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Clinical Record

Adult nasal glioma presenting with visual loss

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Abstract

Objectives: We report a rare case of a nasal glioma found incidentally in an adult, presenting with visual loss, optic nerve oedema and proptosis.

Case report: A 41-year-old woman presented with bilateral proptosis, impairment in visual acuity (6/60 bilaterally) and loss of colour vision. Computed tomography and magnetic resonance imaging showed proptosis, bilateral optic nerve swelling and a heterogeneous mass occupying the left nasal cavity and extending through a skull base defect into the anterior cranial fossa. Biopsy confirmed a nasal glioma. Treatment with intravenous dexamethasone resolved the proptosis, and the patient's visual acuity recovered to 6/9 bilaterally. At the multidisciplinary team meeting, it was felt that the nasal glioma probably represented an incidental finding and was not directly responsible for the patient's proptosis and transient visual loss.

Conclusion: To our knowledge, this is the first report in the English language literature of adult nasal glioma presenting with visual loss. The management of nasal gliomas in adults is contentious and the relevant literature is reviewed. This case was managed conservatively with regular follow up.

Key words: Nasal Glioma; Nasal Glial Heterotopia; Proptosis; Visual Disturbance

Introduction

Nasal glioma is a benign congenital tumour of neurogenic origin. The term nasal glioma is a misnomer, as it implies a true neoplasm with malignant potential. More accurately, the tumour should be termed a 'benign congenital nasal neuroectodermal tumour' or 'nasal glial heterotopia'.[1] It was first described by Reid in 1852,[2] and was termed 'nasal glioma' by Schmidt in 1900.[3] This term found its way into widespread clinical usage, and continues to be used.

We present a rare case of nasal glioma presenting in adulthood with bilateral optic nerve oedema, proptosis and visual disturbance. The patient also had white matter hyperintensity in the left frontal lobe on magnetic resonance imaging (MRI). The clinical manifestations, diagnosis and treatment of this clinical entity are discussed, and the relevant literature is reviewed.

Case report

A 41-year-old Afro-Caribbean woman presented with a one-week history of blurred vision. The patient had no antecedent history of meningism.

Initial examination revealed marked bilateral proptosis with associated severe impairment of visual acuity (6/60 bilaterally) and loss of colour vision. Fundoscopy did not show papilloedema. The patient showed no evidence of congenital deformity. The rest of the neurological and general medical examination was unremarkable. There was no history of hyposmia.

The only abnormality on blood testing was elevation of the C-reactive protein level, to 103 mg/L. Thyroid function test results were normal, and the patient had a negative autoantibody screen. A computed tomography (CT) scan of her orbits showed proptosis and bilateral optic nerve swelling (Figure 1). In addition, a heterogeneous mass was noted to occupy the left nasal cavity and to extend through a skull base defect into the anterior cranial fossa (Figure 2). No obvious dural envelope was seen.

The patient was immediately commenced on a course of intravenous dexamethasone (4 mg thrice daily), and an ophthalmology opinion was sought. Her proptosis resolved within four days, and her visual acuity improved to 6/9 bilaterally.

Further, specific questioning revealed a preceding three-month history of left-sided nasal obstruction and nasal discharge.

Magnetic resonance imaging was performed to further evaluate the nasal mass. This confirmed the earlier CT findings (Figure 3) and also revealed white matter hyperintensity in the left frontal lobe (Figure 4).

Rigid endoscopic examination revealed a pale, non-pulsatile mass obstructing the left nasal cavity emanating from above the middle turbinate. The mass was not compressible and did not transilluminate or change in size during Valsalva's manoeuvre. The Furstenburg test (i.e. compression of the ipsilateral internal jugular vein and observation for any increase in the size of the mass) was negative. There was no evidence of cerebrospinal fluid (CSF) leakage pre-operatively or during peri-operative biopsy.

