CASE REPORT

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Nasopharyngeal glioma in a new-born girl*

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SUMMARY

Nasal gliomas are uncommon tumours of neurogenic origin that occur sporadically. They are diagnosed with MRI and a peroperative biopsy, and surgery is the treatment of choice. Most of the gliomas emerge from the nasal cavity, but only a few cases of nasopharyngeal gliomas have been reported.
We present one case of a nasopharyngeal glioma and two cases of nasal gliomas.

Key words: nasopharyngeal, nasal, glioma, infant

INTRODUCTION

Nasal gliomas (NG) are rare tumours of neurogenic origin, that are non-neoplastic, with limited growth potential and no similarity to the central nervous system gliomas. The nasal glioma is considered to be an encephalocele that in the fetal development has lost its cranial connection (Burckhardt et al., 1999), although 15 - 20% of NG have a fibrous connection, a stalk, to the dura at the time of diagnosis (Karma et al., 1977). They are found in both sexes, are not familial and most are diagnosed in newborns or infants (Juhlin et al., 1989). However a handfull have been diagnosed in adulthood (Heacock et al., 1992; Pasquini et al., 1998).

Clinically, nasal gliomas present as a nasal obstruction or nasal discharge. Extranasal gliomas found at the dorsum of the nose, usually cause a cosmetic deformity and intranasal gliomas cause upper airway obstruction and respiratory distress. The treatment of choice is surgical excision and few recurrences are reported.

Nasal glioma was first described in the literature in 1900 by Schmidt (Harrison et al., 1993), and by the year 2000 more than 250 cases have been reported in the literature. We here report one case of a nasal glioma extending into the nasopharynx in a newborn girl, and two cases of strictly nasal gliomas.

CASE REPORTS

Our department serves one and a half million inhabitants, in the western part of Sweden. In this area about 15 000 children are born every year. In three consecutive years, three newborn girls have been diagnosed with a nasal glioma at our department, and they have all undergone the same surgical treatment, excision by a lateral rhinotomy, by the same surgeon (BP) (Figure 1).

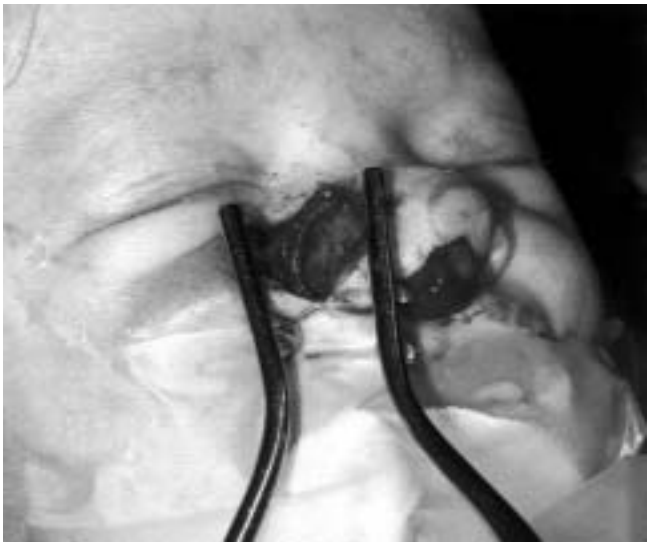

Figure 1. Case three (AO). When a lateral rhinotomy is performed in infants a three millimeter broad bone bridge of the infraorbital margo has to be saved, as well as the suture between the nasal bone and the frontal process of the maxillary bone, in order not to interfere with growth zones. Care is taken not to injure the lacrimal duct and the infraorbital nerve.
The glioma is protruding through the right nostril.

