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Improvement of Visual Acuity Caused by Opticochiasmal Astrocytoma After Treatment for Associated Hydrocephalus

—Case Report—

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Abstract

A 33-year-old male presented with syncopal attacks. He had a history of slowly progressive deterioration of visual acuity in both eyes. His visual deterioration began in the left eye at age 12 years and in the right eye at age 20 years. His left eye was completely blind by age 33 years. He had received no prior treatment for these visual disturbances. Magnetic resonance (MR) imaging on admission showed a large tumor with inhomogeneous intensity occupying the bilateral optic canals and orbital cavities, with extension to parasellar region. Arachnoid cysts were found in the left middle fossa and supracerebellar space, which had caused herniation of the cerebellar tonsils to the foramen magnum. Open biopsy and histology verified that the tumor was pilocytic astrocytoma. Arachnoid cysts associated with the hydrocephalus were treated with a ventriculoperitoneal shunt. Seven years after surgery, visual acuity of the right eye had improved although the left eye remained blind. Follow-up MR imaging demonstrated marked reduction in size of the ventricles and arachnoid cysts, but the tumor size did not change. This case illustrates the benign nature of low-grade glioma in this region in spite of the large size and long history. Deterioration of visual acuity may be reversible in some cases of opticochiasmal astrocytoma. In our case, the improvement in visual acuity probably resulted from growth arrest of the tumor and improvement in the dynamics of cerebrospinal fluid flow.

Key words: arachnoid cyst, hydrocephalus, magnetic resonance imaging, opticochiasmal astrocytoma, ventriculoperitoneal shunt

Introduction

Low-grade astrocytomas in the optic nerve and chiasmatic region are rare tumors with an erratic natural history and are typically seen in young children. Some cases are fatal, despite initial clinical stabilization following radiation therapy. Visual, intellectual, and late endocrinological disabilities are common.[18] Studies covering a large number of cases have focused on the surgical management of these tumors, including biopsy, followed by radiation therapy.[4,17,22] In these studies, the actual survival rate was up to 90% at 15 years with stabilized vision. A high incidence of endocrine abnormalities has been observed in pediatric cases, the majority of which exhibit post-treatment growth hormone deficiencies and significant residual problems associated with radiation therapy.[1,11,17]

Spontaneous regression of large, clinically symptomatic astrocytomas in the optic nerve and chiasmatic region has been reported recently.[15,19] Such a regression may manifest either as overall shrinkage in tumor size or as change in magnetic resonance (MR) imaging characteristics. Improvements in visual function may accompany regression in size.[15] The incidence of spontaneous regression or arrest of growth in these astrocytomas is unknown and unpredictable.

We recently treated a patient with histologically verified pilocytic astrocytoma located in the bilateral optic nerve and chiasma regions who had remained symptomatic but untreated for more than 20 years before the biopsy surgery.

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Case Report

A 33-year-old male was admitted to our hospital for the evaluation of unresolved syncope in 1995. He had a long history of slow, progressive worsening of visual acuity. He first noticed that visual acuity in his left eye had begun to deteriorate at age 12 years. The process was slowly progressive and he lost vision in the left eye at age 17 years. He consulted an ophthalmologist who found atrophic changes in the optic disc of the left eye. No treatment was performed. Visual acuity of his right eye began to deteriorate at age 20 years and continued to progress for years. He developed swelling of the right upper eyelid associated with venous congestion a few months before the onset of syncopal attacks.

On admission, he was alert and well oriented. General physical status was excellent with mature endocrine development. No pathological pigmentation of the skin or subcutaneous tumors were noted. Visual acuity was 1/200 in the right eye and the left eye was blind. The bilateral optic discs were pale in color and atrophic. Exophthalmos was observed in the right eye with congestional swelling and chemosis. Eye movements were full and smooth. No neurological deficits of the motor or sensory systems were noted. His first-degree relatives had no signs of neurofibromatosis.

