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Nasal glioma

鼻膠質瘤

Nasal gliomas are uncommon congenital lesions arising from abnormal embryonic development. Clinically, these masses are firm and incompressible. Histologically, they are made up of astrocytes and neuroglial cells, embedded in fibrous and vascular connective tissue. Proper management of a nasal glioma requires a multidisciplinary approach including an otorhinolaryngologist, radiologist, and neurosurgeon. Radiological investigations such as computed tomography or magnetic resonance imaging should be performed to exclude intracranial extension. The mainstay of treatment is conservative surgical excision because nasal gliomas are slow-growing, rarely recurrent, and have no malignant potential. We report one case of nasal glioma in a Chinese infant. He had an uncomplicated surgical intervention with a good cosmetic result. A review of the clinical features of and diagnostic approach to nasal gliomas is also presented.

鼻膠質瘤是胚胎非正常發展而引致的罕見先天性病症。臨床上，此腫塊堅硬而不會變形；組織學上，此腫瘤由星形膠質細胞和神經膠質細胞組成，外圍包裹著纖維性和血管連接的組織。鼻膠質瘤需要耳鼻喉科醫生、放射治療師和神經外科醫生的跨部門治療，以放射檢查如電腦掃描或磁力共振造影排除顱內擴張的可能。由於鼻膠質瘤生長緩慢、極少復發且不是惡性瘤，因此最普遍是以傳統的外科手術切除。本文報告一名患有鼻膠質瘤的華籍嬰兒，在手術後康復並且復容效果良好。本文亦檢討鼻膠質瘤的臨床特徵和診斷方法。

Introduction

A nasofrontal mass in a newborn is uncommon and there can be many differential diagnoses. An accurate diagnosis permits proper management and prevents potentially life-threatening intracranial complications. Nasal gliomas are one form of the congenital midline nasal masses that usually present at birth. They are rare, benign, congenital tumours, which arise from abnormal embryonic development. Around 15 to 20% of nasal gliomas have a fibrous stalk connecting to the central nervous system. Severe complications such as meningitis or a brain abscess can be avoided if the lesions are removed at an early stage. We report a case of a Chinese infant with a nasal glioma, and discuss the clinical presentation, diagnosis, and management of such a tumour.

Case report

In February 2002, a 51-day-old boy was referred to our department for the evaluation of a swelling over his nasal bridge. The swelling had been present since birth but did not cause any nasal obstruction or epistaxis. The baby also had a cleft lip and palate. He was born at full-term and had a normal vaginal delivery. His elder sibling had no congenital abnormalities and the family history was unremarkable.

On physical examination, the baby had a dysmorphic facial expression with an upper cleft lip and palate, and a 3 x 3 cm superficial mass over his nasal bridge (Fig 1). It was purple in colour and firm in consistency. It was neither tender nor pulsatile. There was no intranasal mass. Computed tomography (CT) and magnetic resonance imaging (MRI) of the brain showed a superficial midline nasal mass with no direct connection to the brain (Fig 2).

Key words:
Glioma;
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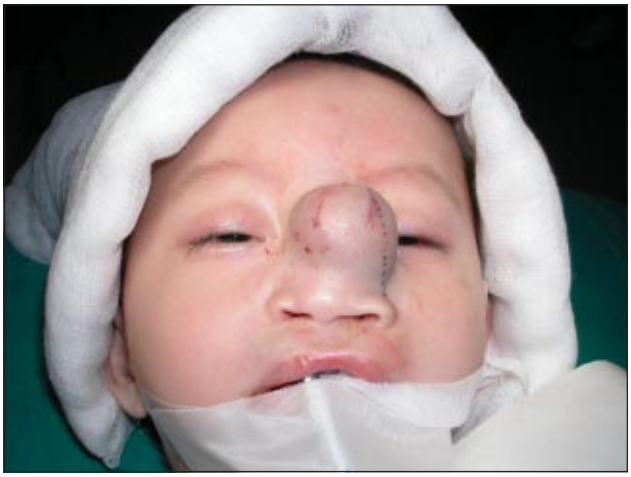

Fig 1. Preoperative view of the external glioma with skin markings for the incision

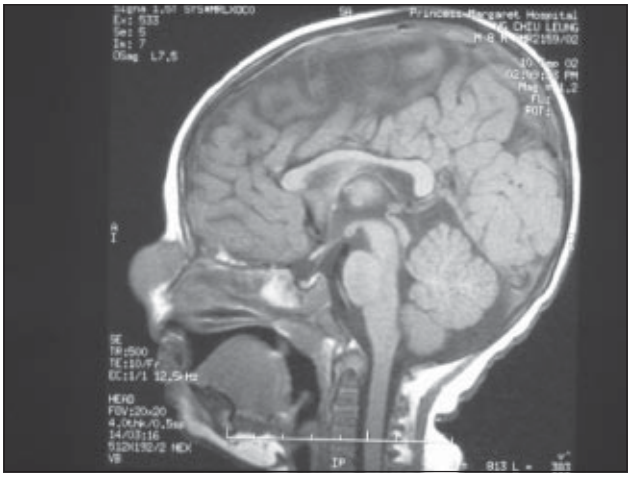

Fig 2. Magnetic resonance imaging demonstrating a soft-tissue nasal mass with no intracranial connection

The baby's cleft lip was repaired at 3 months of age whereas the nasal mass was removed at 1 year. The tumour was excised externally under general anaesthesia, using a left lateral elliptical skin incision. The margin was taken down to the cartilaginous pyramid. The excised mass weighed 8.1 g and measured 3 x 2.5 x 2 cm. Grossly, it appeared as a lobulated, well-circumscribed, and unencapsulated whitish pink soft mass. Microscopic sections demonstrated the presence of glial tissue with a fibrillary background and interspersed fibrovascular stroma. No meningeal or dural tissue was identified. The glial nature of the tumour was confirmed by a positive reaction for S100 protein and glial fibrillary acidic protein (GFAP). The definitive diagnosis was nasal glioma. The patient recovered well after surgery. His cleft palate was repaired when he was 18 months old. Follow-ups performed over the last 3 years using brain MRI showed no residual parts or recurrence of the glioma. The postoperative cosmetic result was satisfactory.

