Solitary primary intracranial extracerebral glioma

Case report

SAMRUAY SHUANGSHOTI, M.D., VIRA KASANTIKUL, M.D., NITAYA SUWANWELA, M.D., AND CHARAS SUWANWELA, M.D.

Departments of Pathology, Radiology, and Surgery (Division of Neurosurgery), Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand

A case is presented of a solitary primary extracerebral mixed glioma occurring in the right suprasellar and parasellar region of a 49-year-old woman who had bilateral temporal hemianopsia for 3 months. At craniotomy, the well demarcated outline and extracerebral location of the tumor suggested that it was a meningioma. However, its gliomatous nature was confirmed by identification of glial fibrillary acidic protein (GFAP) in the tumor cells. Review of nine reported solitary primary intracranial extracerebral gliomas, including the present case, revealed that they tended to occur in the third to fifth decades of life, in patients with an average age of 42 ± 8 years, and without sexual predilection. All were supratentorial with a tendency to be situated in the vicinity of the Sylvian fissure. Only the glioma in the present case was at the cranial base. They were diagnosed as three astrocytomas, two glioblastomas, two oligodendrogliomas, one astroblastoma, and one mixed glioma. A suggestion is made that all these gliomas arose primarily from heterotopic neuroglia in the leptomeninges.

KEY WORDS
- brain tumor
- glioma
- extracerebral tumor
- leptomeningeal glioma

To our knowledge, 17 gliomas arising primarily within the leptomeninges have been reported. Eleven of these were intracranial1,2,5,7,10,11 and six were intraspinal.2,3 Of the 11 intracranial leptomeningeal gliomas, two astrocytomas2,11 and one oligodendroglioma10 spread diffusely over the brain surface creating the leptomeningeal gliomatosis; the remaining eight gliomas were solitary.1,2,5,8 All six intraspinal leptomeningeal gliomas (three astrocytomas,2,3 two ependymomas,2 and one mixed astrocytoma and ependymoma,16) were solitary. The diffuse form of primary leptomeningeal gliomas may produce the clinical picture and gross appearance of meningitis rather than neoplasm, and the prognosis is worse than in the solitary form.2 The latter, on the other hand, may pathologically resemble some non-gliomatous tumors.

We have encountered a rare case of solitary primary intracranial extracerebral glioma possessing macroscopic features suggestive of meningioma. This instance combined with other previously reported examples of solitary primary intracranial gliomas will be analyzed relative to the type of glioma, the age and sex of the patients, and the specific location of the lesions.

Case Report

This 49-year-old woman was admitted to the hospital because of gradual loss of vision. She had worn eyeglasses for 20 years, with lens changes every 2 years. Three months before hospitalization, she complained that her temporal visual fields were blurred, but ophthalmological and neurological examinations disclosed only myopia. Two months later, this symptom had increased in severity. She was hospitalized 1 month subsequently for further investigation.

Examination. Temperature was 37°C, pulse rate 82 beats/min, respiratory rate 20/min, and blood pressure 120/80 mm Hg. The patient was conscious, alert, and cooperative. Both pupils were 3 mm in diameter and reactive to light. Ocular movements were normal in all directions. Papilledema was not observed. Motor power and sensation were intact.

Visual fields reveal bilateral temporal hemianopsia. Carotid angiography demonstrated lateral and anterior displacement and irregularity of the supraclinoid portion of the right internal carotid artery. A right suprasellar and parasellar mass was suspected. Computerized tomography (CT) of the brain exhibited an enhancing
and lobular mass in the right side of the sellar region with extension into the right middle cranial fossa (Fig. 1). It displaced the right internal carotid artery as well as the anterior and middle cerebral arteries.

Operation. A right frontotemporal craniotomy was performed (C.S.). Upon retraction of the right frontal lobe, a pink, well demarcated, lobular, extracerebral neoplasm of about $3 \times 3.5 \times 5$ cm was seen. The tumor invaded the dura mater and cranial base in the region of the right cavernous sinus and encircled the supraclival part of the right internal carotid artery. Biopsy was performed, and as much as possible of the remaining tumor was removed by suction. The operative impression was that of a meningioma.

Pathological Examination. The specimen consisted of multiple small pieces of rubbery tissue; these were fixed in 10% formalin and embedded in paraffin. Sections were stained with hematoxylin and eosin (H & E), Gomori’s reticulin, and Mallory’s phosphotungstic acid hematoxylin (PTAH). The peroxidase-antiperoxidase method was used to localize glial fibrillary acidic protein (GFAP) in formalin-fixed and paraffin-embedded tissue sections. An ependymoma was similarly processed as a control.

Microscopically, a large number of ovoid tumor cells were scattered throughout the specimen, and several of them had delicate blue processes in PTAH preparations. Occasional processes were attached to the vascular walls, forming pseudorosettes. Some neoplastic cells bordered a central space to form a true rosette (Fig. 2).
They had pale cytoplasm and round-to-ovoid nuclei that were moderately rich in chromatin granules. They were interpreted as neoplastic ependymal cells. A moderate number of round tumor cells possessed distinctly clear perikarya and dense, round, centrally placed nuclei creating a "fried egg" appearance. They were considered to be oligodendroglioma (Fig. 3). In the H & E preparations, a small number of neoplastic cells had angular outlines, with occasional thick processes extending from the corners, and ample glassy cytoplasm. The vesicular nuclei were often eccentric. They were regarded as astrocytes. The GFAP-positive tumor cells, mainly astrocytes, were scattered (Fig. 4) as were some ependymal neoplastic cells serving as a control. A few reticulin fibers were present perivascularly but were absent among various tumor cells.