Histological examination of the biopsy specimen showed respiratory-type mucosa with lobules of mature central nervous system parenchymal tissue (with neuronal and

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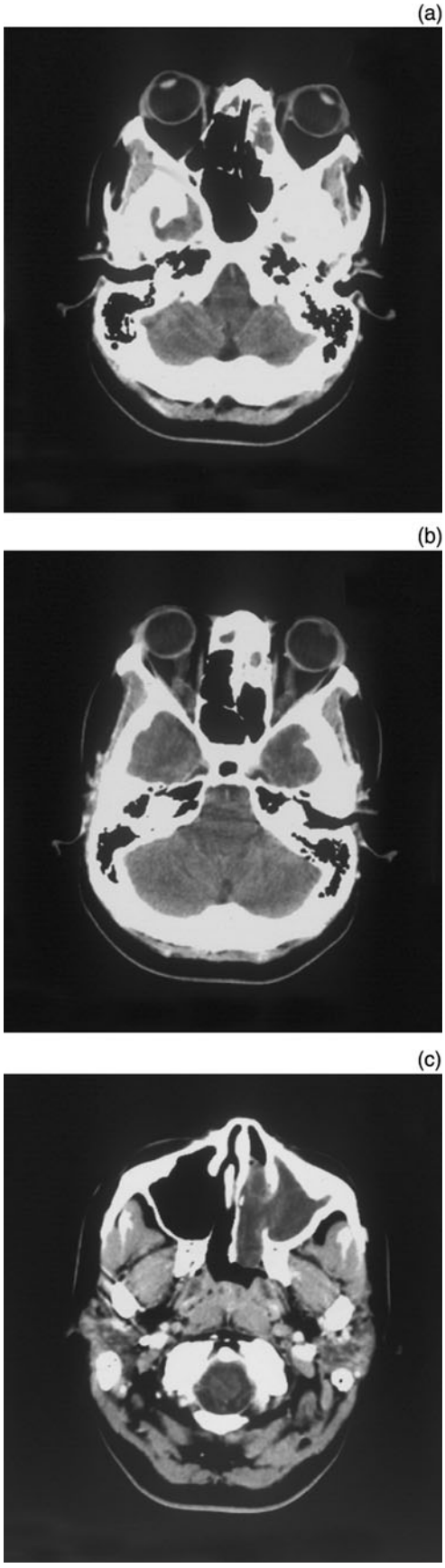

FIG. 1

Axial, non-enhanced computed tomography scans showing (a) bilateral proptosis, (b) optic nerve swelling and (c) a mass in the left nasal cavity.

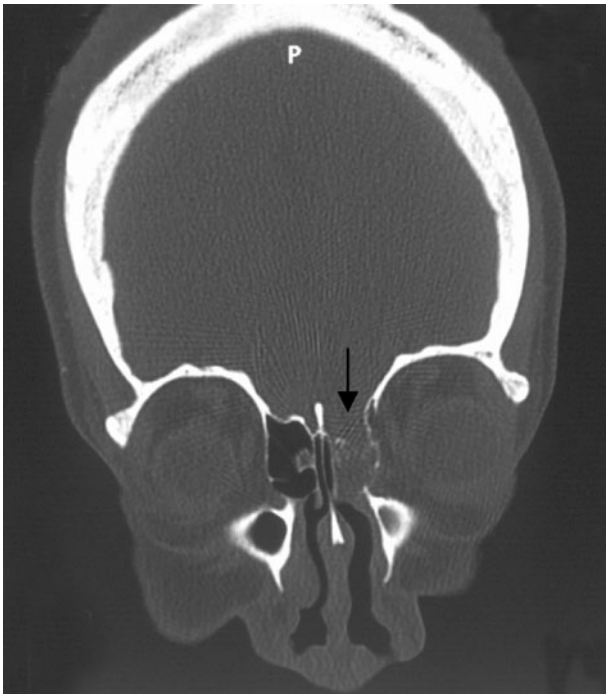

FIG. 2

Coronal, bone window computed tomography scan showing a skull base defect in the left cribriform plate (arrow) and a soft tissue mass extending between the left nasal cavity and the anterior cranial fossa.

glial components) separated by loose connective tissue and chronic inflammatory cell infiltrate (Figures 5 and 6). The glial component showed typical features of a reactive gliosis, with enlarged astrocytes with thick multipolar processes, set within a dense fibrillary matrix. There was no nuclear atypia, mitotic activity or other features of neoplasia within the glial component. The fragments of brain parenchyma were not surrounded by a meningeal covering. Within the connective tissue, there were pigmented cells with a dendritic morphology, in keeping with melanocytes.

