ABSTRACT

Nasal gliomas are rare, benign, congenital, midline tumors. They are seen most commonly in neonates and children but rarely in adults. We encountered in a 28-year-old woman a rare case of nasal glioma presenting as bilateral total blindness. The tumor was present in both nasal fossae. It extended into the parasellar region, with erosion of the cribriform plate and medial wall of the right orbit.

CASE REPORT

Nasal Glioma Presenting as Blindness in an Adult

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Nasal glioma was first described in the literature in 1890.[1] However, the first comprehensive description of the tumor and the first complete case reports were published only in 1950.[2] The authors described that these tumors originated from an encephalocele and also suggested the possibility of teratomatous origin.[2] Since then, these tumors have been described in the literature under various names: fibroglioma, ganglioblastoma, ganglio-neuro-schwanno-spongioblastoma, glioma, nasal glioma, encephalocele, and choristoma.[3] Now the term *nasal cerebral heterotopia* is gradually gaining universal acceptance. The term *nasal glioma* is probably retained due to its widespread clinical use.

These rare tumors are most commonly seen in neonates and children and are extremely rare in adults. Although orbital involvement has been reported, to our knowledge, complete blindness associated with nasal glioma has not been reported in the literature so far. We herein report a case of nasal glioma presenting as bilateral total blindness.

Case Report

A 28-year-old woman reported to the ophthalmology services of our hospital for evaluation of complete loss of vision in both eyes. Apparently she was asymptomatic 6 months earlier. Then she noticed a gradual decrease of vision in both eyes. For the last 2 months she was unable to see at all. On ophthalmic examination, in both eyes there was no perception of light. The

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remaining findings of the ophthalmologic examination were within normal limits. A computed tomographic (CT) scan of the orbit and paranasal sinuses was advised, which revealed a uniformly enhanced mass in both nasal fossae in the superior meatus area. There was erosion of the cribriform plate along with extension of the mass into the parasellar region. The medial wall of the right orbit was also eroded (Fig 1). The patient was referred to otolaryngology services. On nasal endoscopy, a reddish, globular, shining mass with capillary prominence was seen on both sides in the superior meatus. The remaining endoscopic findings were normal. An endoscopic biopsy specimen was taken from the right side, and the procedure produced substantial bleeding. The bleeding stopped with anterior nasal packing.

The histopathologic report established the diagnosis of nasal glioma. The patient was informed about the disease and choices for further management. She refused to undergo any further treatment and had no subsequent follow-up.

Histopathologic Findings

Histologic analysis of the biopsy specimens revealed brain tissue with small areas of capillaries and hemorrhage covered by nasal mucosa (Figs 2 to 5). The tumor mass was unevenly cellular, composed of small fibrillary astrocytes with pleomorphic and hyperchromatic nuclei. Microcystic spaces and mild endothelial proliferation were also present. There were also some areas of necrosis.

Discussion

Nasal gliomas are rare tumors usually seen in children. They are extremely rare in adults and more predominantly found in males.[2] They can be extranasal (60% of cases), intranasal (30%), or a combination of extranasal and intranasal components (10%).[4] On physical examination, an extranasal tumor presents most frequently as a round, firm, red or bluish swelling on either side of the bridge of the nose. Intranasal tumors resemble nasal polyps, but they are slightly firmer in consistency and show increased bleeding tendency on biopsy. In both types there is no pulsation or alteration of size on the Valsalva maneuver.

Nasal glioma are considered to be formed in utero as a herniation of ectodermal neural tissue of the frontal lobe through a bony cleft into the nose ("encephalocele" pathogenesis.[5] Other theories for the formation of nasal glioma are as follows: the amputation of portions of the olfactory bulb during closure of frontal bone sutures[6]; the migration of glial cells along olfactory nerves during embryonic development[7]; the formation of neuroectodermal rests isolated in the nasal cavity early in embryonic development, forming glial ectopia[8]; and teratomatous formation.[9] Of all these theories, the "encephalocele" pathogenesis seems most acceptable due to close association between encephaloceles and nasal gliomas and the frequent occurrence of a stalk to the brain.[10]

