Nasal Glioma and Ala Defects

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Abstract

Congenital nasal mid-line masses are rare, and nasal gliomas account for approximately 5% of all of them. They are benign lesion containing "ectopic brain tissue". Encephaloceles and gliomas are thought to have a close embryological origin and faulty closure of the anterior nasopore is suggested as a possible explanation. We present the case of an eight month old male who presented to us at Great Ormond Street Hospital for Sick Children, London. He had a left sided nasal glioma and also an ipsilateral ala notch. Only two other reference were found commenting on this unusual link between nasal glioma's and ipsilateral ala defects. This association perhaps suggests an alternative more complex embryological origin to the nasal glioma.

Key Words

Nasal Glioma, Ala notch

Congenital nasal mid-line masses are rare and occur in about 1 in 30,000 live birth in the United States, however they are more common in Asian populations, occurring in approximately 1 in 6000 live birth (Hengere AS : Congenital malformations of the nose and paranasal sinuses). Commonly encountered subtypes include encephaloceles, gliomas and dermoids.

Encephaloceles and gliomas are neurogenic tumours and are thought to have a close embryological origin. An encephalocele is a herniation of cranial contents through a defect in the skull. It may include meninges only, termed a meningocele, or it may involve brain and meninges, termed a meningo-encephalocele. Based on their position they can be subdivided into the frontoethmoid type which characteristically presents as an extranasal mass, and a basal type which presents as an intranasal mass. A glioma is thought to be an encephalocele that has lost its intracranial

connection, although 15-20% remain attached to the CNS via a fibrous stalk. They are composed of glial cells and fibrous and vascular connective tissue.

Case Report

An eight month old male child was referred to us to investigate the presence of a nasal mass. The mass had been present since birth. It was not growing in size. There was no nasal obstruction or rhinorrhea. The left eye occasionally watered.

Examination revealed a 3 by 2 cm mass on the left side of the nose in the nasofacial groove (See fig 1). The overlying skin was pigmented and hairy. No obvious midling pit was seen. The mass was not expansile on coughing or crying. The mass felt firm and was non tender. A notch was seen in the ala of the nose on the ipsilateral side. Anterior rhinoscopy was unremarkable and had a good nasal airway bilaterally.

The mass was excised via a modified weber ferguson type incision. The mass was closely tethered to the skin and underlying bone. It was however not extending into the nose. The overlying pigmented skin was excised and primary closure was achieved. Post op-recovery was unremarkable. Repair of the ala defect has been deferred till a later date.

Histopathology confirmed the mass to be a "glioma" with extensive involvement of the dermis.

Discussion

From an extensive review of the literature only two other references were found describing an association of nasal gliomas and ala defects. Tomson et al in 1975 described ipsilateral naso-ocular clefts in three patients with nasal gliomas, over a 28 year period. Ramakrishna et al in 1971 also described 2 cases with ipsilateral ala defects and nasal gliomas. A common feature of all these six cases was extensive dermal involvement.

The exact embryological details of their origin are yet uncertain although as outlined above they are thought to due to faulty closure of the anterior neuropore. The association seen between nasal gliomas and ipsilateral ala defects suggest an alternative more complex embryological origin. Perhaps an aberration in the development of the lateral nasal process and its

cartilage causes protrusion of certain portions of the primitive brain giving rise to the glioma. More

research is clearly needed to map the true embryological basis for the so called "nasal glioma".

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