The laboratory test results were normal. The base line levels of pituitary hormones were within the normal ranges. Chromosomal analysis found no abnormalities. Skull radiography indicated decalcification of the sella. Computed tomography (CT) demonstrated an irregularly shaped tumor extending from the right optic canal to the sella, compressing the hypothalamic region. The tumor had inhomogeneous density, and the intraorbital component was partially calcified. The upper margin of the tumor adjacent to the hypothalamus was enhanced by contrast medium. MR imaging demonstrated two arachnoid cysts located in the left middle fossa and in the infratentorial supracerebellar region (Fig. 1). The latter cyst had compressed the cerebellum downward, resulting in herniation of the cerebellar tonsils to the foramen magnum (Fig. 1). The clivus was deformed, probably due to the long-lasting compression by the ventrally shifted brain stem. The lateral ventricles and the third ventricle were markedly enlarged.

The patient had a long history of ophthalmologic symptoms but the main reason for admission was to treat the syncopal attacks which had started a few weeks before admission. Herniation of the cerebellar tonsils was considered as the probable cause. CT cisternography was first performed to study the

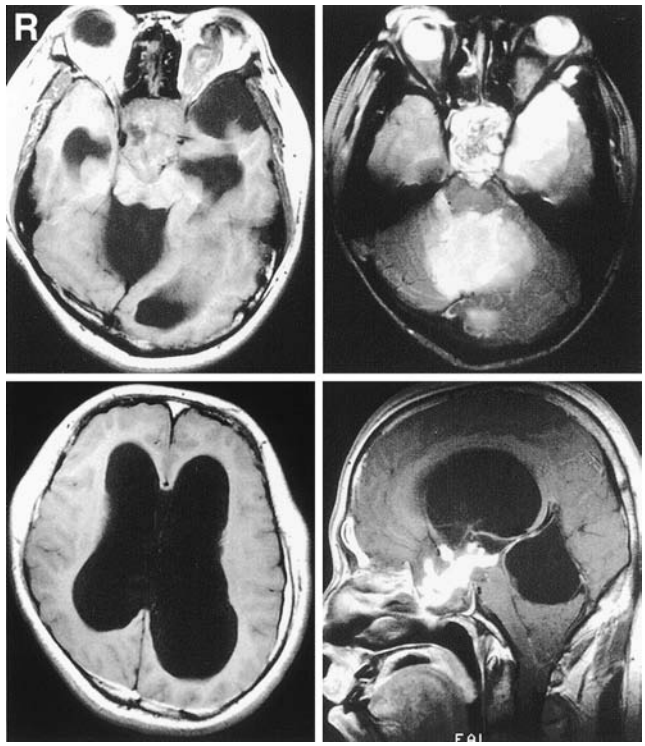

Fig. 1 Axial T_1-weighted (*upper left*) and T_2-weighted (*upper right*) magnetic resonance (MR) images on admission showing the tumor extended intra- and suprasellarly with inhomogeneous intensity, and an arachnoid cyst in the left middle fossa. T_1-weighted MR image (*lower left*) showing marked enlargement of lateral ventricles. Sagittal T_1-weighted MR image (*lower right*) demonstrating an arachnoid cyst in the infratentorial supracerebellar space, and the cerebellar tonsils herniated into the foramen magnum due to compression by the arachnoid cyst.

communication of the arachnoid cysts with the normal cerebrospinal fluid pathway. External drainages were placed under general anesthesia in the right lateral ventricle by the posterior lateral approach, and in the arachnoid cyst in the posterior fossa through a burr hole in the right suboccipital cranium. Intracranial pressure measured from the ventricular drainage tube was approximately 20 cmH_2O. Laboratory examination of the cerebrospinal fluid found protein 63 mg/dl, sugar 94 mg/dl, and cell count 0/3. Injection of contrast medium into the lateral ventricle resulted in rapid filling of the arachnoid cysts in the posterior fossa and the left middle fossa within 3 hours, indicating good cerebrospinal fluid communication between the ventricles and cysts (Fig. 2). To eliminate the

