

Cavernous Malformation in the Anterior Optic Pathways Manifested by an Unilateral Sudden Loss of Vision -A Case Report and Review of the Literature-

Sandra Vuignier, M.D.¹; Takanori fukushima, M.D.²

¹ Department of Neurosurgery, Hokkaido University Graduate School of Medicine, Sapporo, Japan.

² Department of Neurosurgery, Duke Raleigh Hospital and Carolina Neurosciences Institute, North Carolina, United States.

Corresponding author:

sandravuignier@gmail.com

Sandra Vuignier, MD, Place du Gabon, 1971 Grimisuat, 08038563108.

Abstract

We report a case of a 11-year-old boy who presented with sudden loss of vision in the right eye. Two days before he experienced a mild headache when he woke up that vanished during the day. A cerebral computed tomography scan (CT) and an magnetic resonance imaging (MRI) revealed a lesion in projection to the right suprasellar cistern. At the operation a typical cavernous malformation (CM) with old and fresh hematoma was found in the right optic nerve (ON), optic chiasm (OC) and optic tract (OT) with a surrounding hemorrhage. Microsurgical total resection was done and the patient's vision much improved. This case report emphasizes the importance of a rapid diagnosis and reminds us that CM can also occur in young patients presenting as an unilateral sudden loss of vision.

Key words: Cavernous malformation; optic pathways; visual loss; total resection



This article is available from:
www.jneuro.com

Introduction

Lesions in the suprasellar region can interfere with optic nerve functions. The most common lesions are pituitary adenoma, craniopharyngioma, meningioma, carotid aneurysm and optic and hypothalamic gliomas [1, 10, 12, 13, 19]. In addition many uncommon neoplastic and infiltrative diseases affect this area, including germinoma, lymphoma, leukemia, chordoma, metastasis, nasopharyngeal carcinoma, sarcoidosis and histiocytosis X. CM of the ON, OC and OT is a very rare cause of visual deficits. Our case is particular because of the unusual location of the tumor, the unusual presentation of unilateral sudden loss of vision and the first pediatric age case in a series of 50 cases of cerebral cavernomas. Here we describe the case with the clinical and radiological characteristics and review the literature about CMs in the ON, OC and OT.

Case report

History and Examination

A 11-year-old male woke up with right dysopsia and headache. He went to a local eye clinic where left homonymous hemianopsia was highlighted. In the eye examination his right visual acuity was 0.03 and his left visual acuity was 1.2. Other neurological examinations showed no abnormalities and until that event the patient has always been healthy. There is no known family history of intracranial vascular malformations. Two days later he was sent to a local clinic where a cerebral CT and MRI were obtained that showed a heterogeneous mass lesion in the right suprasellar region (**Fig. 1**).

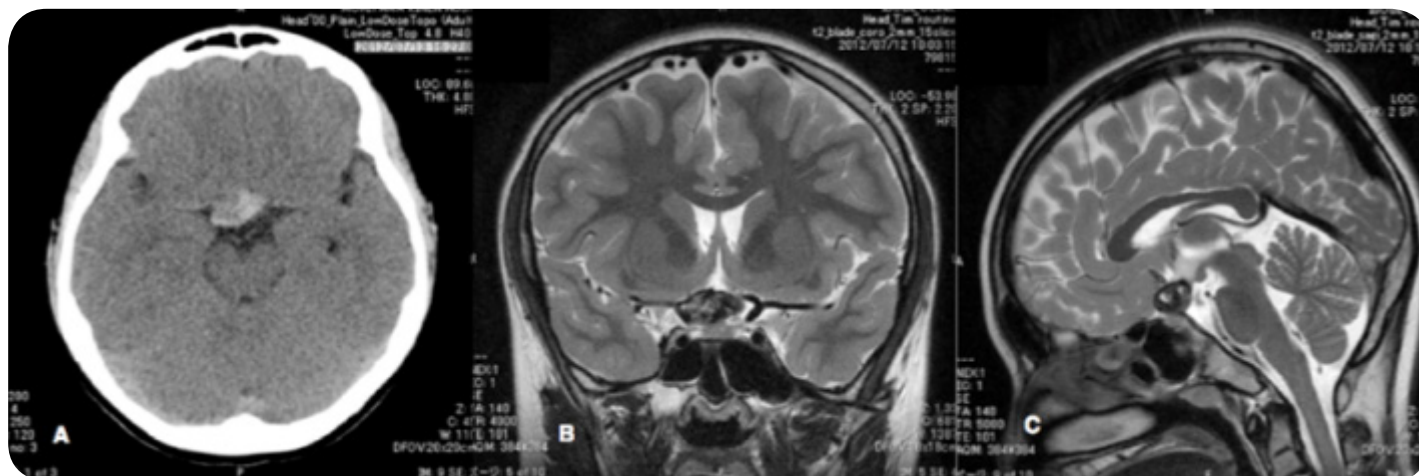


Fig. 1. **A.** Transversal preoperative cerebral CT showing inhomogenous hyperdense lesions on the right suprasellar cistern ; **B, C:** coronal and sagittal preoperative T2-weighted MRI highlighting the lesion on the suprasellar region.

