Case report: A case report of optic nerve glioma in a 5-year-old African girl

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Abstract

Background: Optic nerve gliomas are rare tumors that constitute less than 5% of all pediatric central nervous system tumors.
Case Presentation: We present the case of a 5-year-old girl with complaints of progressive protrusion of the right eye and poor vision of 3 years duration who was referred to the ophthalmologist. Ophthalmological evaluation of the right eye revealed tearing, no perception of light, inferior dystopia, severe proptosis, and a firm non-tender orbital mass palpable posterior to the eyeball. The cranial CT-scan was suggestive of a right optic nerve glioma. Histology showed Optic Glioma with extensive proliferation of meningothelial cells.
Conclusion: Optic gliomas can be associated with extensive proliferation of meningothelial cells that can histologically mimic a meningioma. It is important to be cautious about making a diagnosis of meningioma for tumors in locations that are also characteristic of optic nerve gliomas especially in appropriate clinical settings.

Rapport de cas : Un rapport de cas de gliome du nerf optique chez une fillette africaine de 5 ans Titre en cours - Gliome optique chez une fillette africaine

Résumé
Contexte de l'étude : Les gliomes du nerf optique sont des tumeurs rares qui constituent moins de 5 % de toutes les tumeurs pédiatriques du système nerveux central.
Présentation de cas : Nous présentons le cas d'une fillette de 5 ans présentant des plaintes de protrusion progressive de l'œil droit et une mauvaise vision depuis 3 ans qui a été référée à l'ophtalmologiste. L'évaluation ophtalmologique de l'œil droit a révélé un larmoiement, une absence de perception de la lumière, une dystopie inférieure, une exophtalmie sévère et une masse orbitaire ferme et non douloureuse, palpable en arrière du globe oculaire. Le scanner crânien était évocateur d'un gliome du nerf optique droit. L'histologie montrait un gliome optique avec une prolifération étendue de cellules méningothéliales.
Conclusion : Les gliomes optiques peuvent être associés à une prolifération étendue de cellules méningothéliales pouvant mimer histologiquement un méningiome. Il est important d'être prudent avant de poser un diagnostic de méningiome pour des tumeurs situées dans des localisations également caractéristiques des gliomes du nerf optique, en particulier dans des contextes cliniques appropriés.

INTRODUCTION

Optic nerve gliomas are rare tumors that constitute less than 5% of all pediatric central nervous system tumors (1,2). Most cases of optic nerve gliomas are diagnosed in the first decade of life (1). The tumour usually has a mild behavior and is less likely to be associated with vision loss in children. However, the tumour is more aggressive when found in adults (3). We have not seen a case of optic glioma in our facility in the last ten years. We present a case of optic glioma in a 5-year-old girl who was managed in our centre. This will help to improve the awareness of this tumour in our region of the world.

CASE PRESENTATION

A 5-year-old girl with complaints of painless progressive protrusion of the right eye and poor vision of 3 years duration was referred to the ophthalmologist. There was no history of gait abnormality, no history of seizure, and no history of loss of smell or taste. There was no history of first-degree relatives with similar conditions and no history of skin patches or swelling in any other part of the body.

Ophthalmological evaluation of the right eye revealed a visual acuity of no perception of light, inferior dystopia, severe proptosis, and a firm non-tender orbital mass palpable posterior to the eyeball. The overlying upper lid showed distended veins. There was associated tearing, diffuse hyperaemia, and a clear cornea. There was optic atrophy, optociliary shunts and a pale retina. The findings in the left eye were essentially normal.

The cranial CT-scan shown in figure 1 was suggestive of an intracranial cystic meningioma and right optic nerve glioma. There are no sufficient features required to classify the condition as Neurofibromatosis Type I.

She subsequently had a right enucleation with lateral canthotomy, excision of the orbital mass, orbital implant insertion, and a tarsorrhaphy on the 2nd day of admission. There were no postoperative complications. The resected tissue was sent for histopathologic examination which revealed a tissue with a distorted architecture due to the presence of uniform population of astrocytic cells with delicate hair-like processes imparting a fibrillary appearance to the background. The nuclei were round to oval with bland chromatin. Also present were extensive areas of reactive proliferation of meningothelial cells with associated hyalinized vessels. The resection margins contained tumour cells. The histopathologic diagnosis was consistent with Optic Nerve Glioma. Figures 2 and 3 show the gross and microscopic examination of the mass respectively. She was subsequently referred to another tertiary facility for adjuvant radiotherapy.