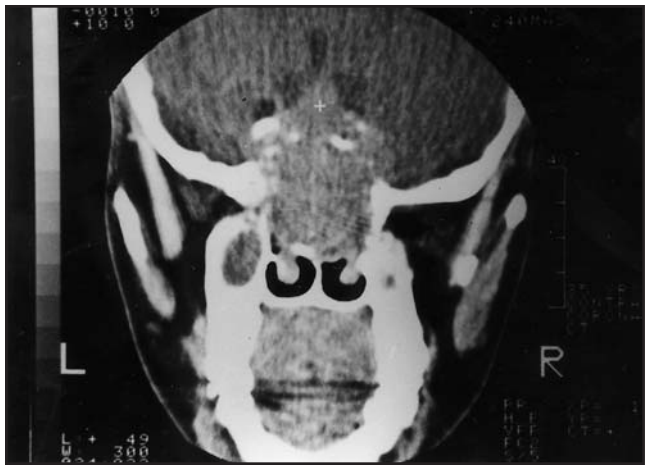

Fig 1.—CT scan of paranasal sinuses and orbit in coronal plane reveals uniformly enhanced soft-tissue mass occupying both nasal fossae, eroding cribriform plate and medial wall of right orbit, and extending into parasellar region.

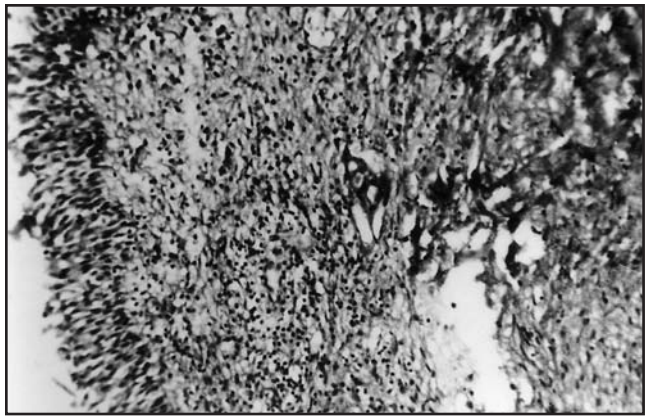

Fig 2.—Nasal epithelium covering brain tissue on biopsy specimen (hematoxylin-eosin, ×60).

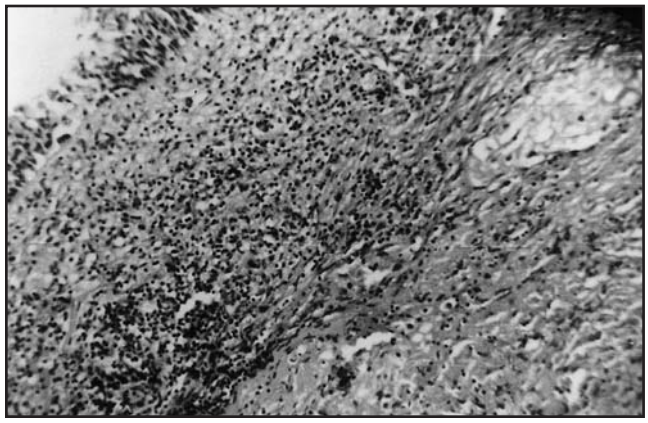

Fig 3.—Another histologic view of tumor showing nasal epithelium underneath, which is gliomatous tumor with unevenly distributed tumor cells, microcystic spaces, and an area of necrosis (hematoxylin-eosin, ×125).

Nasal gliomas are described as being occipital (75% of cases), sincipital (15%), or basal (10%) in location.[11] The sincipital gliomas are located above the dorsum of the nose, orbit, and forehead. These are further divided into 3 subgroups: nasofrontal, nasoethmoidal, and naso-orbital. The nasofrontal type is formed by pro-

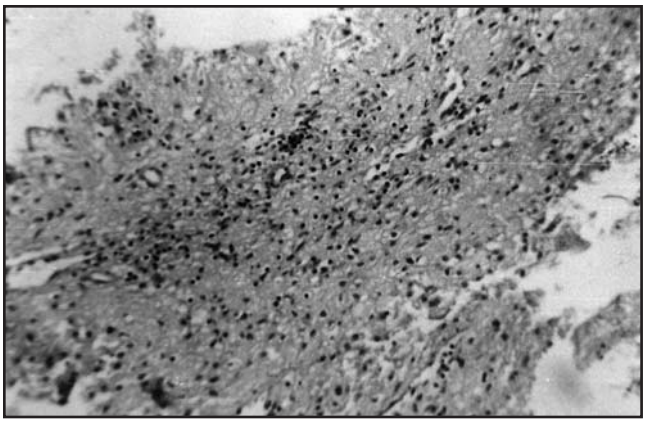

Fig 4.—Area of tumor with typical fibrillary background and astrocytes distributed more evenly. A large number of microcystic spaces are also seen along with mild degree of vascular endothelial proliferation (hematoxylin-eosin, ×125).