At that time optic glioma, craniopharyngioma and germinoma were suspected. He was therefore referred to our hospital for surgical management. We clarified the risks and the benefits of the surgery by explaining that decompression of the right ON is necessary because of the risks of permanent blindness by compression of the mass lesion. Moreover the pathological diagnosis is essential and can only be made if the surgery is performed. The patient and his family accepted the surgery.

Surgery

Operation was performed using a typical right semicoronal skin incision and a frontotemporal craniotomy. Meticulous dissection of the sylvian fissure from distal to proximal was

done. The right middle cerebral artery (MCA), the internal carotid (IC), the ON, OC and OT were exposed. The right ON, OC and OT appeared with a brown and yellowish discoloration (**Fig. 2**).

The lesion consisted in «grapped shaped» vessels that were suggestive of a CM. Later we could confirm the CM by pathologist's examination. The first step of the decompression consisted in the relaxation of the right ON by the resection of the falciform ligament. Secondly, a longitudinal neurotomy on the most discolored part was carried out along ON fibers allowing us to approach the cavity of the angioma. With pre-cautious dissection around the tissues and repeated cutting and coagulation we could remove the lesion totally. Finally duralplasty was done with galeal tissue and the bone flap was repositioned in a cosmetic manner using Fixsorb plates. Skin

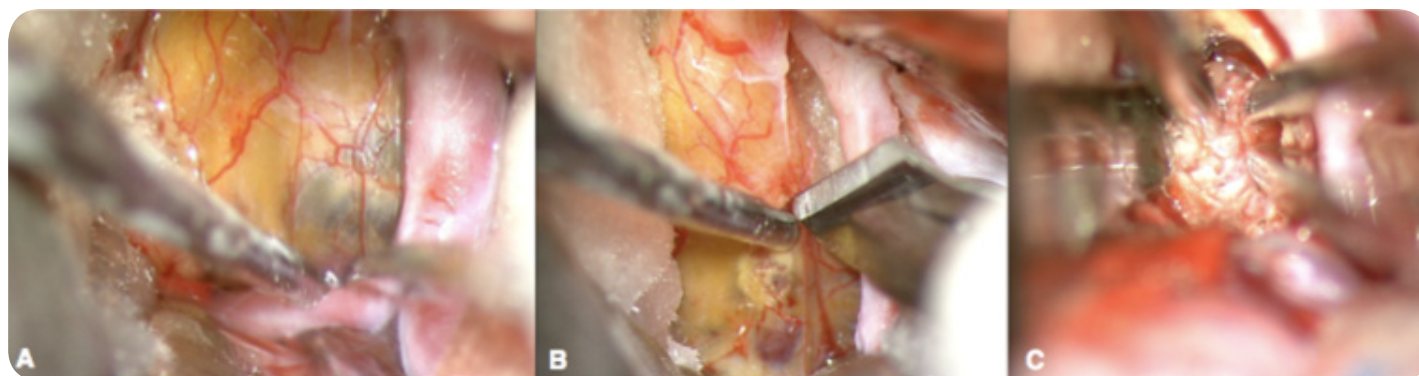


Fig. 2. **A:** intraoperative view showing the right ON with a brown and yellowish discoloration; **B:** longitudinal neurotomy; **C:** Repeated cutting and coagulation on the «grapped shaped» tumor.

