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Case Reports & Case Series

Cavernous malformation of the optic chiasm with continuous hemorrhage in a pregnant woman: A case report

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ABSTRACT

Background: Cavernous malformation of the anterior visual pathway is rare, especially in pregnant woman. Planning a treatment strategy with cross-disciplinary specialists is important.

Case description: A 27-year-old pregnant woman presented with acute hemorrhage around the right optic nerve and chiasm, manifesting as poor vision in both eyes. Examination revealed right-eye deteriorated acuity and bilateral temporal hemianopsia. Computed tomography showed an oval high-density mass in the suprasellar region. Gradient echo-based T2-weighted magnetic resonance imaging showed the lesion to be hypointense (possibly a hematoma) and mainly in the optic chiasm. Fluid attenuated inversion recovery imaging showed a bilateral optic tract surrounding the lesion, which enlarged over 1 week, increasing the loss of visual function. Five days after admission, she delivered a healthy > 2500-g baby by cesarean section (CS). Right frontotemporal craniotomy was performed 7 days after CS. Incision of the right optic nerve's lateral surface revealed clotted blood with abnormal vascular construction from the right side of the chiasm. We removed the hematoma and vascular lesion. Visual evoked potentials were detected only after optic chiasm decompression. Histological evaluation revealed a hematoma-like lesion with capsules and hemosiderin deposition, suggesting cavernous malformation. Her postoperative recovery was uneventful, with right visual acuity returning to normal, and her visual field not deteriorating any more.

Conclusion: Devising a treatment strategy with the obstetrician was important in this case to manage the hematoma and cavernous malformation safely.

1. Introduction

Cavernous malformation of the anterior visual pathway is rare, especially in adults. Hemorrhage associated with cavernous malformation in the optic chiasm represents a serious problem. Surgical management in such cases is based on the rationale that surgery poses less risk than the morbidity associated with possible subsequent hemorrhage [1].

Choosing a treatment strategy for cavernous malformation in pregnant women is difficult [2]. It has been reported that careful advance preoperative planning with cross-disciplinary specialists is needed [3]. It requires considering the safety of both mother and infant. We report a case of hemorrhage affecting the right optic nerve and optic chiasm originating from a cavernous malformation in a pregnant woman.

2. Case report

A 27-year-old pregnant woman had been in her usual state of health until 3 days before admission to our hospital, when mild headache and poor vision suddenly developed. She rested at home, but the symptoms persisted. On the third day, she visited a primary care ophthalmologist, who detected left homonymous hemianopsia (Fig. 1A). She was referred to our university hospital for further examination. After magnetic resonance imaging (MRI) revealed a hemorrhagic lesion in the optic chiasm, she was transferred to our department for surgical treatment.

Abbreviations: VEP, visual evoked potential; CS, cesarean section; MRI, magnetic resonance imaging; T1WI, T1 weighted imaging; T2WI, T2 weighted imaging