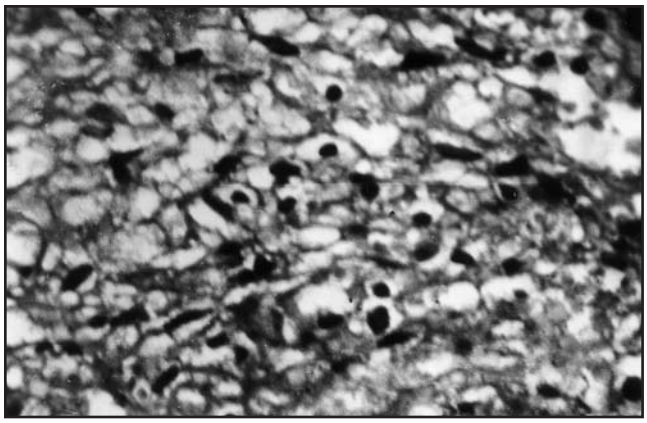

Fig 5.—Another histologic view of tumor showing fibrillary astrocytes with pleomorphic and hyperchromatic nuclei (hematoxylin-eosin, ×500).

trusion through the frontal and nasal bones; nasoorbital protrudes through the medial wall of the orbit, involving frontal, ethmoidal, and lacrimal bones.

The basal encephaloceles present as a mass protruding into the superior meatus of the nasal cavity, nasopharynx, or sphenomaxillary fossa. These are further grouped into transethmoidal, sphenoethmoidal, transsphenoidal, and sphenomaxillary. The transethmoidal type protrudes through a defect in the cribriform plate into the superior meatus. The sphenoethmoidal protrudes into the nasopharynx through a defect between the posterior ethmoidal cells and the sphenoid. The transethmoidal protrudes through a patent craniopharyngeal canal into the nasopharynx. The sphenomaxillary protrudes through the supraorbital tissue, through the infraorbital tissue, and then into the sphenomaxillary fossa, presenting as a mass on the medial side of the mandibular ramus.

Histologically, nasal gliomas are composed of brain tissue in which there is usually a significant and often dominating gliosis. The nest of glial cells is interlaced with a vascular fibrous tissue network or septa. Secondary changes of fibrogliosis or gemistocytic alteration of glial cells are often seen. Ganglion cells and other neural elements may also be present. The gliosis is probably secondary to circulatory impairment, death of cerebral tissue, and obliteration of the subarachnoid and perivascular spaces. Any "bizarre" astrocytes that may be present in the lesion are due to hypoxia and compression and should not be confused with neoplastic cells.

In terms of management, nasal gliomas should be excised completely to avoid any pressure effects of the growing mass. Because associated anomalies are quite commonly observed with nasal gliomas, a complete clinical examination should be performed before any surgical intervention. Computed tomographic scans of the nose, paranasal sinus, nasopharynx, base of the skull, orbit, and the surrounding area are a must. All patients also should be subjected to nasal endoscopic examination before any surgery.

This tumor, although not completely radioresistant, is not radiosensitive; thus, the treatment of choice is complete surgical excision. Surgery should be done by a team composed of the otolaryngologist, neurosurgeon, plastic surgeon, and ophthalmologist. Occasionally the intranasal tumor may be surgically resected by an intranasal approach. A lateral rhinotomy approach provides very good visualization of the cribriform area for evaluation of the presence of any glial stalk. Usually the combination approach of intranasal excision and lateral rhinotomy and craniotomy is preferred. The operating microscope is of immense help in the dissection of all types of nasal gliomas. Inoperable cases or recurrences after surgical excision should be treated by radiotherapy.

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