A Case of Nasal Glioma

Kyun Tae Kim, M.D., Beom Joo Lee, M.D., Sung Ku Ahn, M.D.,
Seung Hun Lee, M.D.* Won Soo Lee, M.D., Soo Im Choi, M.D.**

Department of Dermatology, Yonsei Wonju College of Medicine Wonju,
*Department of Dermatology, Yonsei University College of Medicine, Seoul**
*Taejon Eulji Hospital Taejon**, Korea*

We report a case with nasal glioma, which occurred in a 1-year-old male. He had a slow growing palpable mass on the left epicanthal region since birth.

Histological examination showed fibrillary astrocytes interlaced with fibrous and vascular connective tissue and S-100 stain was positive. The tumor was excised and the defect covered with a full thickness skin graft. (Ann Dermatol 6:(2) 215-218, 1994)

Key Words: Astrocyte Nasal glioma

Nasal glioma is a rare, benign congenital anomaly which is thought to be the result of an error in embryonic development[1]. The origin of this tumor is now accepted as a developmental abnormality by which a frontal encephalocele becomes partially or, more often, completely separated from the frontal lobes by the closure of the cranial sutures[2]. It is found in both sexes with a slight male predominance[3].

This occurs almost invariably as a solitary, firm, incompressible tumor on the root of the nose. It is composed of heterotopic brain tissue, predominantly glial tissue[3-5].

Occasionally a few cases of nasal glioma are referred to the dermatologist as an unusual subcutaneous mass over the bridge of the nose, most often in an infant. These lesions may not be recognized clinically, leading to dangerous mismanagement, and may be misinterpreted by pathologists as a neoplasm, either benign or malignant[4].

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Reprint requests: Kyun Tae Kim, M.D., Depargtment of Dermatology, Yonsei Wonju Collage of Medicine Wonju Korea.

REPORT OF A CASE

An 1-year-old male infant was seen at the department of dermatology with a slow growing palpable mass on the left epicanthal region since birth. Past medical and family history were unremarkable. Physical examination revealed a firm, smooth, red to blue, round and dome shaped mass that was quite large, measuring 1.5 to 2 cm in diameter, and resembling a hemangioma. On roentgenogram of the skull, no bone lesions or defects were demonstrated, and there was no calcification within the mass. Laboratory studies were within normal limits or negative.

The clinical diagnosis was that of hemangioma and the tumor was excised and the defect covered with a full thickness skin graft by the department of plastic surgery. The postoperative result was excellent and there was no evidence of recurrence at the end of three years.

Histological examination of left epicanthal mass showed fibrillary neuroglial cells, mainly large astrocytes and giant mulitinucleated astrocytes, interlaced with variable amounts of fibrous and vascular connective tissue(Fig. 1,2)and the S-100 protein stain was positive(Fig. 3). The pathologic diagnosis was nasal glioma.

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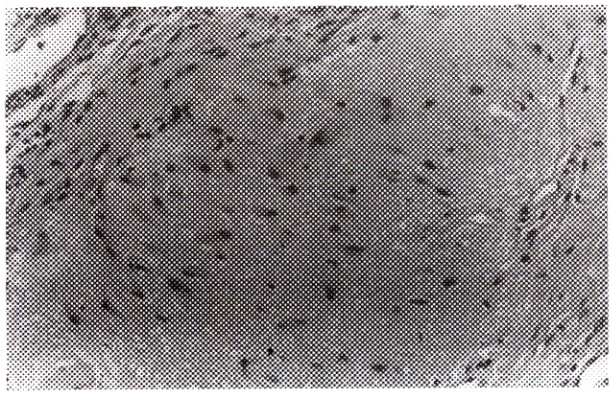

Fig. 1. The tumor is composed of glial cell and loosely arranged glial substance intermingled with scattered lymphoid cells and dilated blood vessels(H&E stain, ×

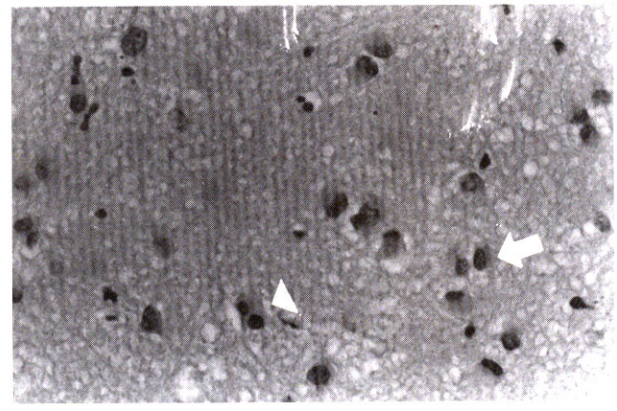

Fig. 2. Astrocytes(arrow) possess fairly light staining, oval nuclei and ample cytoplasm and some multinucleated cells(arrowhead) are observed(H&E stain, × 400).

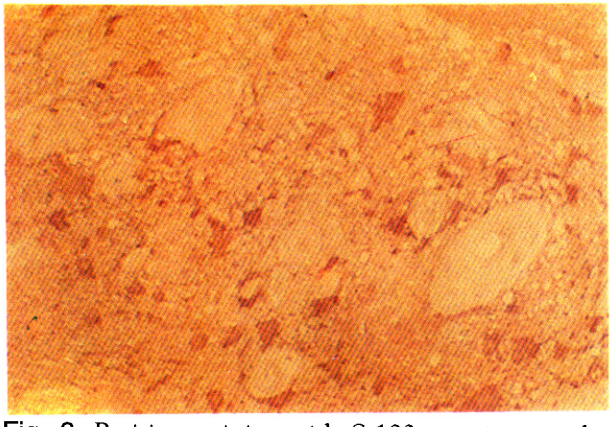

Fig. 3. Positive staining with S-100 protein was observed in tumor cells(PAP stain, × 400).