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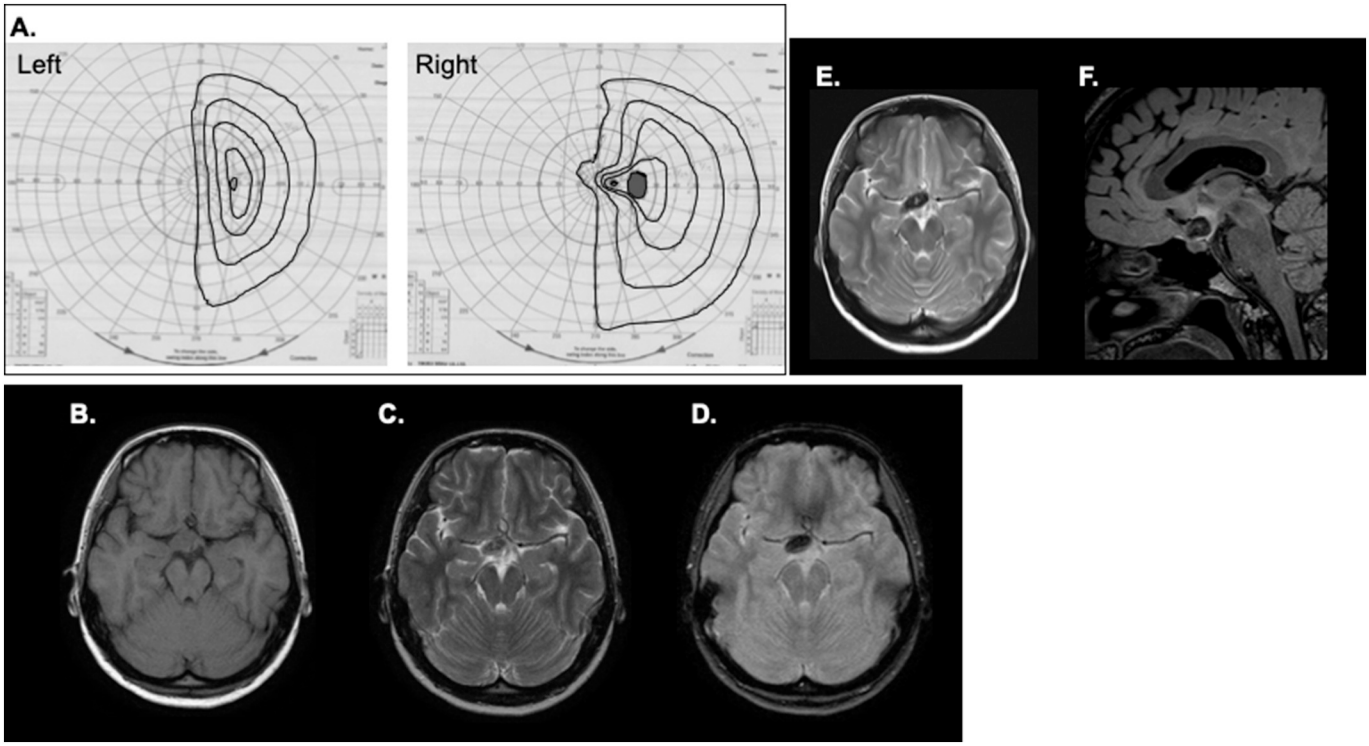

Fig. 1. Examinations before cesarean section. A: Goldmann's perimetry test performed at a primary care ophthalmologist showing left homonymous hemianopsia. T1-weighted imaging (B), T2-weighted imaging (C), and Gradient-echo T2-weighted imaging (D) showing the center of the mass as hypointense on admission. E: T2-weighted image on hospital day 4 showing the hematoma enlargement compared with initial images. F: Sagittal image of fluid attenuated inversion recovery on hospital day 4 showing hyperintensity area extending to the hypothalamus and pituitary stalk.

She had a history of subacute thyroiditis, for which she had been taking thyroid hormone replacement therapy. At the time of admission, she was at 34 weeks' gestation, with an estimated fetal body weight of 2300 g.

On examination, the patient appeared comfortable, although complaining of mild headache. She was alert and oriented, with no apparent neurological impairment except for left homonymous hemianopsia. Visual acuity was 20/286 in the right eye and 20/222 in left, both of which values were slightly worse than those in her premorbid state. Vital signs were within normal ranges. Total blood cell count showed mild anemia caused by hemodilution due to pregnancy. Growth hormone, prolactin, thyroid-stimulating hormone, adrenocorticotropic hormone, arginine vasopressin, and cortisol levels were normal. She had neither a café-au-lait macule nor neurofibromas.

Initial MRI showed an oval mass in the suprasellar region, mainly in the optic chiasm. Both T1-weighted (Fig. 1B) and T2-weighted (Fig. 1C) imaging (T1WI and T2WI, respectively) showed the center of the mass as hypointense. Gradient-echo T2WI also showed a hypointense mass (Fig. 1D). These findings led to a diagnosis of a subacute hematoma caused by hemorrhage within the suprasellar mass. Neither magnetic resonance angiography nor venography showed an aneurysm or vascular malformations.