Case one (AG)

A female infant that shortly after birth showed signs of increasing respiratory distress was brought to the ICU at the age of three days, and intubated. A MRI was done, showing a bone defect in the midline of the sphenoidal bone and from this defect a stalk led to a huge process (3 x 4,5 cm) filling the nasopharynx and protruding into the nasal cavity (Figure 2). No brain parenchyme was detected in the nasophayngeal mass, and the primary diagnosis was nasal glioma. When the girl was

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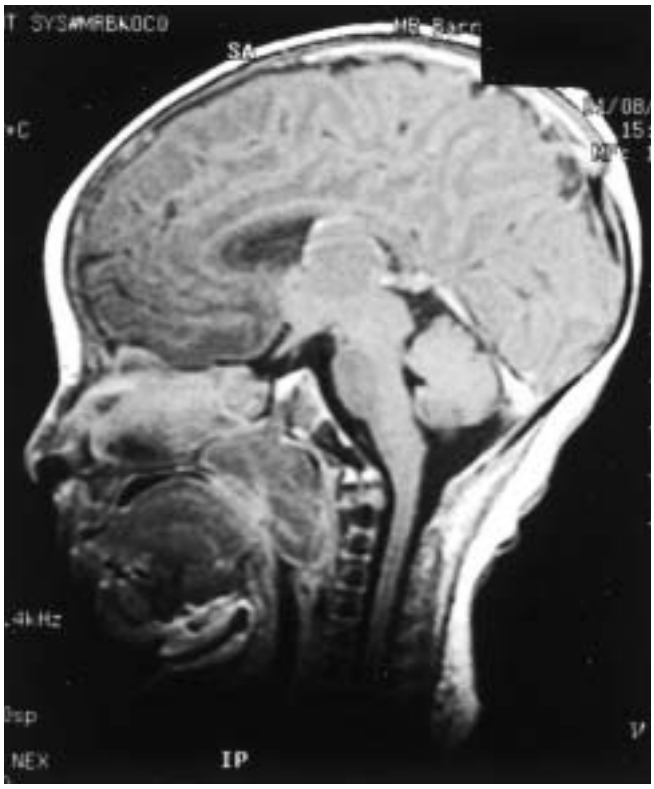

Figure 2. Case one (AG). MRI one week after birth shows how one part of the nasal glioma is protruding behind the soft palate and the tongue and the other part is filling most of the nasal cavity.

one week of age a surgical excision of the process, via a lateral rhinotomy, was done. The glioma-stalk was left, due to its anatomical closeness to the pituitary gland. No postoperative cerebrospinal fluid leakage was noted and after one year the patient has had no recurrence.

Case two (FE)

A female infant was admitted immediatly after birth because of bleeding from the right nostril, which appeared to be blocked. MRI was performed showing an intranasal mass (3 x 1 cm) filling the whole right nasal cavity and was bulging into the nasopharynx. No communication between the mass and the brain was found, nor did the mass contain any brain parenchyme. The patient was operated at two months of age, and via a lateral rhinotomy the glioma was excised. A stalk, leading up to lamina cribrosa, was found and removed. No postoperative cerebrospinal fluid leakage was noted and after 3,5 years the patient has had no recurrence.

Case three (AO)

A female infant, born with a protruding purple process from the right nostril. The mass extended about one cm from the vestibulum. The girl had difficulties breathing. MRI was done showing a 1 x 3 cm strictly intranasal mass with no apparent connection with crista galli, or the brain. At the age of six weeks the girl was operated, and via a lateral rhinotomy the glioma, attached to both the nasal septum and the medial side of the nasal lateral wall was excised. No stalk was found, no postoperative cerebrospinal fluid leakage was noted and after 3 years the patient has had no recurrence

Pathology

In all three cases histological examination of the process showed that it was covered by respiratory mucosa and in the stroma area, small glial cells and scattered atypical, hyperchromatic cells were found. Immunohistochemistry for GFAP (glial fibrillary acidic protein; Figure 4a) and S-100 (Figure 4b) were positive, verifying that the lesions were nasal gliomas. CD-44 and NSE (neuron-specific enolase) also gave a positive reaction. Ki-67 showed scattered proliferating cells. NFP (neurofilament protein) and Synaptophysin were both positive. Transmission electron microscopy verified the presence of glial cells in case one (Figure 3).

