



Nasal glioma

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ABSTRACT

Congenital midline nasal masses are rather rare anomalies that occur in about one in 20 000- 40 000 live births. Nasal gliomas account for approximately 5% of all congenital nasal swellings. The most common are dermoid/epidermoid tumors, nasal cerebral heterotopias (nasal gliomas), and nasal encephaloceles, with clinical significance that some of them have an actual or potential central nervous system connection. We present a case of an 8-year-old boy who complained of slight hearing loss dating 2 month before. Anterior rhinoscopy showed an oval, elastic, smooth, uncompressible mass, at the upper third of the nasal septum, unchanged in size on the Valsalva test. The mass causes breathing difficulties on that left naris. Clinical diagnosis was hemangioma. In the histopathologic laboratory, on gross examination, the mass measured 1.0 x 0.7 x 0.5 cm, was well demarcated, smooth, elastic, homogeneous, firm and whitish-gray in color. On cut section, the mass was homogenous, firm whitish gray in color. It consisted of astrocytic neuroglial cells with fibrous connective tissue and covered by the normal respiratory mucosa. The diagnosis of a nasal glioma was made.

KEY WORDS: Glioma; Nose Neoplasms; Child

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INTRODUCTION

The developmental anomalies of the nose encompass a diverse group of conditions, which include nasal dermoids, gliomas, encephaloceles, nasal clefts, proboscis lateralis, arrhinia, polyrrhinia, nasopharyngeal teratoma, and epignathus (1).

The congenital midline nasal masses are rather rare anomalies, which occur in about one out of 20 000-40 000 live births. The most common are nasal dermoids, nasal gliomas and nasal (meningo) encephaloceles, which are clinically significant because some of them might have an effective or potential connection with the central nervous system (CNS). Nasal gliomas cause approximately 5% of all congenital nasal swellings. They are non-encapsulated CNS masses of neurogenic origin, which have lost their intracranial connections, and present as an obvious external or intranasal mass at birth, without associated surgical symptoms (2,3).

The term "nasal glioma" is a confusing misnomer as it implies a neoplastic condition with malignant potential, which it is not. It should be differentiated from glioma, which is a malignant tumor of the brain. In addition, it should be differentiated from a primary encephalocele, which is herniation of the cranial contents through a bone defect in the skull, through which it retains an intact connection with the central nervous system (4,5).

CASE REPORT

A boy, aged 8 years, complained of slight hearing loss noticed 2 month before. The boy is the first child from the first uneventful term pregnancy of the healthy parents. Since his first year of life, he has had several ear inflammations and has been treated with antibiotics. After the first adenoidectomy performed in his 3rd year, ear inflammations were not so frequent, about once a year. On tonal audiometry, bilateral conductive hearing loss was noted, with a bilateral ear-bone gap of about 40dB. The tympanogram B type was diag-

nosed. The bilateral paracentesis along with re-adenoidectomy was indicated. At the same clinical examination, the anterior rhinoscopy showed a rather normal finding, except for an oval, smooth mass localized at the upper third of the nasal septum. The size of the mass was approximately 1 cm, characterized by elastic consistency and smooth surface; it was uncompressible, unchanged in size on the Valsalva test, and was clinically diagnosed as hemangioma. The rest of the clinical examination gave normal results. There was no history of nasal trauma or repeated nasal bleeding.

The patient was admitted to the Clinic for Otorhinolaryngology for bilateral paracentesis, re-adenoidectomy and for the extirpation of the nasal tumor performed under general endotracheal anesthesia. The mass was resected and sent for examination to the Institute of Pathology and Histology.

On gross examination, the mass measured 1.0 x 0.7 x 0.5 cm, was well demarcated, whitish-gray in color, elastic and smooth. In cross sections, the mass was homogeneous, firm and whitish-gray in color. Histologically, the tumor consisted of astrocytic neuroglial cells surrounded by fibrous connective tissue and covered by a normal nasal, respiratory type of mucosa (Figures 1-3).

The patient recovered well postoperatively, with no evidence of an intranasal disease or cerebrospinal fluid leakage. Now, almost three years after the operation, the patient is of good health condition, without any signs of the relapse.

DISCUSSION

The term nasal glioma has been used to describe a congenital benign tumor of the nasal region containing neural tissue. The nature of these lesions remains open to controversy, because of the different localizations of the heterotopic neural tissue involved, the deficient development of the bony structures and the persistence or lack of the structural relations

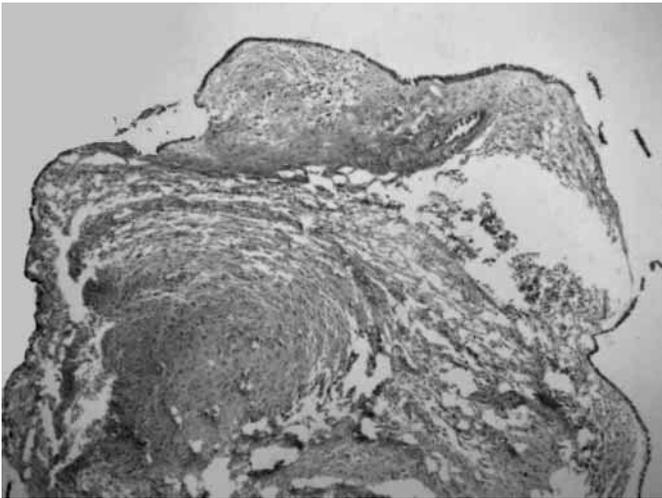


Figure 1. Tumor consisted of astrocytic neuroglial cells surrounded by fibrous connective tissue and covered by a normal nasal, respiratory type of mucosa (HEx 40)

