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Short Report

Nasal Glioma: Report of Two Cases

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Nasal gliomas are rare, benign, congenital masses more accurately referred to as sequestered glial tissue (1). The nasal glioma first described by Schmidt in 1900 (2,3). Sixty percent of gliomas are extranasal, 30% are intranasal, and 10% are both (4). Extranasal gliomas are somewhat movable and usually do not increase in size when the child cries (5). Extranasal gliomas appear near the root of the nose. The overlying skin may be discolored or telangiectatic (6). We described two nasal gliomas, located extranasally. Histologically, the nodular tumor tissue resembled normal glial tissue. Immunohistochemical studies revealed the presence of glial fibrillar acidic protein (GFAP), indicating the glial nature of the tumor.

Case 1

This 5-month-old boy was brought for evaluation of a mass of external swelling on the right side of the nose, which had not increased in size since birth. He was a full-term child, the product of a normal delivery, and had no other abnormalities. There was no history of congenital defects in the three older siblings, and the family history was unremarkable.

Physical examination revealed an approximatelya 2x2 cm, firm, moveable, nonpulsatile mass located at the root of the nose just slightly right of the midline. There was a slight prominence to the vascularity of the overlying skin, but no other changes in the skin surface were seen. There was no intranasal mass. The remainder of the physical examination was unremarkable. The clinical diagnosis was thought to be pilomatrixorna. The patient was referred to the otorhinolaryngology service, where the mass was was excised by elliptical incision under general anesthesia. The mass found to be subcutaneous. The diameter of the tumor was 2x2 cm and appeared without a capsule, lobulated, and fairly firm, with grayish white cut surfaces. Low-power microscopic histologic examination of the hematoxylin and eosin stained sections demostrated pale areas of fine fibrillary tissue, separated by septa of more darkly staining collagen. The glial tissue was distributed haphazardly without any organization into layers or other mature structures (Figure 1,2). The fibrillary stroma was more clearly seen at higher magnifications. The individual glial cells, which had the characteristics of astrocytes, had very regular round nuclei, sharp nuclear membranes, and usually a single conspicuous nucleolus. The tumor cells were stained brown at examination for GFAP (Figure 3). The intense granular staining was generally confined to the cytoplasm of the tumor cells, with very slight staining of nuclei. The patient was well 10 months after surgery.

Case 2

A 1.5 -year-old girl presented with a swelling on the left side of the nose which had gradually increased in size since birth. She had not suffered pain or nose bleeds. There was no nasal obstruction. She was a full-term child. There was no other relevant history.

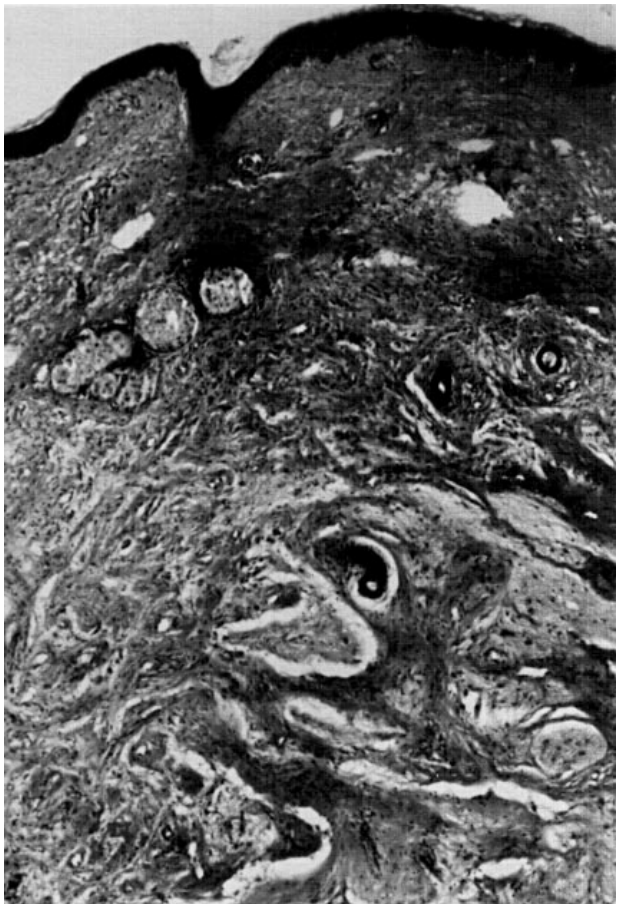

Fig. 1. Lightly stained glial tissue among darkly stained fibers beneath the epidermis (H.E. X40).