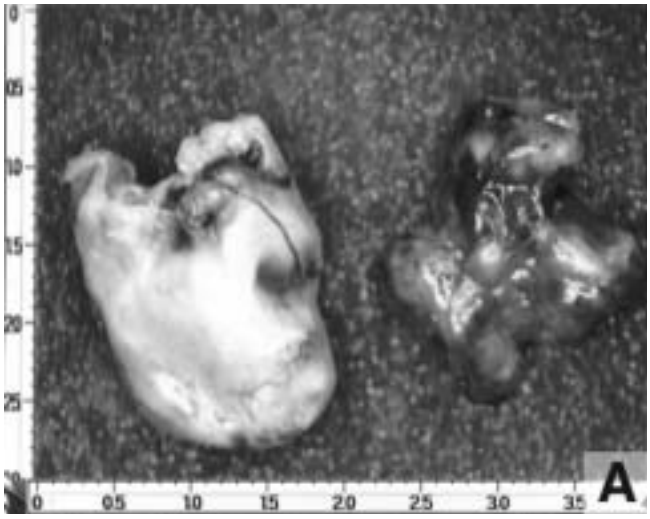

Figure 3a. Macroscopical appearance of the nasal tumour. Ruler indicates mm.

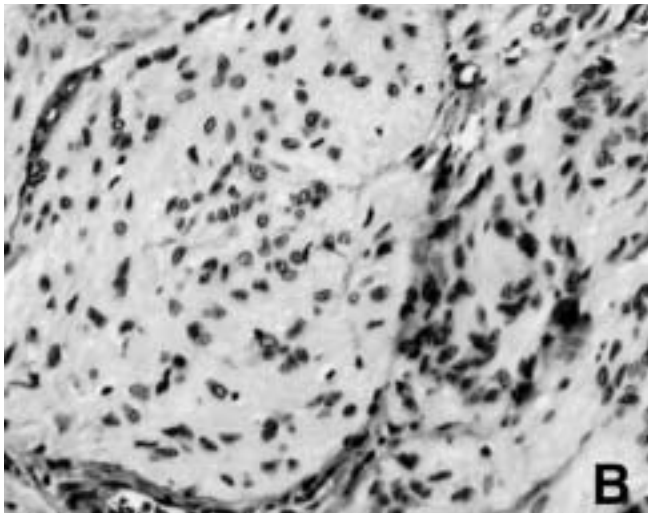

Figure 3b. Histopathological section of the tumour with nests of indolent gliomatous cells (H&E).

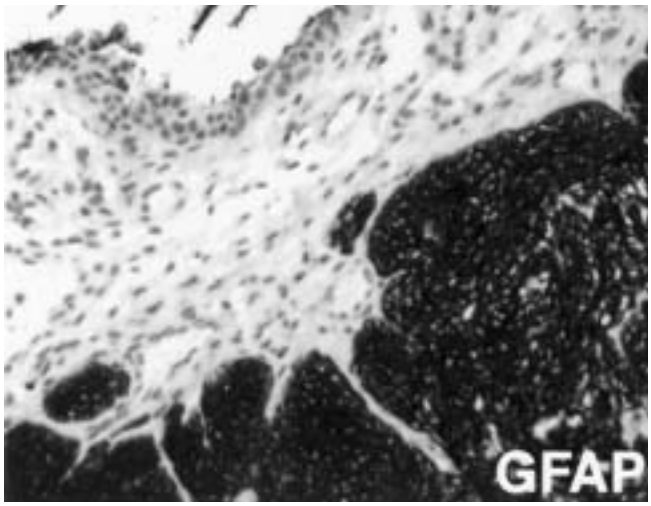

Figure 4a. GFAP: Immunohistopathology staining. Strong reaction over the tumour cell population, but negative stain within normal tissue.