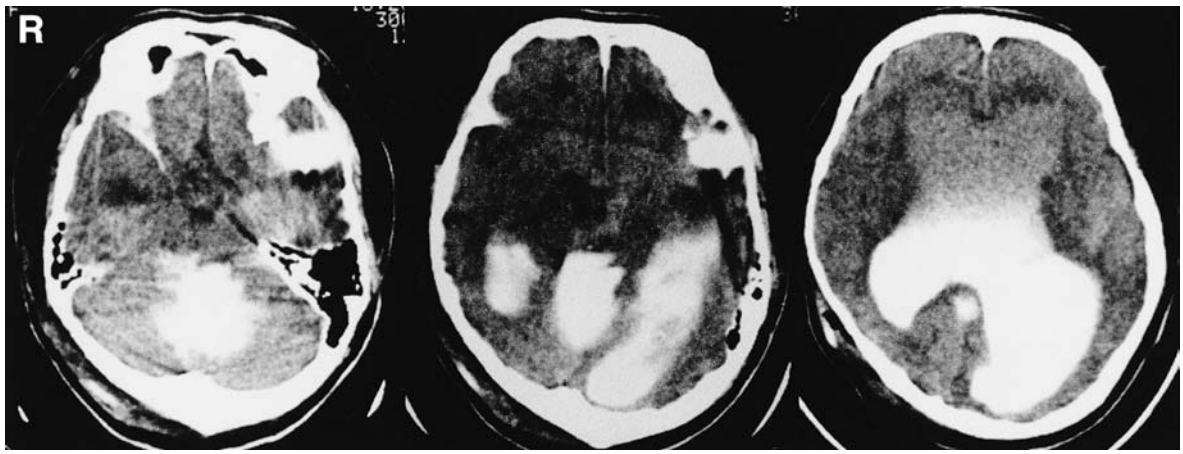

Fig. 2 Computed tomography cisternograms 3 hours after injection of contrast medium into the right lateral ventricle demonstrating rapid filling of the arachnoid cysts in the left middle and the posterior fossae.

herniation of the cerebellar tonsil, a right ventriculoperitoneal shunt was placed followed by left fronto-orbito-temporal craniotomy for biopsy of the tumor.

Intraoperatively, the intraorbital tumor was white and elastic-hard, and was partially removed to reduce the exophthalmos. The intracranial parasellar part of the tumor was soft and yellowish, and surrounded by a thickened arachnoid membrane. The tumor extended through the decompressed left optic canal. There was little bleeding from the tumor. A few pieces from the intracranial side of the tumor were taken for histological study. The postoperative course was uneventful and the patient was discharged after treatment of the swollen eyelid by plastic surgeons. At discharge, the patient's visual acuity was unchanged. Histological study showed bipolar shaped pilocytic astrocytes (Fig. 3). Cells with small round nuclei and a perinuclear halo were also occasionally seen. The pilocytic astrocytes were PTHA-positive, and the MIB-1 index was 0.7%. The final diagnosis was pilocytic astrocytoma associated with oligodendroglioma.

Since the ophthalmological symptoms in both eyes had been slowly progressive for years, blindness in the right eye was strongly expected in the future. Radiation therapy was recommended as an adjuvant therapy but not performed because of the risk of deterioration of the remaining visual acuity due to radiation-induced neuropathy, and the risk of radiation-induced hypopituitarism. The chief complaint of the patient was syncopal attacks as mentioned above and the patient refused further treatment.

Follow-up MR imaging was performed in 2002, as the patient had not visited our hospital for 7 years since the surgery. At that time, his physical condition was excellent, the same as 7 years earlier, and he was still working in his previous agricultural occupation. He had not suffered any episodes of syncope after the surgery. He was still blind in the left eye as before the surgery, but the visual acuity of the right eye was 20/40, showing significant improvement. The visual field of his right eye showed no deterioration. MR imaging revealed no significant change in tumor size (Fig. 4). The size of the ventricles was moderately reduced and the cerebellar tonsillar herniation was relieved. Baseline levels of the pituitary hormones were within normal ranges, so no treatment for endocrine deficiency was required.