The pathological diagnosis was a solitary extracerebral mixed glioma (ependymoma, oligodendroglioma, and astrocytoma) involving the right suprasellar and parasellar region of the cranium.

**Postoperative Course.** The patient received a course of radiotherapy totaling 6000 rads at the surgical area. She was discharged after 2 months of hospitalization and has been seen regularly during the past 2 years, with considerable improvement of her bilateral hemianopsia. There was no tumor recurrence during this 2-year follow-up period.

**Discussion**

The present tumor must be distinguished from meningiomas, pituitary neoplasms, gliomas of the hypothalamus and adjacent regions, and hamartomas of the neural tissue at the base of the brain. The extracerebral location, the well demarcated outline of the tumor, and the invasion of the dura mater and bone suggest it to be a meningioma. Meningiomas occur occasionally in the sellar region, however, the absence of the cellular whorls and reticulin fibers around the tumor cells makes this suggestion unlikely. Furthermore, identification of the GFAP-positive tumor cells indicates the gliomatous nature of the current neoplasm. Meningioma cells have not been known to be GFAP-positive, but GFAP is specific for astrocytes, ependymal cells, and some neoplastic oligodendroglia.

The location of the tumor around the sellar region again suggests it to be a pituitary neoplasm with supra-
FIG. 4. Photomicrographs taken at various places within the tumor to demonstrate neoplastic astrocytes with eccentric vesicular nuclei (arrows). The dark cytoplasm of these astroglia, as compared to other nearby tumor cells, is associated with glial fibrillary acidic protein (GFAP). The astrocyte in A has few processes. GFAP, × 1000.

Sellar and parasellar extension. Some pituitary tumors, especially chromophobe adenomas, may microscopically be similar to an ependymoma or even an oligodendroglia. The presence of GFAP-positive tumor cells again distinguishes our patient’s tumor from a hypophysal adenoma, which is GFAP-negative.22

Gliomas of the region of the hypothalamus and mamillary body may project extracerebrally as well as produce local destruction and functional disturbances of the hypothalamus.13,14 The absence of hypothalamic disorders, such as diabetes insipidus, argues against the location of our patient’s tumor in the hypothalamus or adjacent regions.

Neurogenic hamartomas may protrude from the base of the brain into the leptomeninges. The lesion is usually attached by a thick stalk to the tuber cinereum or the mammillary body, and is often composed of neurons, nerve fibers, and various kinds of neuroglia.13,15 We excluded the possibility of our patient’s lesion (which should be appropriately characterized as a genuine neoplasm) being a neurogenic hamartoma. The capability of the present lesion to invade the dura mater and bone is in accord with neoplastic behavior rather than that of a hamartoma, which manifests as a congenital disturbance in growth of the neural tissue. Neurons are not seen in the current extracerebral tumor.

Review of nine reported solitary primary intracranial extracerebral gliomas, including the present case, disclosed three astrocytomas,1,2,5 two glioblastomas,5 two oligodendroglia,7 one astroblastoma,9 and one mixed tumor (ependymoma, oligodendroglia, and astrocytoma, in our case). There were four male and five female patients. The youngest was a 32-year-old man; the two oldest were female, each aged 49 years. The average age was 42.5 years. All gliomas were supratentorial. Eight of them were on the cerebral convexity at the following regions: two frontal (glioblastoma and oligodendroglia);5 two frontoparietal (astrocytoma);2,5 two Sylvian fissure (astroblastoma and oligodendroglia);5 one frontotemporal (glioblastoma);5
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and one frontoparietal (astrocytoma). Only in our case was the mixed glioma located at the cranial base. We may conclude from this limited number of cases that no definite type of glioma predominated in the series. Nevertheless, if the astroblastoma is included in the category of astrocytomas, it seems that astrocytoma is the most common solitary primary intracranial leptomeningeal glioma. These gliomas tend to occur in the third to fifth decades of life without preference for either sex. They are situated predominantly in the vicinity of the Sylvian fissure of the brain.

Resemblance between these solitary primary extracerebral gliomas and meningiomas was observed grossly in two astrocytomas, one oligodendroglioma, and one mixed glioma (our case). Of these four cases, dural invasion was noted in one astrocytoma and in our mixed glioma. Invasion of bone occurred in one oligodendroglioma and in our combined glioma. One tumor, a solitary primary leptomeningeal astrocytoma of the right frontoparietal region in a 49-year-old woman, metastasized to an ipsilateral cervical lymph node 11 years after the craniotomy.

We suggest that our patient's solitary intracranial extracerebral glioma arose primarily from neoplastic transformation of the neuroglia within the leptomeninges. Heterotopic neural tissue is frequently found in the leptomeninges, and also in the brain substance. It may rarely be present in the wall of a congenital arachnoidal cyst, or form huge extracerebral masses. Heterotopic neural tissue may be composed not only of neuroglia but also of neuroblasts, neurons, and choroid plexus. Its occurrence has been suggested to be associated with aberrant migration of developing neuroepithelial derivatives. Cooper and Kernohan encountered neuronal heterotopia in 1% of a series of 100 consecutive autopsies. This finding is expected to be more common in persons with congenital malformations, particularly of the neuraxis. It is also expected that some extraneuraxial gliomas may arise primarily within the leptomeninges from neoplastic transformation of these ectopic neuroglia.

References


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Address reprint requests to: Samruay Shuangshoti, M.D., Department of Pathology, Chulalongkorn Hospital, Bangkok 10500, Thailand.