After admission, glycerol was administered two or three times a day without surgical intervention because the fetal weight was < 2500 g. A second MRI session was undertaken on hospital day (HD) 4, revealing that the hemorrhagic mass had slightly increased (Fig. 1E). Sagittal fluid attenuated inversion recovery imaging showed hyperintensity extending to the hypothalamus and pituitary stalk (Fig. 1F). At 36 weeks' gestation, on hospital day 6, the fetal volume was > 2500 g, and she delivered a healthy baby via cesarean section. Her visual field test on HD 11 revealed scattered scotomas in the temporal field of the right eye (Fig. 2A), and her right visual acuity was 20/1000, which was worse than that at admission. Computed tomography of the brain on HD 9 revealed an enlarging hemorrhagic lesion (Fig. 2B). A third MRI session on HD 10 showed further (although slight) enlargement of the hematoma. Both T1WI (Fig. 2C) and T2WI (Fig. 2D) showed that the center of the mass was hyperintense and the right edge of the lesion was hypointense, suggesting a mixture of new and old hematoma. T1WI with gadolinium showed enhancement of the rim of the lesion (Fig. 2E). The lesion volume was 1.6 mm^3 as measured with i-Plan Cranial planning software (version 3.0; Brainlab, Munich, Germany), as previously described [4].

Right frontotemporal craniotomy was performed on HD 12, which was 7 days after the CS. We recorded visual evoked potentials (VEPs) with Neuromaster (Nihon Kohden, Tokyo, Japan) to determine visual function [5]. We also used the StealthStation S7 neuronavigation system (Medtronic, Minneapolis, MN, USA) for optical navigation. The right side of the optic chiasm was grossly enlarged with a black dome on the surface but no evidence of external compression (Fig. 3A). Incision of the lateral surface of the right optic nerve revealed clotted blood originating from the right optic nerve and the right side of the chiasm. Abnormal vascular construction was noted in the hematoma cavity (Fig. 3B).

The hematoma was evacuated, and as much of the vascular lesion and membrane-like lesion surrounding the hematoma as possible were removed. VEPs were not detected before incising the right optic nerve, but by the end of the surgery they developed as low, stable, reproducible signals (Fig. 4). Histological examination showed a hematoma-like lesion with capsules and hemosiderin deposition, suggesting a cavernous malformation.

The patient's postoperative recovery was uneventful. She developed temporary diabetes insipidus for a few days after surgery, without requiring oral anti-diuretics. Hydrocortisone was administered as steroid coverage during and after the surgery and was tapered off without withdrawal symptoms. The patient's visual function improved immediately following the surgery, with her visual acuity at 20/133 in the