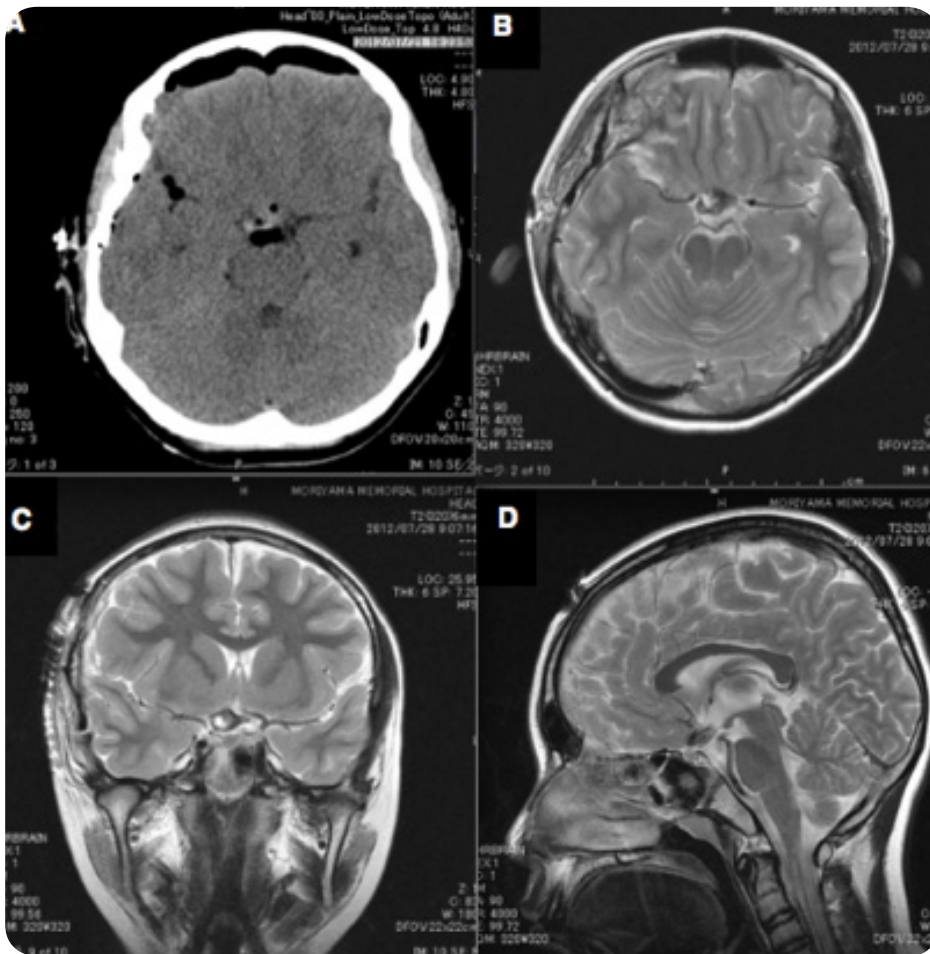


Fig. 3. **A:** transversal postoperative cerebral CT showing the complete removal of the tumor; **B, C, D:** transversal, coronal and sagittal postoperative cerebral T2-weighted MRI.

closure was done in layers and a subdural drain was placed to help ensure that all of the blood products are removed.

Postoperative courseThe postoperative course was uneventful with a dramatic improvement in the right vision. Left homonymous hemianopsia stayed stable. The postoperative cerebral CT and MRI showed total removal of the tumor (**Fig. 3**).

After 9 days the patient was discharged and able to go back to daily life activities.

Discussion

CMs are benign lesions consisting of clusters of thin-walled immature veins [4, 18]. They occur with an incidence of 0.3 to 0.5% and constitute 10 to 20% of all vascular malformations including arteriovenous malformations, venous malformations and capillary telangiectasis [11, 15]. Unlike the arteriovenous malformations, CMs have low flow and are angiographically occluded [2, 4]. These lesions may be observed in either sporadic or familial form [1, 2]. Clinical manifestations

of such tumors lie mainly in the 3rd and 4th decade [16]. Most lesions occur in the supratentorial compartment (80%) followed by infratentorial lesions (15%) and lesions within the spinal cord (5%) [5].

CMs arising from the anterior optic pathways are rare [1, 4, 12, 18]. In the literature we inventoried 60 cases reported by Liu J.K. et al in a study analysis of 65 cases of CM in the optic pathways and the hypothalamus [8]. 46% lied in the left side of the brain, 44% in the right side and 10% extended to both side. Only 2 cases (5%) involved the OC, ON and OT on the right hemisphere, as in our case. One case is from [20]. He described a case of a 43-year-old male with subacute visual loss who underwent surgery with gross total resection. The outcome nevertheless worsened. The other case is from Manz et al. [20]. He reported the case of a 30-year-old male with acute visual loss and headache who underwent a biopsy. The outcome improved.

There is no doubt that our case is rare. The unilateral sudden loss of vision, the excellent outcome after total removal as well as the young age make this case unique.

Conclusion

CMs involving the optic pathways are very rare, so the diagnosis may be difficult. A rapid management decision is essential to avoid permanent blindness. Microsurgical resection is the only one curative management and may provide functional improvement. During surgical excision, the gliotic interspace between the vascular malformation and the surrounding neural tissue provides a plane of cleavage for dissection which permits total excision without causing new deficits. Complete removal of the lesion is essential because of the possibility of recurrence [2-3, 6-7, 12, 14, 17]. This current case emphasises the importance of a rapid diagnosis and surgery of an anterior visual pathway CM. The boy arrived nearly blind and was discharged after 9 days with vision much better improved and could return to his daily life activities.

Aknowledge

The author wants to thank Dr Fukushima for giving her the case.

Conflict of interest

The author declares that they have no conflict of interest.