DISCUSSION

The index case is in her first decade of life which suggests her tumour is more likely to be associated with Neurofibromatosis type 1 (NF1). It is widely known that optic nerve gliomas in children are likely to be detected during the first decade of life. The patient's age is consistent with reports from other authors (1,4-6). We were unable to conclude that the patient had NF1 as we could not do genetic studies.

Many researchers have found a higher incidence of optic nerve gliomas in females. Our finding of this rare tumour in a young girl is not surprising as females have higher probability of developing the optic nerve glioma. Females with NF1 are also known to have a much higher chance of developing optic nerve gliomas (7,8). They are also more likely to have brain MRI for visual symptoms and more likely to require treatment for defects in vision than male patients (9).

Many authors have reported different locations of optic nerve gliomas. These include the intravitreal, orbital, and chiasma/hypothalamus (4,5). Studies done by many researchers show the intra-orbital mass as the most common form of presentation. Presumably, it will be difficult to differentiate gliomas involving optic radiation from other gliomas in children since the most common histologic variant for all intra-cerebral gliomas is Pilocytic astrocytoma.

Neurofibromatosis type 1 is a rare tumor suppressor syndrome known to be inherited in an autosomal dominant pattern (10,11). Hence, we expect the patient to have had a history suggestive of neurofibromatosis in the parents or siblings if this is NF1. It is possible that the disease has variable expressivity in the family since penetrance is commonly complete, or the information gathered is inadequate (6,12). It will be necessary to further evaluate the genetic characteristics of the patient to directly confirm her actual status.

Many reports of optic nerve gliomas show that when symptomatic, it commonly presents with protrusion of the eye due to the displacement effect of the mass on the eyeball. Our index case presented with inferior dystopia and severe proptosis. This is consistent with well-established facts. It has been reported that optic nerve glioma is one of the most common causes

of proptosis in children (13). It is important that family physicians and pediatricians' endeavor to detect proptosis for prompt evaluation by ophthalmologists for early diagnosis and appropriate management of the tumor.

Optic nerve gliomas can be associated with difficulty in closing the eye and excessive lacrimation as seen in the index patient. The pressure effect on the optic nerve can lead to vision loss as was seen in this patient. However, this vision loss and eye removal could have been avoided if the patient had presented earlier to the hospital for treatment.

Our patient had distended upper lid veins. This is not unusual as the pressure effect of the mass will reduce the venous drainage in the area involved by the tumour.

Microscopic examination of the tumour in the index patient showed reactive proliferation of meningothelial cells with associated hyalinized vessels. The proliferation of meningothelial cells was widespread and could mislead an unwary pathologist into making a diagnosis of meningioma. It is necessary to take histologic sections of many areas of the tumour to see the proliferating glial cells. It is important to be cautious about making a diagnosis of meningioma for tumors in locations also characteristic of optic nerve gliomas especially in appropriate clinical settings. Optic nerve sheath meningiomas are most found in older patients (14).

CONCLUSION

It is important that family physicians and paediatricians' endeavor to detect proptosis for prompt evaluation by ophthalmologists for early diagnosis and appropriate management of the Optic nerve gliomas. The tumour can be associated with vision loss, difficulty in closing the eye, and excessive lacrimation as seen in our patient. Optic gliomas can be associated with extensive proliferation of meningothelial cells that can histologically mimic a meningioma. It is important to be cautious about making a diagnosis of meningioma for tumors in locations that are also characteristic of optic nerve gliomas especially in appropriate clinical settings.

Ethical Statement: We obtained informed consent from the patient according to the guidelines of Ethical and Research Committee of Obafemi Awolowo University Teaching Hospitals Complex. A signed consent form is available on request. We followed the ethical principles outlined in the declaration of Helsinki in preparation of this manuscript.

Conflict of Interest: We declare no conflict of interest.

Availability of data and materials: The tissue blocks are available for future use.

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Authors' contributions: OOO, OCC, and EEE reported the pathologic findings. ABA managed the patient and reported the clinical findings and operation notes. All authors contributed to the discussion and the final write up. All authors approved the final write up.