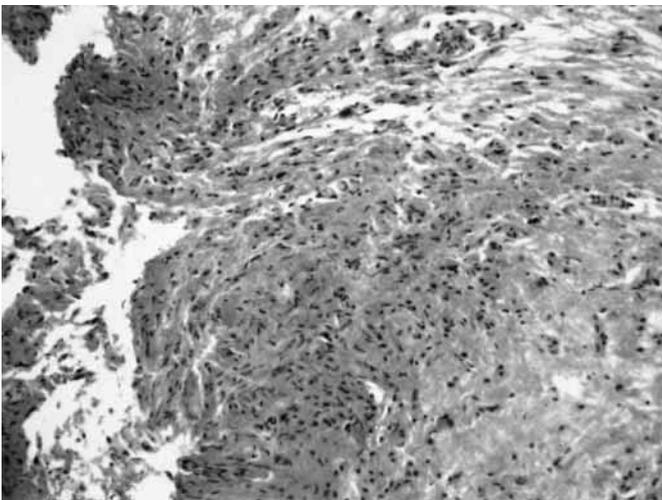


Figure 2. Neuroglial cells mixed with fibrous connective tissue (HEx 200)

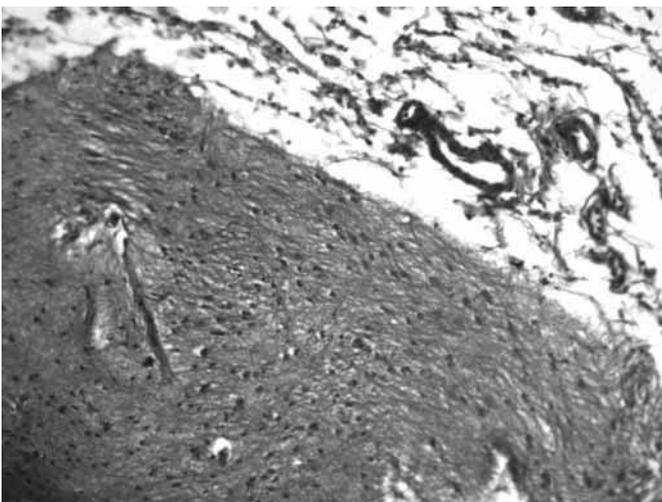


Figure 3. Neuroglial cells surrounded by fibrous connective tissue (HEx 200)

with the central nervous system. Terms that are more recent define these lesions as ectopic nervous tissue. "Nasal glioma" is a valid descriptive term defining a congenital benign tumor composed of heterotopic neural tissue within the nasal region and covered by skin or respiratory mucosa that may recur in up to 10% of cases, following incomplete surgical excision.

There are intranasal (30%), extranasal (60%), or combined (10%) types of nasal gliomas (6). Most nasal gliomas are diagnosed at birth or during early childhood, with relative peaks of occurrence between 5 and 10 years of age. Although the majority of patients present with signs and symptoms during the first year of life, a later presentation may be due to a specialist's failure to recognize a subclinical lesion in childhood or because of a trauma (2,5). Unlike dermoids, they do not necessarily occur in the midline, or attach to sinuses or skin. Gliomas form an uncompressible mass that does not increase in size on the Valsalva testing and does not transilluminate. Extranasal gliomas are usually located at the glabella level, but they may be present laterally. Intranasal gliomas are associated most frequently with the middle turbinate or higher structures, and may mimic nasal polyps. Combined intra/extranasal gliomas have a typical dumbbell shape with a connecting band. Fifteen percents of gliomas are connected with the dura, either through the foramen cecum or through the fonticulus. Patients may present with a unilateral nasal obstruction, unilateral nasal mass, epistaxis, or cerebrospinal rhinorrhea (4,5).

As gliomas are nonencapsulated accumulations of glial cells situated outside the CNS, the possible theories of development include the following: 1) sequestration of glial tissue of the olfactory bulb entrapped during cribriform plate fusion; 2) ectopic neural tissue cells; 3) pinched encephalocele; and 4) inappropriate closure of the anterior neuropore (fonticulus frontalis), with the failure of mesoderm to enter the region, resulting in an inadequate bone formation (7,8).

The clinical differential diagnosis includes several various congenital and acquired disorders, which could be manifested by nasal masses. Nasal glioma should be differentiated from several common or important lesions, among which: 1) Nasal dermoids, which are epithelial-lined cavities or sinus tracts with variable numbers of skin appendages, including hair follicles, sebaceous glands, and eccrine glands. They constitute the most common congenital nasal anomaly. 2) Encephaloceles point to the herniation of neural tissue through defects in the skull. They may contain meninges (meningocele) or brain matter and meninges (encephalomeningocele), or they may communicate with a ventricle (encephalomeningocystocele). Encephaloceles have an etiology similar to that of gliomas. Twenty percents of all encephaloceles occur in the cranium. Of those, 15% are nasal. 3) Hemangioma, which are the most frequent benign vascular tumors in infancy (9-11).

The diagnosis is corroborated by CT scan, which shows a bony defect. MR imaging is obligatory in patients with suspected glioma to determine possible intracranial extension of the nasal cavity lesion and the brain (12,13).

The treatment of choice is a complete surgical excision of the glioma to avoid the recurrence. The choice between endoscopic intranasal, lateral rhinotomy and frontal craniotomy depends on the lesion size and existence of CNS connections. The intranasal removal has an aesthetic advantage, but it carries the risk of incomplete resection.

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