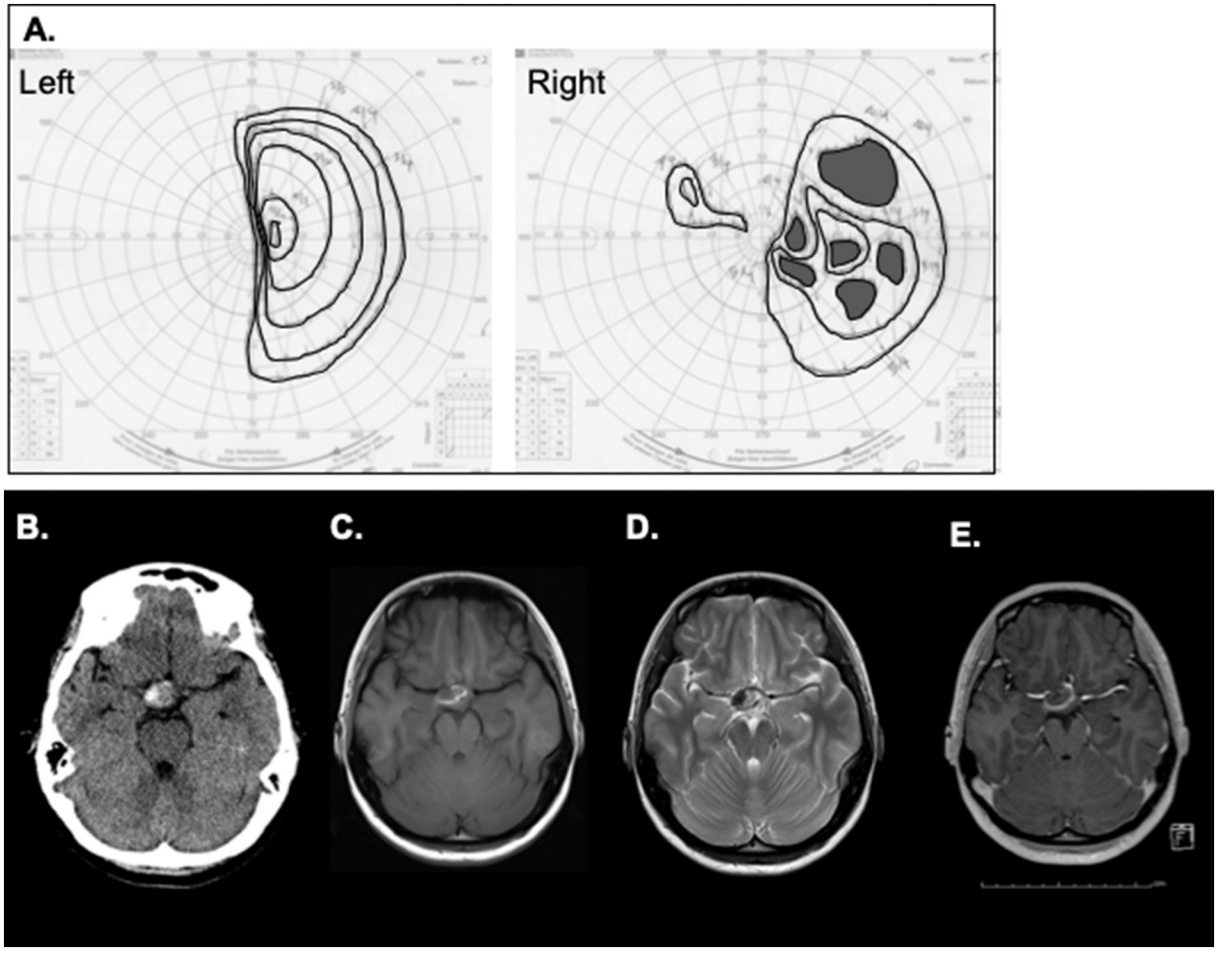

Fig. 2. Examinations before craniotomy. A: Goldmann's perimetry test shows left homonymous hemianopsia with scattered scotomas in the temporal field of the right eye. B: Computed tomography of the brain shows the increased size of the hemorrhagic lesion. T1WI (C) and T2WI (D) show the center of the mass as hyperintense and the right edge of the lesion as hypointense. E: T1WI with gadolinium shows enhancement of the rim of the lesion.

right eye and 20/100 in the left eye—levels equivalent to her premorbid readings. A visual field test showed that the scattered scotomas in the temporal field of her right eye had diminished (Fig. 5A). At 4 days after surgery, MRI showed complete disappearance of the hematoma (Fig. 5B) and of the hyperintense area that had extended to the hypothalamus and pituitary stalk (Fig. 5C).

