In this case presented, the tumour appears to arise from ectopic arachnoid tissue in the anterior inferior orbit.

Intra-orbital meningiomas are seen most frequently in young and middle aged women with most cases occurring between the ages 30 to 50 years (73-84% of cases occur in females).[3,5,13,14] The higher incidence in females is thought to be related to the presence of estrogen and progesterone receptors in these tumours.[14] These tumours rarely occur in children, however neurofibromatosis is a known risk factor of orbital meningioma in children, and these neurofibromatosis associated tumours are more aggressive.[3,5]

Neurofibromatosis is also associated with bilateral intraorbital meningiomas.[13] This case was a young female who fits the profile of patients with intra-orbital meningioma. The patient has no clinical features suggestive of neurofibromatosis, however genetic tests were not carried out to confirm.

Clinical features of meningioma are dependent on the location of the tumour. Patients with tumours arising from the optic nerve sheath usually present with progressive visual loss while proptosis is more commonly seen in patients with secondary intra-orbital meningiomas.[3,5,12] Diplopia, pain and extraocular muscle dysfunction are also commonly observed.[3] The index case presented with gradual loss of vision and protrusion of the eye ball with resultant marked improvement in visual acuity after surgery. This marked improvement of visual acuity was possible because the symptoms resulted from compression of the eye ball rather than damage to the optic nerve as is commonly seen in many primary intra-orbital meningiomas. The associated double vision, restriction in eyeball movement, conjunctival redness, and inability to close the eye lids were all reversed after surgery.

The WHO classification of tumours of the nervous system recognizes 15 histological subtypes and 3 grades of meningioma. Grading of meningiomas is based on pathologic subtype, mitotic index and invasiveness. The diversity of subtypes and grades encountered within central nervous system is not usually encountered in primary intra-orbital meningiomas.[5,12,14] Higher grade

tumours have a greater risk of recurrence. Most intra-orbital meningiomas are WHO grade I tumours, grade II tumours are categorized as atypical and grade III malignant. The meningothelial and transitional subtypes are most frequently seen within the orbit.[14] The index case is a WHO grade I tumour (meningothelial meningioma) which is the most common variant encountered worldwide.[14] The tumour in this patient was therefore not aggressive with complete surgical removal being curative.

Orbital meningiomas are difficult to remove surgically and recurrence after surgical intervention is high, recurrences range from 10-23%.[3] Use of fractionated radiotherapy and particularly stereotactic radiation has shown excellent short and long term prognosis in intra-orbital meningiomas.[5] Radiation therapy is better suited for cases involving the optic nerve and surgery more suited for secondary tumours.[13] The index case was surgically removed and appeared to be completely excised, there was no involvement of the optic nerve or other features such as residual disease to warrant consideration of radiation therapy.

Conclusion

Orbital meningiomas are rare ocular tumours which appear to be even rarer in the negroid population. The rarity in the black population may be genetic or as a result of under-diagnosis. Further research and surveillance is necessary to assess the true incidence in the black population.

References

1. Perry A, Louis DN, Budka A, Von Deimling A, Sahn F, Rusling EJ et al. Meningioma. In: Hiroko DNL, Otmar O, Webster WK, Cavanee, eds. WHO classification of tumours of the central nervous system. 4th ed. Lyon: IARC press. 2016: 232-245
2. Huang X, Tang D, Wu T, Jian T, Sun F. Ectopic Orbital Meningioma: a retrospective case series. BMC Opthalmol. 2018;296:1-7.doi.org/10.1186/s12886-018-0959-z.
3. Silva CE, Freitas PE, Romero ADCB, Pereyra TM, Fonseca VF, Martins WA et al. Orbital Meningiomas. J Bras Neurocirurg. 2010; 21(1):31-38.
4. Garcia GA, Choy AE, Hasso N, Mincler DS. Malignant orbital meningioma originating from the frontal lobe. Ocul Oncol Pathol. 2018;4:186-190. Doi 10.1159/000481509.
5. Jain D, Ebrahimi KB, Miller NR, Eberhart CG. Intraorbital Meningiomas: A pathologic review using current world health organization criteria. Arch Pathol Lab Med 2010;134:766-770
6. Verdijk RM, Porcerella I, Mooy CM, The Orbit, including the lacrimal gland and lacrimal drainage system. In: Heegaard S, Grossniklaus H, eds. Eye Pathology; An illustrated guide. Heidelberg: Springer-Verlag. 2015:639-642.
7. Umar AB, Ochicha O, Ilyasu Y. A pathologic review of ophthalmic tumours in Kano, Northern Nigeria. Niger J Basic Clin Sci. 2012;9(1):23-6.
8. Charles NC, James EV. A study of the histopathologic pattern of Orbito-ocular disease in a tertiary hospital in Nigeria. Sahel Med J. 2014; 17:60-4
9. Chinda D, Samaila MO, Abah ER, Garba F, Rafindadi AL, Adamu A. A clinicopathological study of Orbito-ocular tumors at Ahmadu bello University Teaching Hospital, Shika-Zaria, Nigeria: A 5 year review. Clin Cancer Investig J. 2012;1:145-7
10. Odebode TO, Uyanne IA. Primary intra-orbital optic nerve sheath meningioma in an adolescent: a case report. Nigerian Journal of Opthalmology. 2006;14(1):35-37.
11. Akuaku-Dogbe E. Histopathological features of tumours of the orbit and adnexia seen in Korle-Bu teaching hospital. WAJM. 2012;31(1): 58-62.
12. Boulos PT, Dumont AS, Mandell JW, Jane JA. Meningiomas of the orbit: contemporary considerations. Neurosurg Focus. 2005; 10(5):1-10.
13. Furdova A, Babal P, Kobzova D. Orbital nerve sheath meningioma. Czech and Slovak Ophthalmology. 2018;74(1):23-30.
14. Cummings TJ, van der Valk, Optic nerve. In: Heegaard S, Grossniklaus H, eds. Eye Pathology; An illustrated guide. Heidelberg: Springer-Verlag. 2015:250-259