References

1. Christoforidis, GA., Bourekas, EC., Baujan, M., Drevelangas, A., Tzalonikou, M. Neuroradiology case of the day. *AJR Am J. Roentgenol* 2000; 175: 888-890.
2. Deshmukh, VR., Albuquerque, FC., Zabramski, JM., Spetzler, RF. Surgical management of cavernous malformations involving the cranial nerves. *Neurosurgery* 2003; 53: 352-357.
3. Ercan, O., Orhan, K., Kemal, Y., Serafettin, C. Optochiasmatic Cavernous Angioma. *Neurol Med Chir.* 2007; 47: 128-131.
4. Newman, H., Nevo, M., Constantini, S., Maimon, S., Kesler, A. Chiasmal Cavernoma: A rare cause of acute visual loss improved by prompt surgery. *Pediatric Neurosurgery* 2008; 44: 414-417.
5. Hassler, W., Zentner, J., Wilhelm, H. Cavernous angiomas of the anterior visual pathways. *J Clin Neuroophthalmol* 1989; 9: 160-164.
6. Hempelmann, RG., Mater, E., Schröder, F., Schön, R. Complete resection of a cavernous haemangioma of the optic nerve, the chiasm, and the optic tract. *Acta Neurochir.* 2007; 149: 699-703.
7. Lehner, M., Fellner, FA., Wurm, G. Cavernous haemangiomas of the anterior visual pathways. Short review on occasion of an exceptional case. *Acta Neurochir.* 2006; 148: 571-578.
8. Liu, J.K., Lu, Y., Raslan, A., Gultekin, S.H., Delashaw, J.B. Cavernous malformations of the optic pathway and hypothalamus: Analysis of 65 cases in the literature. *Neurosurg Focus* 2010; 29.
9. Manz, HJ., Klein, LH., Fermaglich, J., Kattah, J., Luessenhop, AJ. Cavernous hemangioma of optic chiasm, optic nerves, and right optic tract. *Virchows Arch A Pathol Anat Histol.* 1979; 383: 225-231.
10. Maruoka, N., Yamakawa, Y., Shimauchi, M. Cavernous hemangioma of the optic nerve. Case report. *J Neurosurg* 1988; 69: 292-294.
11. Moriarity, JL., Wetzel, M., Clatterbuck, RE., Javedan, S., Sheppard, JM., Hoenig-Rigamonti, K. et al. The natural history of cavernous malformations: A prospective study of 68 patients. *Neurosurgery* 1999; 44: 1166-1173.
12. Muta, D., Nishi, T., Koga, K., Yamashiro, S., Fujioka, S., Kuratsu, JI. Cavernous malformation of the optic chiasm: Case report. *Br J Neurosurg* 2006; 20: 312-315.
13. Ozer, E., Kalemci, O., Yücesoy, K., Canda, S. Optochiasmatic cavernous angioma: Unexpected diagnosis. Case report. *Neurol Med Chir.* 2007; 47: 128-131.
14. Pollock, BE., Garces, YI., Stafford, SL., Foote, RL., Schomberg, PJ., Link, MJ. Stereotactic radiosurgery for cavernous malformations. *J Neurosurg.* 2000; 93: 987-991.
15. Porter, RW., Detwiler, PW., Spetzler, RF., Lawton, MT., Baskin, JJ., Derksen, PT. et al. Cavernous malformations of the brainstem: Experience with 100 patients. *J Neurosurg.* 1999; 90: 50-58.
16. Robinson, JR., Awad, IA., Little, JR. Natural history of the cavernous angioma. *J Neurosurg.* 1991; 75: 709-714.
17. Son, DW., Lee, SW., Choi, CH. Cavernous malformation of the optic chiasm: Case report. *J Korean Neurosurg Soc.* 2008; 44: 88-90.
18. Werner, H., Zentner, J., Peterson, D. Cavernous Angioma of the Optic Nerve, case report. *Surg Neurol.* 1989; 31: 444-447.
19. Wilms, G., Lammens, M., Marchal, G., Van Calenbergh, F., Plets, C., Van Fraeyenhoven, L. et al. Thickening of dura surrounding meningiomas: MR features. *J Comput Assist Tomogr.* 1989; 13: 763-768.
20. Yoshimoto, T., Suzuki, J. Radical surgery on cavernous angioma of the brainstem. *Surg Neurol.* 1986; 26: 72-78.

Follow us:



Medicalia.org

Where Doctors exchange clinical experiences, review their cases and share clinical knowledge. You can also access lots of medical publications for free. **Join Now!**

<http://medicalia.ning.com/>

Publish with iMedPub

<http://www.imedpub.com>

✓ Journal of Neurology and Neuroscience (JNeuro.com) is a hybrid, peer-reviewed journal that considers articles concerned with any aspect of clinical neurosciences such as neurology, psychiatry and neurosurgery, as well as basic research on neuroscience where neurologists and neuroscientists publish together.

Submit your manuscript here:

<http://www.jneuro.com>