Immunohistochemistry using the proliferation marker Ki67 identified inflammatory cells but did not demonstrate any proliferating glial cells (Figure 7). This was in keeping with the benign nature of the lesion, which was identified as heterotopic neuroglial tissue, compatible with a diagnosis of nasal glioma.

Post-operatively, the patient had no CSF leakage and her constellation of symptoms quickly settled. Her case was discussed at the multidisciplinary team meeting; on balance, it was felt that the nasal glioma probably represented an incidental finding and was not directly responsible for the patient's proptosis and transient visual loss. For this reason, and at the patient's request, a conservative approach was adopted.

The patient was discharged home without headache, visual disturbance, nausea or vomiting, and was fully orientated with a Glasgow Coma Scale score of 15/15. She continued to be reviewed as an out-patient, and remained asymptomatic at 18 month follow up.

Discussion

Nasal gliomas form part of a larger group of congenital midline masses which include encephalocoeles and dermoids. The incidence of congenital nasal masses is one in every 20 000 to 40 000 live births.[4] Approximately 250

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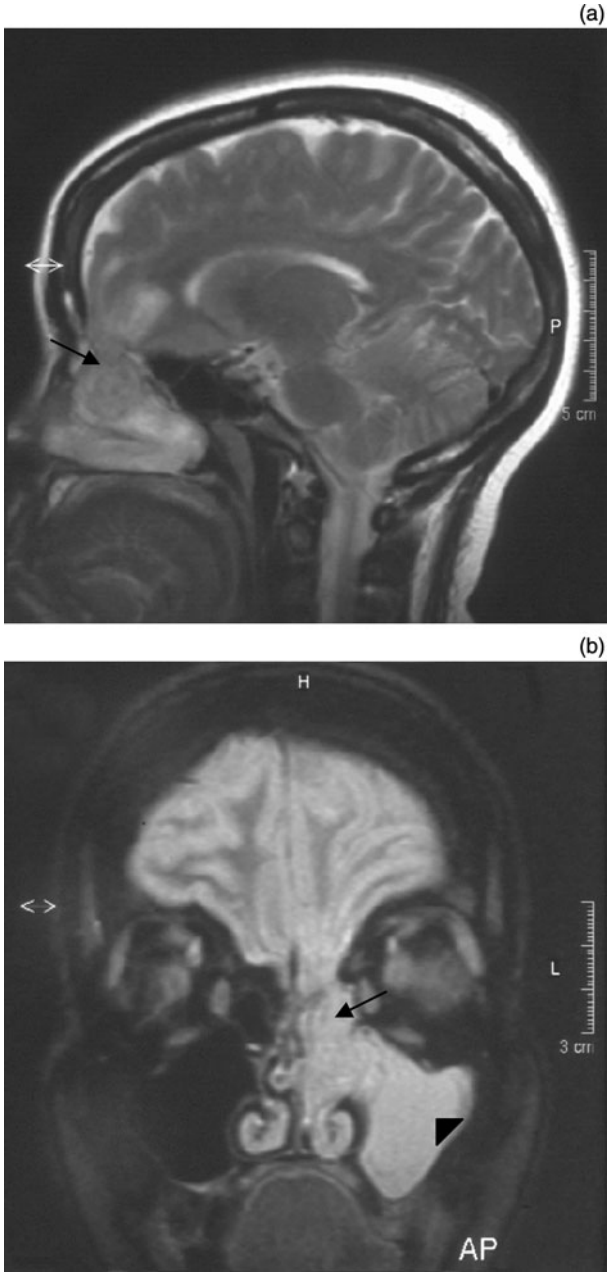

