Frontonasal glioma – diagnostic difficulties

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Abstract

Nasal glioma or nasal glial heterotopia represents a rare rare benign tumor of the median line. Prenatal and even postnatal imagingc diagnosis of congenital frontonasal masses is difficult. We describe the case of an infant presenting with a lateral nasal mass. The characteristics of the prenatal magnetic resonance imaging and the postnatal clinical examination argued in favor of a congenital hemangioma. The non-regressive aspect of hemangioma under Propranolol therapy pleaded for a possible non-involuting congenital hemangioma. To rule out other diagnosis we performed a postnatal magnetic resonance imaging that indicated a tissue lesion and ruled out an encephalocele. Surgical treatment was performed. The histologic examination revealed glial tissue and connective tissue, with immunohistochemistry confirming nasal glial heterotopia. Nasal glioma can be misdiagnosed as hemangioma. Magnetic resonance imaging and pathology with immunohistochemistry are the mainstay of diagnosis.

Keywords: nasal glioma, nasal glial heterotopia, hemangioma, MRI

Introduction

Congenital frontonasal masses represent a group of very rare benign lesions that occur in one of every 20,000 to 40,000 live births [1]. Differential diagnosis of the tumors from the median line include tumors having a mesodermal origin (congenital and infantile hemangiomas, lymphangiomas, angiofibromas, lipomas), an ectodermic origin (dermoid cysts), or a neurogenic origin (encephaloceles, nasal glial heterotopias), and teratomas [2].

Nasal neuroglial heterotopia (NGH, previously known as nasal glioma) and encephaloceles are included in this group and often discussed together given their similar neural origins. One of the main differential diagnosis of NGH is congenital hemangioma, due to their similar clinical presentations [3-6]. Prenatal imaging diagnosis of these entities is difficult.

Imaging examinations are essential for diagnosis and to rule out a possible communication of the lesion with intracranial space. Complete surgical excision is advised early in life in order to avoid complications.

We present the case of an infant with a prenatal imaging of a median line tumor and the value of fetal and postnatal magnetic resonance imaging (MRI) correlated with excised tissue pathology and immunohistochemistry in the final diagnosis and treatment of the nasal mass.

Case report

A 7 months female patient presented for a left paranasal mass. The child was born through cesarian section at 38 gestational weeks from a primipara with a prenatal MRI diagnosis of nasal hemangioma. The clinical examination after birth also suggested a congenital hemangioma. Propranolol treatment was started.

The non-regressive aspect argued for a possible non-involuting congenital hemangioma. To rule out other tumor diagnosis we performed a postnatal MRI, which indicated a tissue lesion. The MRI was not typical for hemangioma and ruled out an encephalocele.

The lesion was resected and sent to the pathology analysis. The histopathology

DOI: 10.15386/mpr-2225

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and immunohistochemistry confirmed the diagnosis of nasal glial heterotopia. Due to the slow progression of NGH, the recurrence risk and the esthetic concerns in the region, clinical and imaging monitoring is compulsory.

Discussion

Congenital midline nasal masses are very rare anomalies in pediatric pathology. The most common entities are hemangiomas, nasal gliomas, encephaloceles and dermoid cysts [7]. During the embryonic development, as a result of the abnormal closure of the nasal and frontal bones there is an extracranial settlement of ectopic glial tissue [8].

Nasal glioma or nasal glial heterotopia represents a non hereditary, congenital malformation, benign, embryologically related to encephaloceles [9].

There are a few theories described regarding the development of nasal gliomas: encephaloceles with lost of the intracranial connection and meningeal continuity; inappropriate closure of the anterior neuropore (fonticulus frontalis); sequestration of glial tissue of the olfactory bulb during cribriform plate fusion; ectopic neural tissue cells [9,10]. In about 15–20% of patients there is a fibrous stalk that might connect them to the intracranial space [9].

Nasal glioma was first described by Reid in 1852 and then named by Schmidt in 1900 [11]. Nasal glioma or nasal neuroglial heterotopia is not a real tumor. Most authors prefer to use the terminology of nasal glial heterotopia, which more correctly describes the pathogenic mechanism and avoids the inappropriate connotation of a neoplastic lesion [12].

Nasal gliomas are usually located in the region of the glabella, but they can extend toward the nasal tip. They may be located extranasally (60%), intranasally (30%), or both (10%) [1,2,9,12]. Rarely they have been found in the nasopharynx, paranasal sinuses, tonsillar fossa, scalp, and orbit [13].

The clinical presentation of nasal gliomas will depend on its location. The extranasal gliomas present as incompressible masses, most often in the glabellar region. These masses are firm in consistency, noncompressible, nonpulsatile, grayish or purple lesions, often with a telangiectatic surface, which may orientate the diagnosis toward a hemangioma [14]. In our patient the lesion was round and well limited. The characteristics of the prenatal magnetic resonance imaging and the postnatal clinical examination argued in favor of a congenital hemangioma. Intranasal and mixed gliomas arise from the nasal septum or lateral nasal wall and may present with symptoms of nasal obstruction [14].

Nasal gliomas do not change their size with compression of the internal jugular vein based on the absence of a patent connection with the central nervous system [13].

Prenatal and post natal imaging by ultrasound, MRI or CT scan can offer an important role for obstetricians, radiologists, pediatricians and surgeons in the diagnostic and therapeutic approach. Biopsy or fine needle aspiration of nasal masses are contraindicated. The treatment of choice is complete surgical excision [8,15].

Histologic examination proves neuroglial tissue (astrocytes with fibrillary glial processes and fibroconnective tissue). The glial tissue can be confirmed by immunohistochemistry for glial fibrillary acidic protein (GFAP) or S100 protein [1,8,9,13,16].

The non-regressive aspect of our patient hemangioma under Propranolol therapy argued for a possible non-involuting congenital hemangioma. In our patient, to rule out other tumor diagnosis it was performed a postnatal MRI, that indicated a tissue lesion. The MRI was not typical for hemangioma and ruled out an encephalocele. The lesion was resected and sent to the pathological analysis. The histopathology and immunohistochemistry confirmed the diagnosis of nasal glial heterotopia.

Close follow-up after surgical resection is essential. Nasal gliomas are benign conditions, but incomplete excision results in a 4–10% recurrence rates [9,10,13,17].

Conclusion

Nasal neuroglial heterotopia and hemangioma are rare lesions that must be considered in the differential of congenital nasal masses. Prenatal and postnatal imaging are necessary for evaluation of these lesions. Treatment consists in surgical resection. Magnetic resonance imaging and pathology with immunohistochemistry are the mainstay of diagnosis. Close follow-up of these patients is of paramount importance, with a rare possibility of recurrence.

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