Discussion

In the present case, the actual course of tumor growth was unclear because no evaluation was performed for more than 20 years after the onset of ophthalmological symptoms. As demonstrated by the progressive ophthalmological symptoms, the tumor had probably grown indolently since age 17 years. Low-grade gliomas in the location of the optic chiasma were suggested to be non-neoplastic and self-limiting tumors, and surgical removal of the tumor does not prolong life and should be undertaken only to control proptosis in a blind eye in 1969.[9] However, various studies have since emphasized the efficacy of surgical intervention as well as radiotherapy and chemotherapy.[18,22] Evaluating the size and the extent of this rare tumor was difficult before the introduction of MR imaging, and so

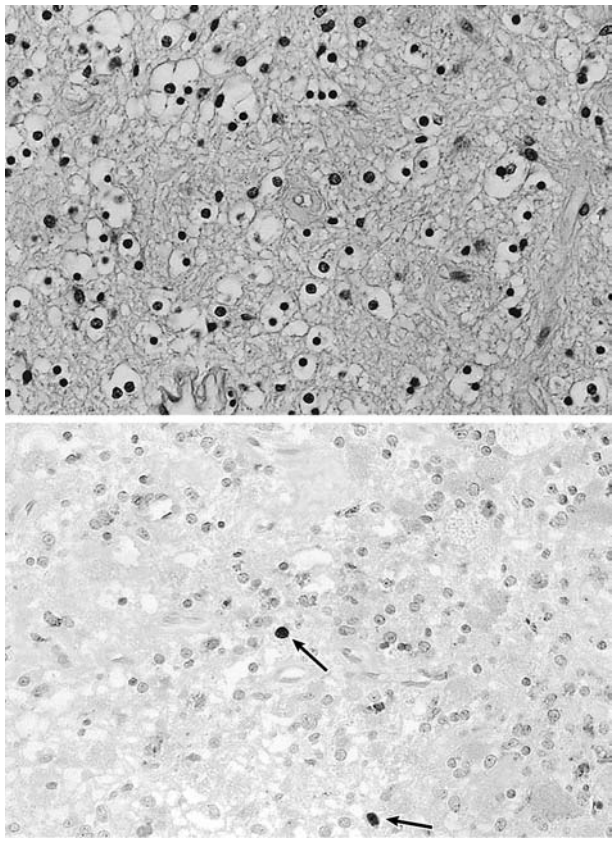

Fig. 3 Photomicrographs of the tumor specimen showing bipolar shaped pilocytic astrocytes. Cells with small round nuclei and a perinuclear halo are also occasionally seen (*upper*). MIB-1-positive cells are also visible (*arrows*). The MIB-1 index was 0.7% (*lower*). Hematoxylin and eosin stain (*upper*) and anti-MIB-1 immunostain (*lower*), original magnification ×200.

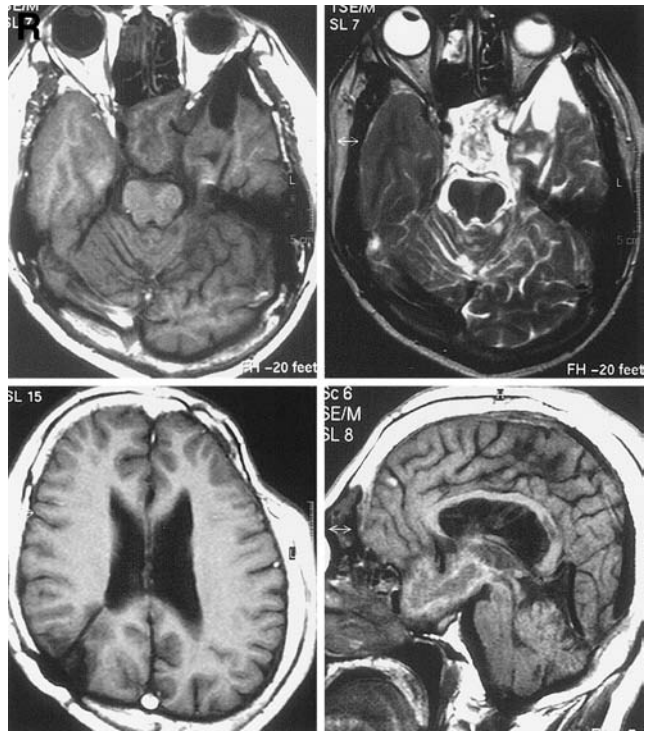

Fig. 4 Axial T_1-weighted (*upper left*) and T_2-weighted (*upper right*) magnetic resonance (MR) images 7 years after surgery showing no changes in the size or intensity of the tumor. Axial T_1-weighted MR image (*lower left*) showing reduction in the size of both lateral ventricles. Sagittal T_1-weighted MR image (*lower right*) demonstrating improvement in the cerebellar tonsillar herniation.

