Does nasal neuroglial heterotopia represent a risk for the newborn during delivery?

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Submitted: 2014-09-23 Accepted: 2014-11-08 Published online: 2014-11-30

Key words: **nasal glioma; neuroglial heterotopia; magnetic resonance imaging; newborn**

Neuroendocrinol Lett 2014; **35**(Suppl. 2):211–213 **PMID:** 25638389 NEL351014C01

Abstract

Neuroglial heterotopia is a rare developmental abnormality. Most frequently the diagnosis is established at birth or in early childhood by a typical clinical presentation. Neuroglial heterotopia can be intracranial or extracranial. A typical example of extracranial heterotopia is nasal glioma, which can be isolated or can communicate directly with the intracranium. The most sensitive investigation for the confirmation of its site is magnetic resonance imaging. Histological investigation is crucial in establishing the diagnosis.
The authors present the case of postnatally assessed nasal glioma. They emphasize the importance of detailed prenatal investigation as most important in preventing birth trauma and consequent complications.

INTRODUCTION

Heterotopic neuroglial tissue can be intracranial or extracranial. Both types can be isolated or communicate with the surrounding structures. Kau *et al.* (2011) described intracranially located transsphenoidal extension of heterotopic glioneuronal tissue. Ozgen *et al.* (2007) published the case report of a patient with intracranial extracerebral brain tissue.

Nasal neuroglial heterotopia is a rare congenital developmental abnormality. It is thought to represent encephaloceles that become sequestered on the extracranial side of the skull base (Choudhury *et al.* 1996; Husein *et al.* 2008). It is seen in a wide age range (Penner & Thompson 2003; Sequel & Gonzales 2003; Ohta *et al.* 2010; Vilarinho *et al.* 2000). The incidence is very low. Most frequently the diagnosis is established at birth or in early childhood; a few cases have been found in adults.

Penner and Thompson (2003) presented a group of 10 patients with a high mean age of 8.6 years at presentation (range 0.2 to 44 years).

Of the **clinical manifestations** of nasal neuroglial heterotopia, the typical finding is the presence of a polyploid mass in the nasal cavity (inner form) or in the area of the nasal bridge (external form) (Hussein *et al.* 2008; Ohta *et al.* 2010; Kardon 2000). According to the site of presentation, nasal glioma can have a direct connection with the central nervous system, yet mostly it is without connection. Penner and Thompson (2003) confirmed a direct communication of nasal glioma with the central nervous system only in one out of ten patients presented.

CASE REPORT

Histological investigation of the „mass" usually confirms the presence of varying proportions of astrocytes, neurons, ependymal cells, neuroglial fibers, fibrovascular connective stroma, calcifications, inflammatory cells and varying degrees of fibrosis (Penner & Thompson 2003; Ohta *et al.* 2010).

CASE REPORT

The patient was the mother's first child from her third pregnancy (1 abortion, 1 interruption of pregnancy). The delivery was in the 39th week of gestation, spontaneous in head position. Birth weight was 3 300 grams, birth length was 49 cm, the value of Apgar scoring system was 10/10 points. Amniotic fluid was clear (rupture of membranes occurred 14 hours before delivery). Screening for beta hemolytic streptococcus was negative (smear from the vagina). The process of direct postnatal adaptation was without problems.

The male newborn was admitted to our neonatal department at the age of 2 hours. The most dominant feature was the presence of numerous minor bleedings in the skin of the forehead and an intact prominent "mass" in the area of the nasal root (Figure 1). Sonography confirmed hypoechogenicity of this "mass" without the presence of color Doppler signal. The "mass" was well separated from its surroundings (2.09×1.94×0.92 cm). Magnetic resonance imaging of the central nervous system established the size of the mass: in transversal level 15×10 mm, in sagittal projection 17×15 mm, the basis was wide but without communication with the intracranium. The brain, ventricles, subarachnoid spaces were of normal values, without dilatation and asymmetry.

Fig. 1. Nasal glioma in a term newborn.

Rhinoscopy confirmed the narrowing of the left part of the nasal cavity caused by deviation of the nasal septum. The otorhinolaryngologist did not confirm a direct presence of coele in the nasal cavity. On neurological investigation, a slight decrease of muscle tone without asymmetry was found. Complete excision of the "mass" was performed. Histological investigation confirmed the presence of neuroglial tissue. Immunostain for glial fibrillary acid protein showed areas of neuroglial tissue. The patient is now 3 years old in good condition.

DISCUSSION

Nasal glioma is a tumor of extracranial neuroglial tissue with or without intracranial connections. This mass is firm and incompressible. It has a low incidence yet a wide age range at presentation. The presence of the "tumor" mass in our patient was not confirmed prenatally, only immediately after birth. The delivery was spontaneous in head position.

The size of the "mass" in extracranial position may cause acute problems during delivery (rupture, infection, hemorrhage, asphyxia). In the case of our patient, due to the presence of numerous small bleeding into the skin of the forehead, we can assume that the newborn had problems during delivery but the pressure was not so severe as to cause rupture of the nasal glioma.

Nasal glioma can be joined with either respiratory problems due to obstruction of airways or enlargement due to direct communication with the inner structures of the brain. Our patient did not have respiratory problems; the breathing was spontaneous, not requiring oxygen therapy or ventilatory support. Cases of large nasal glioma may be associated with manifestations caused by obstruction of the airways (Husein *et al.* 2008; Ohta *et al.* 2010; Kau *et al.* 2011).

The imaging techniques can help to localize the glioma and provide the important information whether it has communications with the inner structures of the brain. The most sensitive techniques are magnetic resonance imaging and high-resolution computed tomography (Ohta *et al.* 2010; Kau *et al.* 2011).

In differential diagnosis, it is very important to distinguish glial heterotopias of the nose from nasofrontal encephalocoeles (Choudhury *et al.* 1996).

The condition requires surgical treatment. The result depends on the size and localization of the nasal glioma.

CONCLUSION

The care of a child with a mass of unknown origin must be multidisciplinary. Establishment of the presence of anomalous mass before labor and prevention of delivery complications are of utmost importance. The next step would be the decision of the most advantageous mode of birth, considering that a nasal glioma is firm and incompressible. The aim of optimal management of

the newborn after delivery is to decrease the spectrum of complications, such as birth trauma, bleeding, infection, damage of the nervous system, etc.

Early diagnosis improves the strategy of health care management of the patient. It is necessary to ensure regular long-term monitoring of the clinical status, involving the care of a neonatologist, pediatrician, neurologist, and neurosurgeon.

ACKNOWLEDGEMENTS

The work was supported by the Ministry of Education, VEGA 02/0081/11.

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