Optochiasmatic Cavernous Angioma with Rapid Progression after Biopsy Despite Radiation Therapy

Kwanag Wook Jo, M.D., Sang Don Kim, M.D., Ph.D., Eun Yong Chung, M.D., Ph.D., Ik Seong Park, M.D., Ph.D.
Department of Neurosurgery, Anesthesiology and Pain Medicine, St. Mary’s Hospital, College of Medicine, The Catholic University of Korea, Bucheon, Korea

We present a rare case of optochiasmatic cavernous angioma (CA) that progressed despite radiation therapy. A 31-year-old female patient presented with sudden loss of left visual acuity and right homonymous hemianopsia. Magnetic resonance imaging (MRI) revealed a suprasellar mass and findings compatible with a craniopharyngioma or an optic glioma with bleeding. An open biopsy was conducted using the transcranial approach, and histological examination revealed gliosis. During the one-year follow-up period, imaging suggested intratumoral bleeding and the mass continued to grow. We recommended re-operation, but the patient refused due to fear of surgery. Consequently, the patient received fractionated radiation therapy (3,000 cGy) to the parasellar area. Despite the radiotherapy, the mass continued to grow for the following 6 years. The final MRI before definitive treatment revealed a multifocalated, multistage hematoma with calcification in the parasellar area, extending into the third ventricle and midbrain. The patient ultimately underwent reoperation due to the growth of the tumor. The mass was completely removed with transcranial surgery, and the pathologic findings indicated a cavernous angioma (CA) without evidence of glioma. As shown in our case, patients may suffer intratumoral hemorrhage after biopsy and radiotherapy. This case places the value of biopsy and radiotherapy for a remnant lesion into question. It also shows that reaching the correct diagnosis is critical, and complete surgical removal is the treatment of choice.

Key Words: Optic Pathway - Cavernous Angioma - Radiotherapy.

INTRODUCTION

Cavernous angiomas (CAs) are usually found in the cerebral parenchyma, principally in the frontal and temporal lobes[10]. CAs are extremely rare and can develop within the optic nerve, optic tract, and optic chiasm. The presenting symptoms of optochiasmatic CAs are variable and include episodic visual symptoms, headache, retro-orbital pain, and nausea[11,12]. These symptoms often develop due to intratumoral hemorrhage. The goals of surgery for optochiasmatic CAs are prevention of rebleeding, optic decompression, and to preservation e and improvement of visual function[12]. In most cases, complete resections are conducted. In one reported case, stereotactic radiotherapy was done administrated for a remnant CA[13]. We present a rare case of optochiasmatic CA with rapid progression after biopsy despite radiation therapy.

CASE REPORT

A 31-year-old female patient presented with a sudden loss of visual acuity. Neuro-ophthalmologic examination revealed right homonymous hemianopsia and decreased left visual acuity. Only perception of hand movement was possible with the left eye. A fluid-fluid level was present on non-enhanced sagittal T1-weighted magnetic resonance imaging (MRI), suggesting intratumoral bleeding. A gadolinium enhanced MRI showed a nonenhancing, well-demarcated mass in the suprasellar subchiasmatic area. Both optic nerves were laterally displaced by the mass; however, the pituitary gland occupied the normal position and was normally oriented (Fig. 1). The mass was 15x15 mm, and the initial differential diagnosis included a craniopharyngioma and/or optic glioma with bleeding.

Exploration and biopsy of the lesion were performed via the periorbital approach. A hematoma cavity was noted in the optic chiasm, and no abnormal vessels or masses were identified. Simple decompression for the hematoma and a biopsy were conducted. Pathological examination indicated gliosis, and the patient's visual acuity improved after the operation. Finger counting at 30 cm was possible with the left eye. Unfortunately, the patient complained of sudden visual loss of the left eye nine...
months after the operation. Repeat MRI showed evidence of repeated bleeding, and the mass had extended to the left optic track (Fig. 2). We pursued conservative treatment because the patient refused the surgery. The visual loss improved, and finger counting at 30 cm was again possible. A third bleeding episode occurred five months later. The mass continued to grow progressively and appeared to be a malignant tumor with repeated bleeding; however, the patient refused to undergo surgery for tissue confirmation. After this, the patient received 3,000 cGy of fractionated radiation therapy to the parasellar area and was followed-up at a local clinic for five years. During that time, she lost vision in both eyes and was ultimately transferred to our hospital after developing gait disturbance and dementia. MRI at that time revealed hydrocephalus and a multilobulated, multistage hematoma with calcification in the parasellar area extending to the third ventricle and midbrain (Fig. 3). Due to the lack of response to radiotherapy and progressive visual loss, mass removal was selected decided for treatment. A multistage hematoma, with a so-called cobble stone appearance, and a whitish granulomatosus aggregated mass was noted found in the parasellar area (Fig. 4). Both optic nerves were displaced and tightly adherent to the mass. Although the mass was completely removed using the supraorbital approach (Fig. 5), a ventriculoperitoneal shunt was required to relieve the pressure associated with her hydrocephalus. The neuropathology report indicated angioma without evidence of glioma or sarcoma (Fig. 6). Although the patient's bilateral visual loss remained, her hydrocephalus symptoms, including dementia and gait disturbance, were improved.