FIG. 3

(a) Sagittal, T2-weighted magnetic resonance imaging (MRI) scans (Echo time (TE) = 120 ms, Repetition time (TR) = 1942 ms) and (b) coronal, T1-weighted Short tau inversion recovery (STIR) MRI scans (TE = 18 ms, TR = 1238 ms). There is opacification of the left maxillary (arrowhead) and ethmoidal sinuses, and a soft tissue density within the left nasal cavity (arrow). This occupies the middle and superior meatus and extends through the cribriform plate and fovea ethmoidalis into the anterior cranial fossa, abutting the left frontal lobe. P = posterior; L = left; AP = anteroposterior

cases of nasal glioma have been reported.[5] The tumour has a male-to-female ratio of 3:2, and there is no recognised familial tendency or association with other developmental anomalies.[6,7]

It is widely accepted that these lesions result from aberrations in normal embryonic development.[4] In particular, nasal gliomas are thought to represent basal or sincipital encephalocoeles that have lost their intracranial meningeal connection.[6] Unsurprisingly, differentiating nasal gliomas

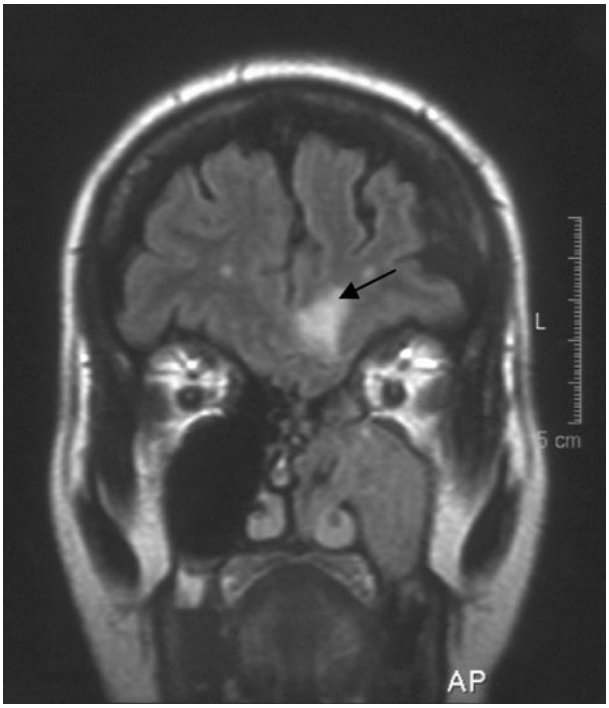

FIG. 4

Coronal, T2-weighted Fluid attenuated inversion recovery (FLAIR) magnetic resonance imaging scan (TE = 150 ms, TR = 7000 ms) showing white matter hyperintensity in the left frontal lobe (arrow). L = left; AP = anteroposterior

from encephalocoeles is difficult, and is based on clinical and radiological features. Encephalocoeles have an intracranial connection via a bony defect in the skull base and are covered by meninges. Nasal gliomas can have a fibrous stalk connecting them to the dura but not breaching it.[1] The only other differential diagnosis for mature neuroglial tissue in the nasal cavity is a teratoma. This would however contain tissue from all three germ cell layers, and could be excluded by microscopic examination of the entire specimen.

Sixty per cent of nasal gliomas manifest extranasally, 30 per cent are solely intranasal and 10 per cent are mixed.[6]

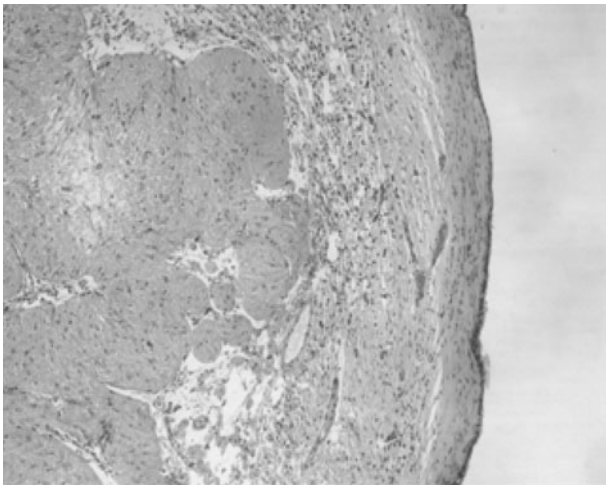

FIG. 5

Photomicrograph of the nasal glioma showing nasal mucosa with underlying lobulated neural tissue. (H&E; ×40)