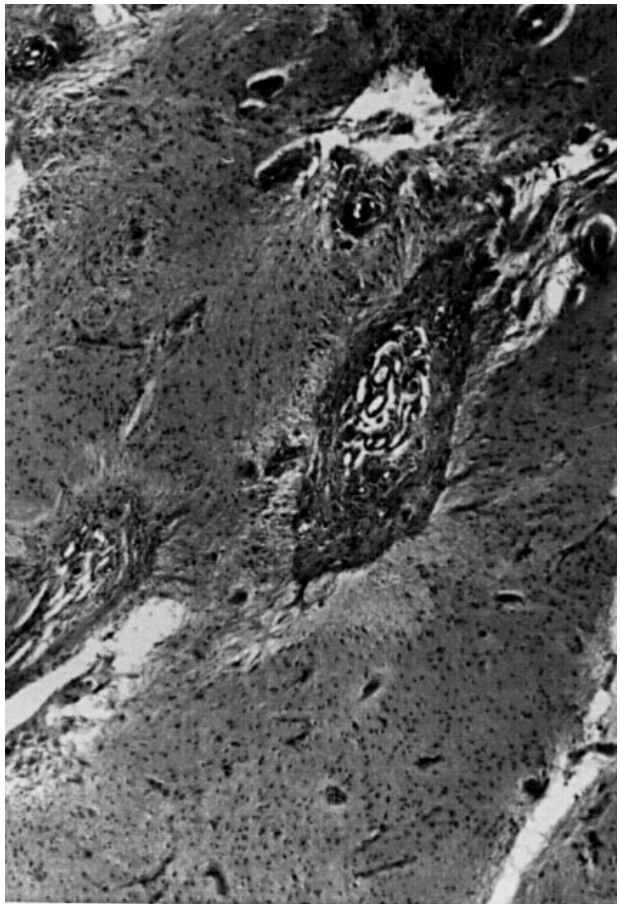

Fig. 2. Mature neuroglial tissues embedded in a fibrovascular stroma (H.E. X100).

Physical evaluation revealed that the subcutaneous swelling was 2x1 cm in size, on the left side of the nose. It was not tender, mobile, or pulsatile. There was no a mass inside the nose. The mass did not increase in size when the child cried. Plain skull films and a CT scan showed only a subcutaneous mass, without any intracranial extension or evidence of defect at the base of the skull (Figure 4).

The mass was excised with a lateral rhinotomy incision under general anesthesia. The lateral rhinotomy exposed an unencapsulated subcutaneous mass which extended intranasally through a gap between the nasal bone and the upper lateral cartilage. The distal nasal bone on the left and the upper nasal cartilage were found to have eroded. The histological appearance of the excised mass was the same as in the first case. Also, the tumor cells were positive for GFAP. The patient was well six months after surgery.

Gliomas are locally aggressive lesions noticed at birth or during early childhood, but which may be present at any age (4,6). The skin covering them may have telengiectasia. Nasal gliomas are seen more often in females, with a female:male ratio of 3:1 (6). Our patients were an 18-month–old female and a 5-month-old male.

Extranasal gliomas are skin-covered nodules most often located at the bridge or root of the nose, although they may also be found at the nasal tip. They are often located slightly to one side of the midline and range in size from 1 to 5 cm (7). In our patients, tumors were located at the root of the nose, just slightly to one side of the midline, and there was a nodular-like appearance.

Nasal encephaloceles and nasal gliomas have a similar embryological origin but, as the nasal encephalocele is a herniation of the intracranial contents, it must have an intracranial connection through a bone defect. The nasal

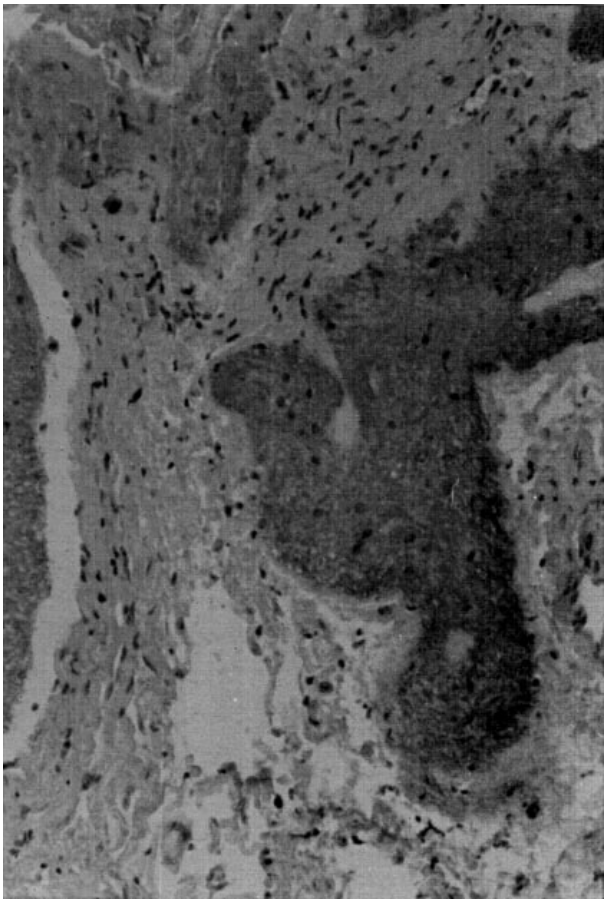

Fig. 3. The GFAP positive neuroglial tissue and the negative surrounding mesenchymal structures (GFAP, X100).

glioma, however, is ectopic sequestrated tissue and not a herniated structure, although a connection with CNS is present in 15-20% of cases. It is considered important to distinguish nasal gliom as from nasal encephaloceles because of the risk of infection spreading inward along the intracranial communication in the latter to produce meningitis (3,8).

