

Prenatal diagnosis of a nasal glioma in the mid trimester

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ABSTRACT

We report a case of fetal nasal glioma diagnosed at 21 weeks of gestation. The glioma appeared as a moderately hypoechoic mass arising from the junction between the medial aspect of the left orbit and the lateral aspect of the nose, and showing no internal vascularization on color and power Doppler ultrasonography. Fetal magnetic resonance imaging (MRI) excluded the possibility of an encephalocele by ruling out underlying bone defects. After an uneventful pregnancy, the nasal glioma was resected without complications at 4 months of age. The differential diagnosis of fetal paranasal facial masses is discussed. Copyright © 2006 ISUOG. Published by John Wiley & Sons, Ltd.

CASE REPORT

A 37-year-old primigravida was booked for an anomaly scan at 21 weeks of gestation. She had been diagnosed with epilepsy at the age of 21 years, and was on valproic acid treatment. She reported smoking 10 cigarettes per day during the pregnancy. She had undergone genetic amniocentesis for advanced maternal age at 15 weeks, with a normal female karyotype. Amniotic fluid alpha-fetoprotein was 0.89 multiples of the median (MoM).

Ultrasound examination revealed the presence of a moderately hypoechoic mass, 7 mm in diameter, arising from the junction between the medial aspect of the left orbit and the lateral aspect of the nose (Figure 1a and b). The mass showed no internal vascularization on color and power Doppler ultrasonography. No other associated abnormalities were detected. In particular, intracranial structures and bony structures of the head and face appeared normal. The couple were informed that the most likely diagnosis was that of a nasal glioma. However, they were informed that alternative diagnoses

such as dacryocystocele, encephalocele, retinoblastoma, hemangioma or teratoma could not be completely excluded antenatally. Prenatal fetal magnetic resonance imaging (MRI) was offered and performed at 21 weeks (Figure 2a). Fetal MRI excluded the presence of an encephalocele by ruling out underlying bone defects, but could not provide a specific diagnosis of the nature of the lesion. With continuation of the pregnancy, the mass increased in size, reaching a diameter of 20 mm at 32 weeks' gestation (Figure 1c).

A female fetus was uneventfully delivered at 38 weeks of gestation, with a birth weight of 3390 g. The presence of a pink non-compressible mass between the internal canthus and the nasal bridge was confirmed (Figure 3). The nasal fossae were patent. Postnatal MRI suggested the diagnosis of a nasal glioma with minimal intranasal extension (Figure 2b). An uncomplicated resection was performed at 4 months of age, and pathological examination of the specimen confirmed the presence of nasal glial heterotopia mainly consisting of astrocytes with rare neuronal cells.

DISCUSSION

Nasal gliomas are benign non-hereditary congenital midline malformations, composed of heterotopic masses of neuroglial tissue. These rare lesions are estimated to occur in 1 in 20 000 to 1 in 40 000 live births, and present with a male:female ratio of 3:2¹. Approximately 60% of the lesions are extranasal, 30% are intranasal and 10% have extranasal and intranasal components. Abnormal closure of the anterior fontanel between the nasal and frontal bones during embryonic development can cause incomplete separation of ectodermal and neuroectodermal elements. Nasal glioma may develop if neuroglial tissue remnants persist extracranially following closure of the anterior fontanel, while anterior encephaloceles are due to the herniation of the meninges

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Figure 1 Prenatal ultrasound appearance of the nasal glioma at 21 weeks in left parasagittal (a) and oblique (b) sections. Small arrows indicate the margins of the glioma. (c) Coronal section of the fetal face at 32 weeks' gestation.

(with or without brain tissue) through an improperly closed fontanel².

Antenatally, the differential diagnosis of nasal glioma includes not only encephalocele, but also teratoma,