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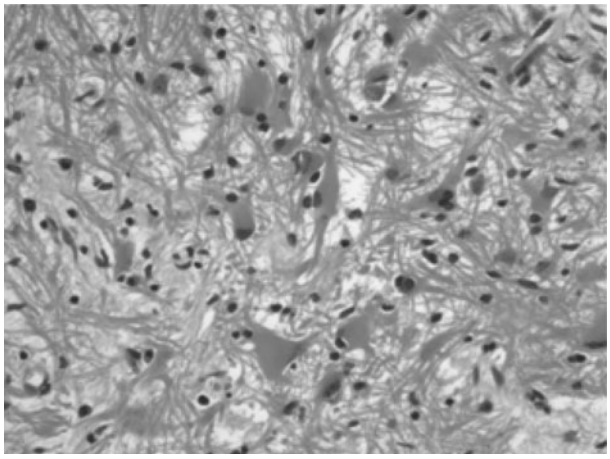

FIG. 6
Photomicrograph of the nasal glioma showing astrocytes with coarse multipolar processes and abundant eosinophilic cytoplasm. (H&E; ×200)

The base of the intranasal glioma generally arises from the lateral nasal wall at or above the level of the middle turbinate, and occasionally from the nasal septum.[8] A minority of cases, 10–25 per cent, may have a fibrous stalk extending toward the skull base, with an underlying bony defect.[4,8]

The diagnosis of nasal glioma is facilitated by the appropriate use of cross-sectional imaging studies. Computed tomography is useful in visualising bony defects in the anterior skull base,[9] while MRI provides complementary information regarding the fluid or soft tissue characteristics of the mass.[10] MRI also provides a three-dimensional view and can determine the presence or absence of any intracranial extension.[9] However, pre-operative radiological evaluation should not lead to a false sense of security if no bony defect is demonstrated.[8]

On macroscopic examination, nasal gliomas are generally pale, firm, noncompressible masses that do not transilluminate or expand with the Valsalva manoeuvre or crying. The Furstenberg test, performed by compressing the ipsilateral internal jugular vein and observing the mass for expansion or pulsation, should be negative.[9] A positive result may be evident in cases of encephalocoele which are directly connected with the subarachnoid space and may contain CSF.

Histologically, nasal 'gliomas' are composed of unencapsulated collections of astrocytic, eosinophilic neuroglial cells in a connective tissue matrix.[4] The neural tissue within is thought to be non-functioning, as there have been no reports of any neurological deficit resulting from surgical extirpation. Mitoses are characteristically absent.[11] Any possible intracranial connection is usually fibrous and does not breach the dura. Nasal gliomas have no meningeal envelope and are covered only by skin or nasal respiratory mucosa.[7] Secondary changes of fibrogliosis or gemistiocytic alteration of glial cells are often seen,[12] probably due to compression and infarction of neural tissue. This can make nasal gliomas histologically indistinguishable from encephalocoeles; in this case, differentiation is possible only based on radiological or surgical evidence.[13,14] In the case above, the absence of a meningeal covering layer (either macroscopically or histologically) and the failure to provoke a CSF leak during deep biopsy suggested that the lesion was a nasal glioma rather than an encephalocoele.