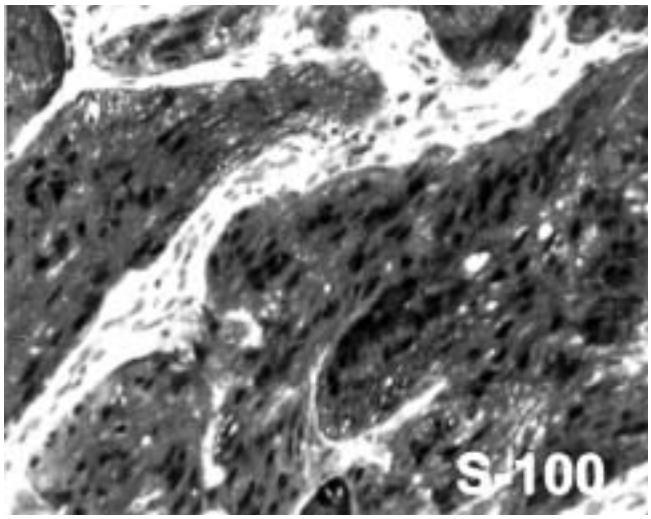

Figure 4b. S-100: Immunohistopathology staining for S-100 protein. Very strong positive reaction over the tumour cells.

DISCUSSION

Nasal glioma is a rare, benign congenital tumour situated in the nasal cavity, nasopharynx or paranasal sinuses (Karma et al., 1977). Sixty percent of the gliomas are extranasal, 30% are intranasal, and 10% are mixed intranasal and extranasal. Extranasal gliomas are usually found under the skin on the dorsum of the nose, most of them are diagnosed shortly after birth because of a cosmetic deformity: a broadened nasal bridge or the eyes appearing wider apart.

The intranasal lesion usually arises from the lateral wall of the nose, near the middle turbinate but occasionally it may also arise from the nasal septum (Gorenstein et al., 1980). On intranasal examination, the tumour appears firm, pale, and covered with a normal nasal mucous membrane. Only about 20 nasal gliomas have been reported to be found in the nasopharynx (Braun et al., 1992; Uemura et al., 1999; Sidebottom et al., 2000).

Histologically nasal gliomas are composed of globules of fibrillary or protoplasmic astrocytes surrounded by a fibrovascular stroma and a pseudocapsule. In about 10% of the cases scattered neurons have been identified. Caracteristically mitoses are absent (Dini et al, 1998; Tashiro et al., 1995).

CT in combination with MRI is essential in the diagnostic and pretherapeutic evaluation, notably to avoid missing an ectopic hypophysis (Braun et al., 1992). Nasal gliomas are usually isodense on CT, and on MRI they are often hyperintense on T2W images and hypo-, iso- or hyper-intense to the grey matter on T1W (Shah et al., 1999).

Preoperative biopsies and other nasal interventions should be avoided, as it could result in cerebrospinal fluid leak and meningitis. Diagnosis is thus confirmed by histological evaluation after extirpation.

Surgical removal of the extracranial portion of the tumour is the treatment of choice. This should be done in early childhood to prevent the tumour from deforming the developing facial bones. When an intranasal glioma has signs of an intracranial connection, a neurosurgical approach through a frontal craniotomy should be performed. Otherwise a lateral rhinotomy approach (Harrison et al., 1993), or an endoscopic approach is recommended (Burckhardt et al., 1999); the former preferably when the glioma is found posteriorly in the nasal cavity. The incidence of recurrence due to inadequacy of primary excision ranges from 4-10% (Dini et al., 1998).

There are many reasons for nasal obstruction in new-borns and infants, i.e. choanal atresia as well as several different intranasal masses like; dermoids, inflammatory polyps, papillomas, cysts of the lacrimal apparatus, carcinoma, hemangioma, fibroma, meningioma, lymphoma, sarcoma, as well as encephalocele, meningocele, and teratoma (Heacock et al., 1992).

As nasal gliomas are very rare we would like this article to be a reminder about this malformation in new-borns with upper respiratory tract obstruction. A MRI easily gives the correct diagnosis, and radical surgery cures.

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