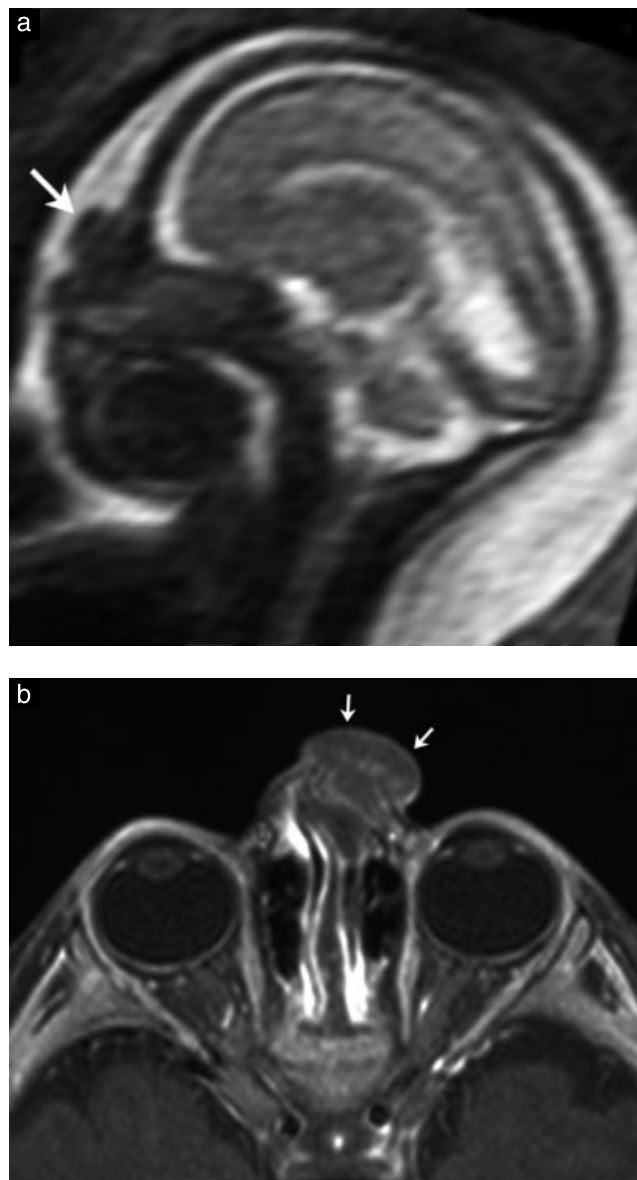


Figure 2 Prenatal (a) and postnatal (b) magnetic resonance findings. (a) Sagittal express turbo spin-echo T2-weighted image (20 000/140/1, repetition time/echo time/acquisitions; slice thickness 5 mm; acquisition time 20'') showing the mass lesion at the level of the root of the nose (arrow). (b) Contrast-enhanced, fat-suppressed axial spin-echo T1-weighted image (520/10/2, repetition time/echo time/acquisitions, slice thickness 3 mm, acquisition time 3' 42'') showing that the mass is slightly paramedian to the left and does not show contrast enhancement (arrows).

dacryocystocele, retinoblastoma and hemangioma³. Anterior encephaloceles are accompanied by a calvarial defect and frequently by ventriculomegaly. Teratomas have a complex internal ultrasound structure, and do not usually display internal Doppler signals⁴. Dacryocystocele is by far the commonest lesion in this group. Its prenatal ultrasound features are characteristic, as it presents as a cystic mass located inferiomedially to the orbit and is frequently shown to be in communication with the latter⁵. Retinoblastoma originates from the eye and has an irregular echogenic structure, surrounded by a sonolucent



Figure 3 Preoperative appearance of the mass.

area and covered by a membrane⁶. Hemangiomas can be septate or solid, and display a characteristic Doppler flow pattern with high flow velocities during arterial diastole, as opposed to the low flow velocity during diastole demonstrated in some pediatric cases of glioma⁷.

A previous case of prenatal diagnosis of nasal glioma was reported by Chmait *et al.* in 2002⁸. At 31 weeks of gestation, they described a left paraorbital cystic mass measuring 19 × 15 mm not communicating with any of the surrounding structures and not displaying internal Doppler signals. A provisional diagnosis of dacryocystocele was proposed, although the patient was informed of the differential diagnosis. Postnatally, the mass was diagnosed as a nasal glioma and successfully treated at 3 months of age. It must be noted that in both our case and in that of Chmait *et al.*, the masses showed a bright external contour (probably representing the skin) and an internal low-echogenicity but not completely anechoic content. Particularly if the mass is small, it can be very difficult to ascribe these findings to a solid content

as opposed to a cystic structure, thereby complicating the differential diagnosis.

The present report demonstrates that although the diagnosis of nasal glioma can be performed as early as 21 weeks, the prenatal findings should always be confirmed postnatally and parents must be informed about the uncertainties of the diagnosis. If the most severe abnormalities can be excluded with some confidence *in utero*, careful postnatal assessment of paranasal facial masses is crucial in planning the treatment of these lesions. In particular, gliomas may be difficult to differentiate from hemangiomas^{2,7}, and evaluation by computed tomography or MRI of their intranasal extension is useful in order to tailor surgical treatment and reduce the risk of incomplete resection.

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