DISCUSSION

Nasal glioma was first reported by Reid in 1852, although the term "glioma" was coined by Schmidt in 1900[1]. There have only been a few reports of this case including this one from Korea[6-8].

This tumor is a developmental abnormality of neurogenic origin with no malignant potential. It is a benign congenital lesion usually recongnized at birth and rarely in adults. Its growh is usually about the same rate as that of the rest of body[5,9]. The sex ratio is 3:1 in male to female[2,3]. Most commonly, external nasal gliomas(60% of the reported nasal gliomas) are polypoid, firm, smooth, incompressible, red or bluish non-tender bulge to either side of the nasal dorsum and do not swell with the Valsalva maneuver. The nasal roof may be broad with slight hypertelorism and tearing usually occurs on the involved side. Intranasal nasal gliomas are seen as polyp-like masses in the nose or pharynx, usually with obstruction of the upper respiratory tract. Other locations for intranasal gliomas are the nasopharynx, maxillary antrum, and frontal sinus. They may communicate with intracranial contents, usually through a defect in the region of the nasal attachment of the frontal bone. Thus, biopsies or attempts to remove the intranasal glioma may cause a CSF leak. 10% of all nasal gliomas reported were both extranasal and intranasal[4,10,11]. Awareness of the mass, without associated symptoms, is the most common manifestation, but intranasal gliomas may have a watery nasal discharge(CSF rhinorhea), nasal stuffiness, epistaxis, frontal headaches, epiphora caused by obstruction of the nasolacrimal duct, and respiratory distress[11].

The pathogenesis of nasal glioma is unknown so far, but Schmidt[1] proposed the evagination of embyonic neuroectodemal tissue during fetal develpoment through the nasofrontal fontanelle, under the developing nasal and frontal bones similar to an encephalocele. Closure of the craniofrontal sutures, before the intracranial dura retraction, would isolate a mass of neuroectodermal tissue. An incomplete closure of the cranial sutures may result in a stalk of fibroglial tissue attached to the neuroectodermal mass through the foramen cecum. This seems to be the most logical and reasonable pathogenic mechanism[2,10]. Other theories[11] are the following : (1) the amputation of portions of the olfactory bulb during the closure of frontal bone sutures, (2) the migration of glial

cells along olfactory nerves during embryonic development, (3) glial ectopia formation, and, (4) teratomatous formation.

Histopathologically, there are interweaving stands of neural and fibrous tissue beneath a flattened epidermis. In addition, there may be dilated blood vessels. The neural tissue consists of glial cells or astrocytes, and loosely textured intercellular glial substance. The astrocytes posses a fairly light-staining, oval nuclei, and nucleoli are usually absent. The cells may be multinuclear but mitoses are rare[12,13].

When the presence of nasal glioma is suspected at birth, a tissue specimen is necessary for establishing a definite diagnosis. Incisional biopsy may not show definitive findings and also may be accompanied by the danger of CSF rhinorrhea if there is a preexisting intracranial connection. Excisional biopsy is the best method for histopathologic diagnosis and definitive treatment. Before surgical exploration other diagnostic procedures are helpful, although not conclusive. The Furstenberg test, done by compressing the ipsilateral jugular vein and observing the tumor for expansion or pulsation that occurs when there is a communication with subarachnoid or ventri cular space, is of questionable value in the case of external nasal gliomas[10]. Plain skull roentgenograms, pneumoencephalograms and computerized tomography may be desirable if a bony defect is present[11,14]. It is important that this lesion is considered in the differential diagnosis of a subcutaneous nasal mass in childhood, because the consequences of failing to recognize the possible existence of an intranasal component or open communication with a frontal lobe can be dangeros, and venturous treatment may result in meningitis or CSF leakage. Nasal gliomas must be differentiated from meningocele, encephalocele, ganglioneuroma, olfactory neuroblastoma, hemangiomas, dermoid cysts, and teratomas. A meningocele consists of meningeal tisssue only, while a true encephalocele, containing meninges, glial tissue and sometimes neurons, retainis an open and contiguous connection, composed of cerebral tissue, with the underlying brain. Ganglioneuroma, a rare tumor in children, is very uncommon at this site and histologically contains mature ganglion cells. Olfactory neuroblastoma is shown by primitive small cells forming a rosette parrern. Other nasal masses in childhood, such as hemangiomas, dermoid cysts, and teratomas, will obviously not cause any histological diagnostic difficulty[4,13].

Treatment is adequate surgical extraction, because gliomas are not radiosensitive. The extranasal glioma without evidence of intranasal component or intracranial connection may be simply excised. Intranasal gliomas may be treated by a combination of intranasal excision and that of excision with neurosurgical exploration are indicated, owing to the risk of meningitis[11,15]. A multidisciplinary approach to nasal gliomas, including neurosurgical, radiological, and probably plastic surgical advice, is mandatory[4,15].

In our case, a slow growing palpable mass was seen on the left epicanthal region since birth. On roentgenogram of the skull, no bone lesions or defects were demonstrated. The clinical feature was similar to that of hemangioma. The tumor was excised and the defect covered with a full thickness skin graft. Histological examination revealed fibrillary neuroglial cells, mainly large astrocytes and giant multinucleated astrocytes. S-100 stain was positive. The presence of S-100 protein in nasal glioma was consistent with the presence of neural cells in this tumor. The final diagnosis was nasal glioma.

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