- **The term 'nasal glioma' is a misnomer as it implies a neoplastic condition with malignant potential, which it is not; the term is however widely used clinically**
- **The vast majority of nasal gliomas present in early childhood with nasal obstruction and feeding difficulties**
- **A rare case of an incidental nasal glioma in an adult is reported**
- **This patient presented with optic nerve oedema, proptosis and visual loss (a previously unreported presentation for this tumour), presumed to be inflammatory as symptoms improved with intravenous steroids**
- **Although nasal gliomas can be excised endoscopically, their management in adults is contentious. Due to patient choice and the fact that the finding was incidental, this case was managed conservatively with regular follow up**

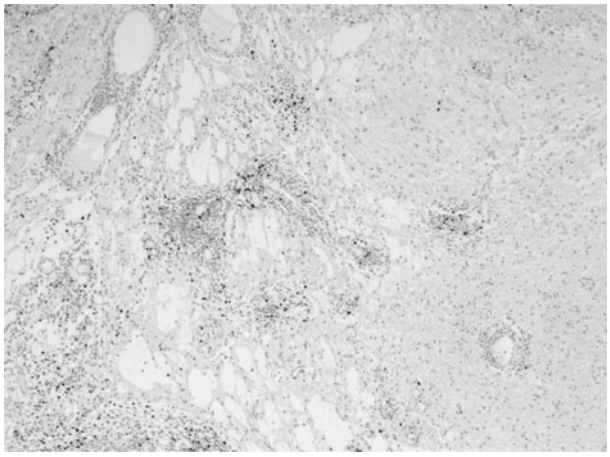

FIG. 7
Photomicrograph of the nasal glioma, immunohistochemically stained with the Ki67 proliferation marker, identifying inflammatory cells but no proliferating glial cells. (×20)

Intranasal gliomas predominantly occur in the paediatric population, causing nasal obstruction and difficulty with feeding.[9] Only nine cases have been reported in adults.[6,11,12,15–20] Most of these presented with non-specific nasal symptoms; two presented with meningitis. One presented with bilateral total blindness due to extension of the 'glioma' into the orbits.[12] It is difficult to understand why our patient had optic nerve swelling and proptosis, given the absence of any retro-orbital extension of the nasal glioma. The fact that she improved on commencing dexamethasone implies an inflammatory process, but no specific diagnosis could be reached by blood testing, including autoantibody analysis. She refused a lumbar puncture, and therefore a secondary diagnosis of multiple sclerosis cannot be completely excluded. However, the white matter hypersensitivity in the left frontal lobe, seen on MRI, is not typical of multiple sclerosis, and probably represents vasogenic oedema secondary to the adjacent glioma extending through the cribriform plate. Alternatively, it may represent a local area of cytotoxic oedema secondary to local ischaemia caused by compression from the glioma.

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In children with nasal glioma, the general consensus on management recommends complete extirpation via a functional surgical approach, in order to prevent the mutilating effects of this lesion on the growing facial bones.[17] Whilst early reports suggested the use of incisional biopsy,[21] more recent articles have advocated the avoidance of such biopsies, due to the increased risk of CSF leak and meningitis.[9,17,18] In terms of surgical technique, the endoscopic approach can be used successfully for the treatment of nasal glioma.[9] Although this approach offers an aesthetic advantage and quicker recovery, a craniofacial approach may be required if there is significant intracranial connection. Whatever the approach, the risk of recurrence following inadequate primary excision is between 4 and 10 per cent.[6]

The management of nasal gliomas in adults is more contentious. The diagnosis usually comes as a histological surprise following incisional biopsy for a presumed nasal tumour. Nasal gliomas are relatively radioresistant,[12] and radiotherapy is therefore reserved for inoperable cases or recurrences after surgical excision. There is controversy as to whether adult cases should undergo surgery or be managed with a more conservative approach, especially in asymptomatic individuals. Nasal gliomas have a slow growth rate approximating that of the surrounding tissue,[7] and malignant degeneration has not been described. In view of this and, more importantly, the patient's wishes, a conservative policy was adopted in the present case.

Conclusion

The management of nasal gliomas in adults is contentious. We report a rare case of an incidental nasal glioma found in an adult, which was managed conservatively. The patient presented with optic nerve oedema, proptosis and visual loss, which has not previously been reported for this condition. These were presumed to be inflammatory, as her symptoms improved on commencing intravenous steroids.

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