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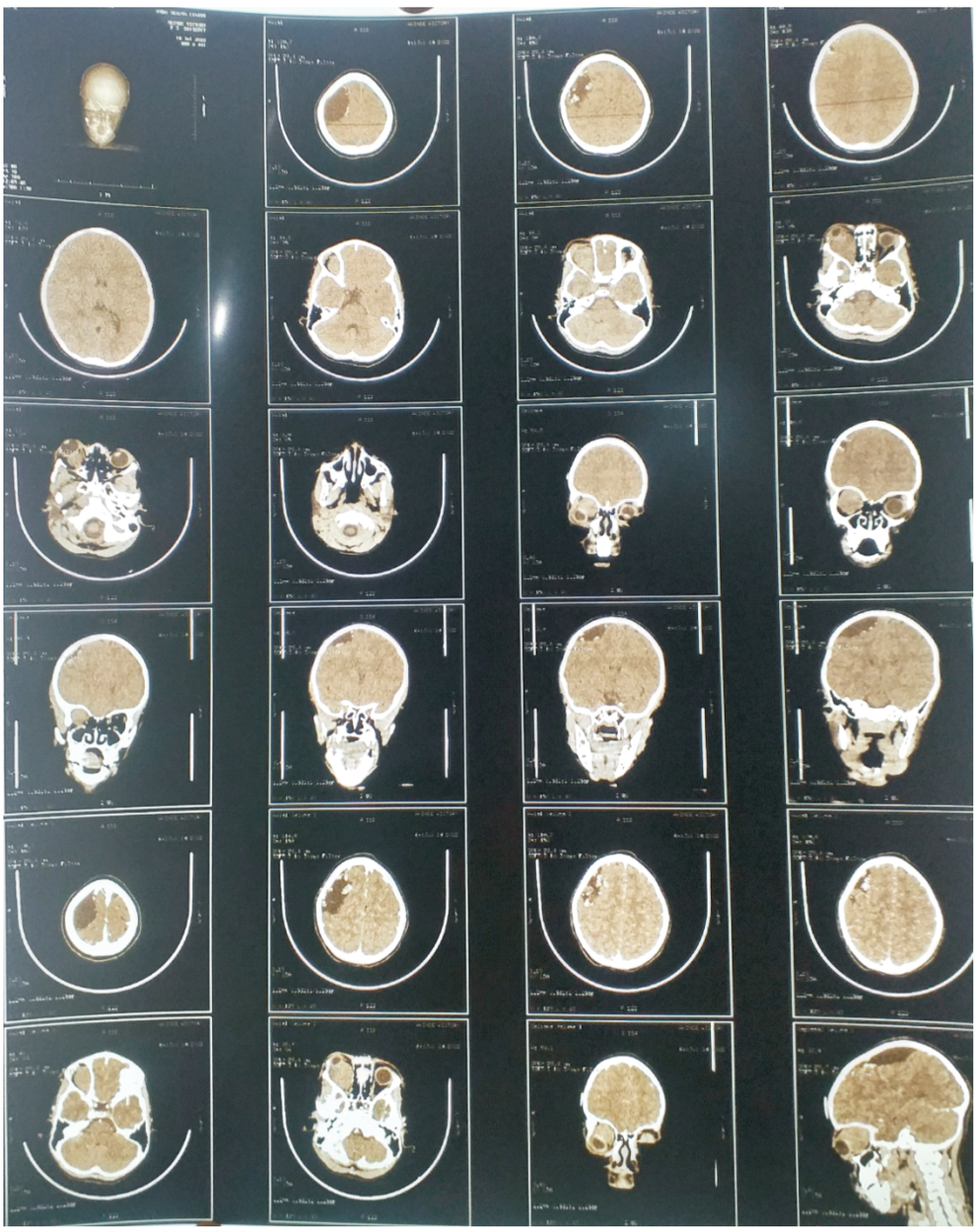

Figure 1 Cranial CT-Scan
The CT-Scan (axial, coronal and sagittal views) shows a right sided orbital mass and an intracranial space occupying lesion.