Histologically, nasal gliomas consist of unencapsulated rests of glial cells, predominantly astrocytes, embedded in varying amounts of fibromuscular stroma (9,10,11). Multinucleated giant cells are often seen, but mitotic figures are rare or absent. Neurons have been identified in 10%-60% of cases in the series reported. Reactive changes and local calcifications as seen in some nasal gliomas may reflect the relatively poor blood supply to these heterotopias, which may also explain the paucity of neurons (11). In our patients, neuroglial cells in fibrovascular stroma were seen. No mitotic figures, multinucleated giant cells or neurons were observed.

The immunohistochemical demonstration of GFAP has been shown to be capable of identifying neuroglial cells with a high degree of specificity. Because of this specificity, this method succeeds in distinguishing glial cells within mixed cells populations or varying histological structures of neural origin where glial elements have lost their characteristic features of differentiation (12). GFAP staining of neuroglial cells was positive in our patients. On rare occasions, meningiomas can be seen in the nose and elsewhere outside the cranial cavity. Immunohistochemical identification of S-100 protein and

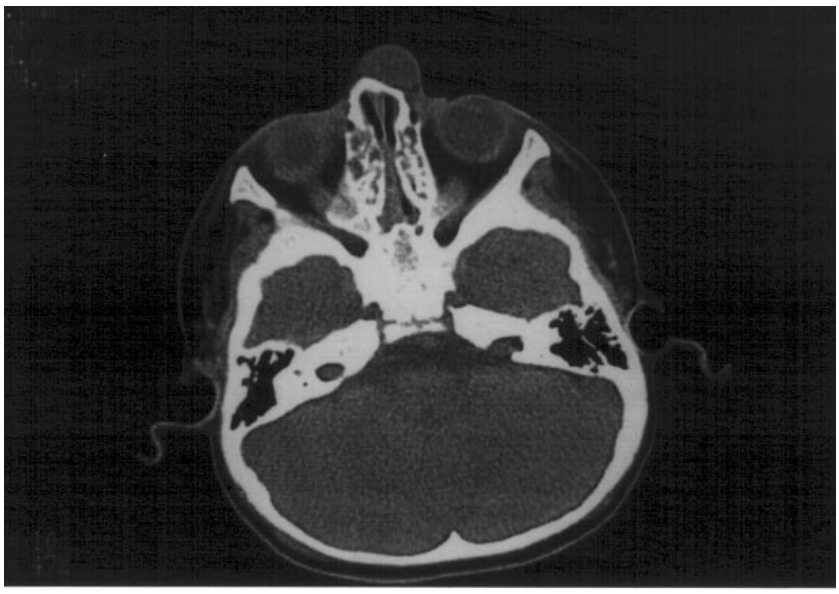

Fig. 4. CT scan shows a subcutaneous swelling, 2x1 cm in size, on the left side of the nose, and eroded distal nasal bone on the left.

GFAP helps to distinguish these lesions from one another (2,13).

Diagnosis of both intranasal and extranasal gliomas involves a detailed CT study of the nasofrontal area and anterior cranial fossa to rule out intracranial connections. Needle aspiration of these masses is to be avoided, because of the danger of iatrogenic meningitis. Other intranasal neural tumors, including neurofibromas, and neurilemmomas may be distinguishable from intranasal gliomas. Diagnosis is usually made by biopsy after the integrity of the anterior base of the skull has been determined (5).

Extranasal gliomas with no obvious CNS connection may be excised externally, using either a vertical elliptic midline incision or a horizontal incision over the dorsum of the nose. Both types of incisions have yielded equally good cosmetic results. Postsurgical defects of the nasal bones may spontaneously fill in over time or may require bone grafting at a later date (5). If a CSF leak is encountered, a bifrontal craniotomy approach may be required. Intranasal gliomas usually arise from the lateral nasal wall and can be approached via a lateral rhinotomy incision. If an intracranial connection is found, a craniotomy or an external ethmoidectomy may be necessary (6). Elliptical incision was used in the first patient, and lateral rhinotomy incision in the other patient.

Thomson et al. (1) suggested total excision of the skin adhering to the mass if glial elements are seen within the dermis on frozen section. They reported that no recurrences were seen after this new treatment protocol. During the 10-month follow-up period for the first patient, and during the six-month follow-up for the second patient, no recurrences were seen.

The basic rule for midline nasal masses is never to perform a biopsy until one has ascertained whether there is intracranial extension that might cause significant intracranial complications if a biopsy is done (14).

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