

Case Report

Optochiasmatic Cavernous Angioma with Rapid Progression after Biopsy Despite Radiation Therapy

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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 multilobulated, multistage hematoma with calcification in the parasellar area, extending into the third ventricle and mid-brain. 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⁽⁰⁾. 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^(4,13). These symptoms often develop due to intratumoral hemorrhage. The goals of surgery for optochiasmatic CAs are prevention of rebleeding, optic decompression, and to preservation and improvement of visual function⁽¹²⁾. In most cases, complete resections are conducted. In one reported case, stereotactic radiotherapy was administered for a remnant CA⁽⁵⁾. 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 15×15 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 pterional 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

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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 granulomatous 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 needed 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 lobes¹⁰. 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 seizures¹¹, but optochiasmatic CAs may show have various manifestations. The

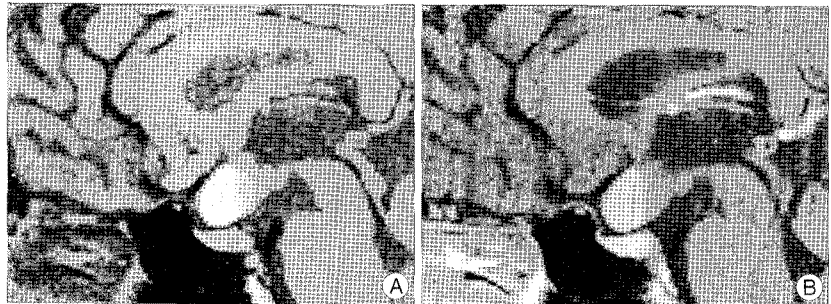


Fig. 1. A fluid-fluid level is noted on non-enhanced sagittal T1-weighted magnetic resonance images (MRI), suggesting intratumoral bleeding (A). A gadolinium enhanced MRI shows a nonenhancing, well-demarcated mass in the suprasellar subchiasmatic area. Both optic nerves are laterally displaced by the mass (B).

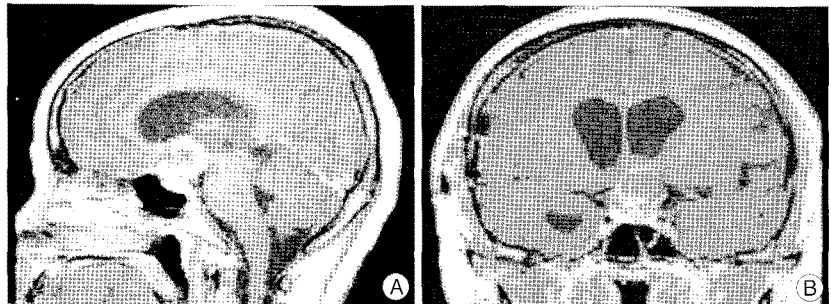


Fig. 2. Nine months after the operation, enhanced T1-weighted MRI shows growth of the mass lesion which now extends to the third ventricle roof and left optic track with evidence repeated bleeding (A : sagittal view, B : coronal view).



Fig. 3. Six years after the initial diagnosis, MRI shows a multilobulated parasellar mass, extending to the posterior superior part of the third ventricle. T1- (A) and T2-weighted MRI (B) shows multistage hematomas mixed with a high cellularity mass. A typical dark signal is noted on the gradient echo image (C) and there is little contrast enhancement (D).

most common symptoms are episodic visual symptoms as well as headaches, retro-orbital pain, and nausea, the so-called chiasmal apoplexy^{4,13}.

The bleeding rate of intracerebral CAs is about 0.6%/yr and the rebleeding rate is 4.5%/yr². Hemorrhages can be extralesional or intralesional. Extralesional hemorrhages are common in cases where enlargement results in progressive symptoms¹².



Fig. 4. Intra-operative photograph shows a multistage hematoma, with a so-called cobble stone appearance, and a whitish granulomatous aggregated mass in the parasellar area.



Fig. 5. The high-density mass around the parasellar area is not visualized on post-operative enhanced computed tomography.



Fig. 6. Histopathologic examination reveals lumens lined with endothelium and filled with thrombus. There are no elastic fibers or muscle in the walls of the lumen. These findings suggest an angioma (H & E $\times 40$).

The reported symptomatic hemorrhage rate per year is less than 1% for all lesions⁹.

The differential diagnosis of an optochiasmatic CA includes arteriovenous malformation, aneurysm, optic glioma, craniopharyngioma, other neoplasms, pituitary apoplexy, and infiltrative and inflammatory conditions².

MRI is the imaging modality of choice for the identification and follow-up of optochiasmatic CAs^{5,8,14}. MRI is a more sensitive and specific modality than CT for the diagnosis of CAs and the differential diagnosis of the causes of chiasmal apoplexy³. 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 enhancement⁶. 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 function¹². In the present case, the operative strategy employed at the first operation was a combination of simple biopsy and partial removal of the CA^{6,11}. 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 CAs¹². 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 remnant². One of the case studies reviewed had good results with postoperative radiotherapy, without long-term recurrence¹⁵. While stereotactic radiotherapy has been used to treat CAs throughout the brain with good results², there are recognized tolerance limits of the anterior visual pathway beyond which the risks of optic neuropathy increase unacceptably⁷. Our patient did receive radiotherapy; however, the CA did not respond and showed progressive features. Existing data are insufficient to reach definite 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 adequate²⁾. 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.

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