accurate observation of tumor size during long-term follow up was unlikely. Postoperative regression of a partially resected opticochiasmatic astrocytoma was demonstrated radiologically in 1984.[23] More than 30 such low-grade gliomas in the optic or opticochiasmal region have since been found to regress after surgical debulking or after biopsy, and all tumor diagnoses were confirmed histologically.[3,12,15,19,23] Spontaneous regression without surgical intervention has been also reported, but the diagnosis was based on radiological findings.[2,8,12,13,15,16,21] Regardless of the method used for diagnosis, subgroups of this entity are clearly self-limiting and can demonstrate spontaneous regression.

Several mechanisms underlying the regression or growth arrest of low-grade glioma have been suggested, including the activation of host immunity after surgical intervention, apoptosis or programmed cell death, and endocrine-assisted reduction in vascular engorgement of the tumor.[3,8,15] The present case was typical low-grade astrocytoma with low MIB-1 index. Cell death or specific vascular changes were not observed. Tumors that may show spontaneous regression or growth arrest have onset in childhood and regress at a young age. Soon after the diagnosis is established, the visual symptoms stabilize or improve and no aggressive treatments are performed. The unique aspect of our case was that growth arrest occurred in middle age after the progression of ophthalmological symptoms over 20 years beginning in childhood.

The prechiasmatic optic pathway is involved in 26–36% of cases and the chiasma, optic tracts, and/or third ventricle in 64–73% of cases.[22] Lesions located in the optic nerves occur frequently in children, whereas opticochiasmal lesions are commonly seen in adolescents.[3] Progression-free survival is

more common in tumors located in the optic nerve than in opticochiasmal locations.[6,10] Our case was sporadic and not neurofibromatosis type 1 (NF-1)-related. In general, low-grade gliomas related to NF-1 in this region show earlier regression than sporadic cases, and symptomatic sporadic optic gliomas show impaired vision more frequently and are more aggressive than NF-1-related optic gliomas.[20]

Any relationship between the pathogenesis of multiple arachnoid cysts and long-standing optic gliomas is not clear and could simply be incidental. The associated hydrocephalus is likely to be caused by intermittent obstruction by the tumor of the cerebrospinal fluid pathway at the craniocervical junction rather than direct obstruction of the foramen of Monro. Our patient's syncopal attacks disappeared immediately after the shunt surgery so that the cause of syncope was probably compression of the craniocervical junction by the herniated cerebellar tonsils. Such reversible tonsillar herniation due to supracerebellar arachnoid cysts has been treated by direct approach by craniectomy.[5]

Spontaneous improvement of visual acuity without remission is known[7,14] and our case may be in the same category. Growth arrest of the tumor might result in reduction of compression of the optic tract. Change in cerebrospinal fluid dynamics by placement of the ventriculoperitoneal shunt might be another reason for the improvement of visual acuity. The intracranial pressure was found to be slightly elevated intraoperatively and this could result in enlarged ventricles, leading to distortion and/or mechanical compression of the optic tract, which could have contributed to the deterioration of visual acuity. Ventriculoperitoneal shunting has not guaranteed the improvement of visual acuity in cases of gliomas of the optic tract.[6,9] We performed the shunt placement operation originally only to treat the syncopal attacks. Retrospectively, in view of the large size of the tumor and the absence of aggressive treatment, the improved quality of daily life of the patient was remarkable over many years. We emphasize that growth arrest of the tumor, even with a long history, should be recognized as a possible outcome, and that less aggressive treatment or an indirect approach to the tumor can potentially result in improvement of ophthalmological symptoms.

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