Discussion

Congenital midline nasal masses are rare anomalies, with an incidence of one in 20 000 to 40 000 live births.[1-4] The common forms are dermoid cysts, nasal gliomas, encephalocoeles, and haemangiomas.

Schmidt was the first scientist to describe the comprehensive nature of the nasal glioma in 1900. However, the term he used is a misnomer.[3,5] Nasal gliomas are not true neoplasms; they originate from ectopic glial tissue left extracranially following abnormal closure of the nasal and frontal bone during embryonic development.[1-4,6] Therefore, some authors recommend using the term 'glial heterotopia' instead.[3] Around 15 to 20% of nasal gliomas have a fibrous stalk connecting them to the subarachnoid space of the brain. Sometimes, this fibrous connection may be associated with severe complications like cerebrospinal fluid leakage or meningitis.[1,7]

Nasal gliomas generally present at birth, rarely in adults, as a mass without associated nasal symptoms. They have a 3:1 male predominance, with no familial or hereditary predisposition and no malignant potential.[3,5,7] The tumour growth rate is consistent with the patient's body growth.[7] About 60% of gliomas are extranasal, 30% are intranasal, and 10% are mixed lesions.[5]

Extranasal gliomas are firm, incompressible masses that often occur along the nasomaxillary suture or near the glabella. The overlying skin may have telangiectasia, and they may easily be confused with haemangiomas. The nasal bridge may be broadened and the space between the eyes may be widened.[1,4] Intranasal gliomas usually present as a pale mass with septal deviation or nasal obstruction. They often arise from the lateral nasal wall or, less often, from the nasal septum. In all gliomas, pulsation or expansion of the lesions is absent during crying, coughing, straining or even compressing the jugular vein (Furstenberg's test). Furstenberg's test is positive in encephalocoeles, owing to the intracranial connection.[3]

Histologically, nasal gliomas are composed of astrocytes and neuroglial cells, embedded in fibrous and vascular connective tissue.[5] They have no true capsule and mitosis is rarely noted. Multinucleated or gemistocytic astrocytes may be present but it is rare to find neurons. The presence of abundant neurons raises the possibility of an encephalocoele. The glial nature of the cells can be further confirmed by immunohistochemical demonstration of S100 protein and GFAP. These two proteins can identify neurological cells with high specificity, and help to distinguish nasal gliomas from other tumours such as meningiomas and granular cell tumours.[5,8] In our case, both S100 and GFAP staining were positive. Hence, a definitive diagnosis of nasal glioma was made.

Proper management of nasal gliomas requires a multidisciplinary approach, including an oto-rhinolaryngologist, radiologist, and neurosurgeon.[3] A complete medical history review and a full physical examination should be performed for patients with a nasal glioma. Any other congenital abnormalities should be searched for. Our patient had both a nasal glioma and a cleft lip and palate; no clear association between the two abnormalities was found in the literature. Nasal endoscopy should be performed to delineate the site and extent of any intranasal mass. When a nasal glioma is suspected, no invasive procedure nor surgery should be performed until an intracranial connection has been excluded using CT scanning or MRI, due to the risk of meningitis or cerebrospinal fluid leakage.[5] The preferred form of imaging is MRI. Computed tomography is good for delineating bony abnormalities but not for detecting an intracranial component in nasal gliomas. Magnetic resonance imaging is better for delineating soft tissue details, including any intracranial connection. It also minimises the level of exposure to ionising radiation, particularly in infants.[4] A neurosurgeon should be consulted whenever the tumour communicates with the brain, as a craniotomy may be necessary.

Surgical excision is the mainstay of treatment and is required for a definitive histopathological diagnosis. A thorough preoperative imaging study must be performed prior to any attempt at removal of the glioma. The extent of the surgery is dictated by the exact size, location, and contents of the lesion.[4] Early surgical intervention is also important, since a delay can lead to severe complications such as meningitis or a brain abscess; or cosmetic problems such as distortion of the septum and nasal bone.[6] For pure extranasal gliomas, an external incision using either a vertical elliptical midline incision or a horizontal incision over the dorsum of the nose can yield equally good cosmetic results.[5] Some authors recommend a conservative and cosmetic incision using an external rhinoplasty approach, because nasal gliomas are benign and rarely recurrent.[3] For pure intranasal gliomas, a transnasal endoscopic approach is recommended for complete removal of the intranasal mass with no postoperative facial deformity.[3] In our case, the 3 x 2 cm extranasal glioma was excised via a lateral elliptical skin incision, and with a good cosmetic result.

Conclusion

A nasofrontal mass in an infant is a rare anomaly and may result in severe intracranial complications. When nasal gliomas or encephalocoeles are suspected, appropriate cross-sectional imaging such as CT or MRI of the brain must be done to exclude any intracranial connection before performing an invasive procedure. Conservative surgical excision is the mainstay of the management of an extranasal glioma, as it is slow-growing, rarely recurrent, and has no malignant potential.

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