**DISCUSSION**

The incidence of CA is 5-13% in all intracranial vascular malformations. CAs can occur anywhere in the brain and spinal cord, but have been reported most frequently at subcortical sites in the frontal and temporal lobes. However, CAs are extremely rare and can develop within the optic nerve, optic tract, and optic chiasm. The clinical presentation of CAs is determined by the location and presence or absence of hemorrhage. In overall, the most common presentation of symptomatic CAs is seizures, but ophtalmal CAs may show variable various manifestations. The
The most common symptoms are episodic visual symptoms as well as headaches, retro-orbital pain, and nausea, the so-called chiasmal apoplexy. The bleeding rate of intracerebral CAs is about 0.6%/yr and the rebleeding rate is 4.5%/yr. Hemorrhages can be extracranial or intracranial. Extracranial hemorrhages are common in cases where enlargement results in progressive symptoms. The reported symptomatic hemorrhage rate per year is less than 1% for all lesions.

The differential diagnosis of an optochiasmatic CA includes arteriovenous malformation, aneurysm, optic glioma, craniopharyngioma, other neoplasms, pituitary apoplexy, and infiltrative and inflammatory conditions.

MRI is the imaging modality of choice for the identification and follow-up of optochiasmatic CAs. MRI is a more sensitive and specific modality than CT for the diagnosis of CAs and the differential diagnosis of the causes of chiasmal apoplexy. The typical MRI pattern is as follows: a central focus of methemoglobin, a peripheral rim of hemosiderin, an adjacent focus of acute or subacute hemorrhage, and minimal to no enhancement. This focal heterogeneity occurs because of varying degrees of alteration of blood collected at different times.

The surgical indications for optochiasmatic CAs are to prevent rebleeding, relieve optic compression, and preserve and improve visual function. In the present case, the operative strategy employed at the first operation was a combination of simple biopsy and partial removal of the CA. For patients presenting with apoplectic symptoms, a procedure involving minimal manipulation of the optic apparatus already at risk should be the goal in order to minimize the prospects of an iatrogenic visual deficit. Therefore, evacuation of the hematoma, a biopsy to confirm diagnosis, and resection of part or all of the CAs would be advised only if the pathological anatomy appeared highly favorable. Biopsy and decompression are considered to have been an appropriate technique in the present case, because the initial impression was a suprasellar mass such as a craniopharyngioma or an optic glioma with bleeding. However, biopsy has been proposed as a factor that may potentially increase both the growth of the lesion and the likelihood of bleeding in optochiasmatic CAs. The CA remnant must be considered to have a risk of symptomatic bleeding, and some patients, especially those with good visual outcomes, will merit further treatment of the remnant. One of the case studies reviewed had good results with postoperative radiotherapy, without long-term recurrence. While stereotactic radiotherapy has been used to treat CAs throughout the brain with good results, there are recognized tolerance limits of the anterior visual pathway beyond which the risks of optic neuropathy increase unacceptably. Our patient did receive radiotherapy; however, the CA did not respond and showed progressive features. Existing data are insufficient to reach definitive conclusions regarding the efficacy of radiotherapy in such cases.

Because the present case exhibited repeated bleeding and progression despite radiotherapy, a direct and less extensive cranial approach, such as a subfrontal approach, was preferred for the removal of the mass. Possible surgical approaches included a variety of subfrontal approaches such as the eyebrow, orbitozygomatic, and more lateral approaches, including the pterional approach. The surgical approach must be dictated by the laterality of the lesion, i.e., for more lateral lesions, a pterional ap-
proach may be adequate. In the present case, a keyhole approach was used because the CA was a suprasellar lesion located in the midline.

CONCLUSION

Optochiasmatic CAs are very rare, and diagnosis may be difficult. The indications for surgical intervention are decompression of the optic nerve pathway and prevention of lesion growth and rebleeding. Diagnostic biopsy is not always helpful and may result in enlargement of the lesion. In addition, the efficacy of radiotherapy for treatment of the remnant lesion has not been adequately demonstrated. Obtaining the correct diagnosis is critical, and complete surgical removal is the treatment choice for optochiasmatic CAs.

References