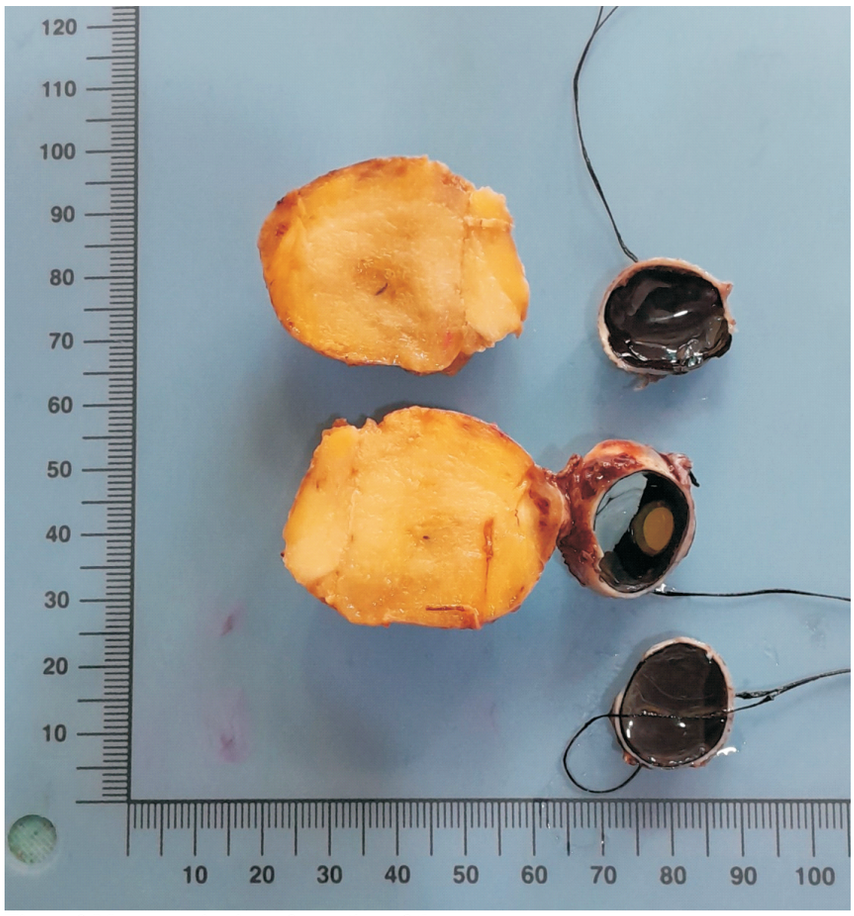

Figure 2 Macroscopic view of a section of the mass
The optic nerve is enlarged by an encapsulated mass extending through its entire length. It measures 4.1x3.5x2.7cm. The cut surface shows pale white to greyish white solid tissue.

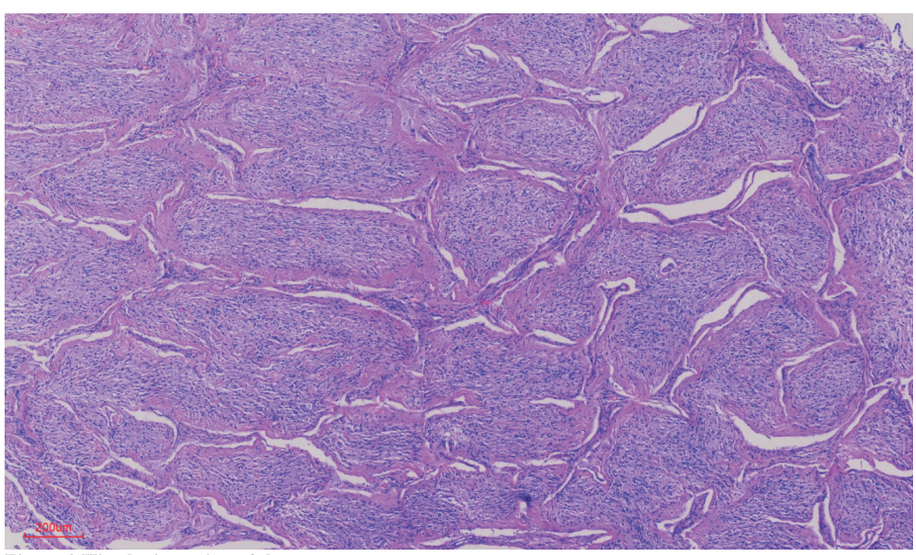

Figure 3 Histologic section of the mass
There is infiltration of the optic nerve by neoplastic astrocytic cells. Other areas showed features of pilocytic astrocytoma.

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