3. Discussion

We treated an unusual case of cavernous malformation that originated in the optic chiasm. The patient was a pregnant woman who presented with continuous hemorrhage and developed left homonymous hemianopsia in her right eye, resulting in poor vision. Because several days had already passed from the onset of the symptoms, and she was in late pregnancy, we decided to treat her conservatively until

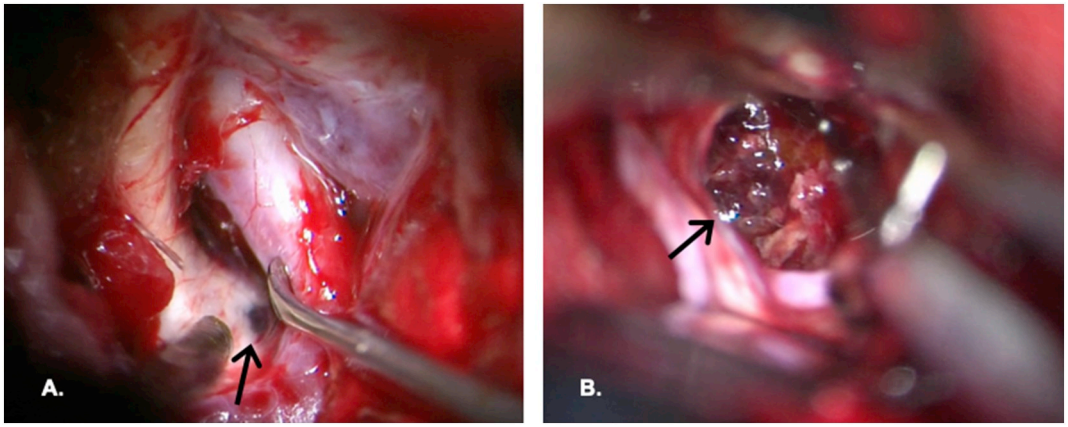

Fig. 3. Intraoperative views via right frontotemporal osteoplastic craniotomy. A: A black dome is present on the right surface of the grossly enlarged right optic nerve (arrow). B: A blood clot was exposed after incising the lateral surface of the right optic nerve (arrow).

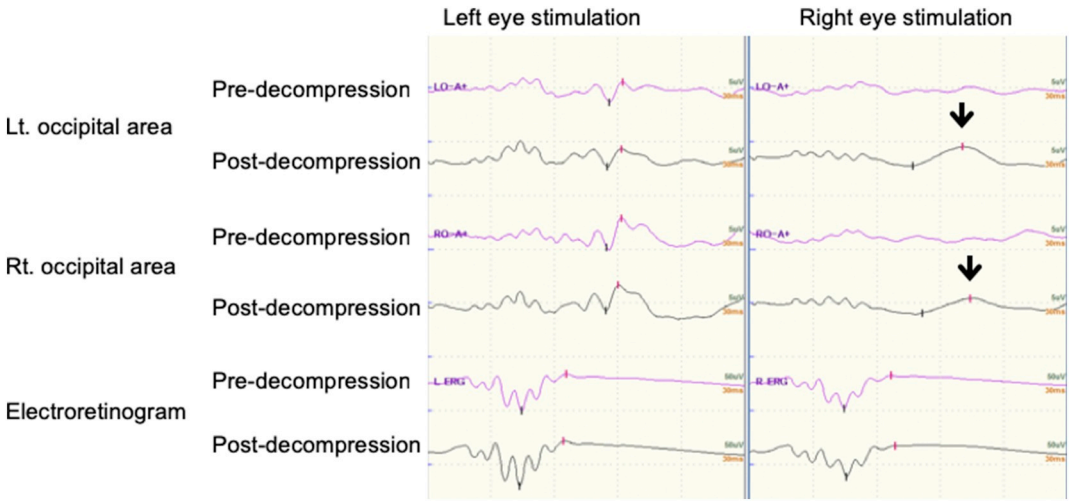

Fig. 4. Intraoperative record of visual evoked potentials (VEPs). A normal VEP pattern in the left eye stimulation both before (pink line) and after (black line) decompression. For the right eye stimulation, VEPs were detected only after decompression of the optic chiasm (arrow). Electroretinography pattern did not change between before and after decompression in either eye. Lt.: left; Rt.: right.

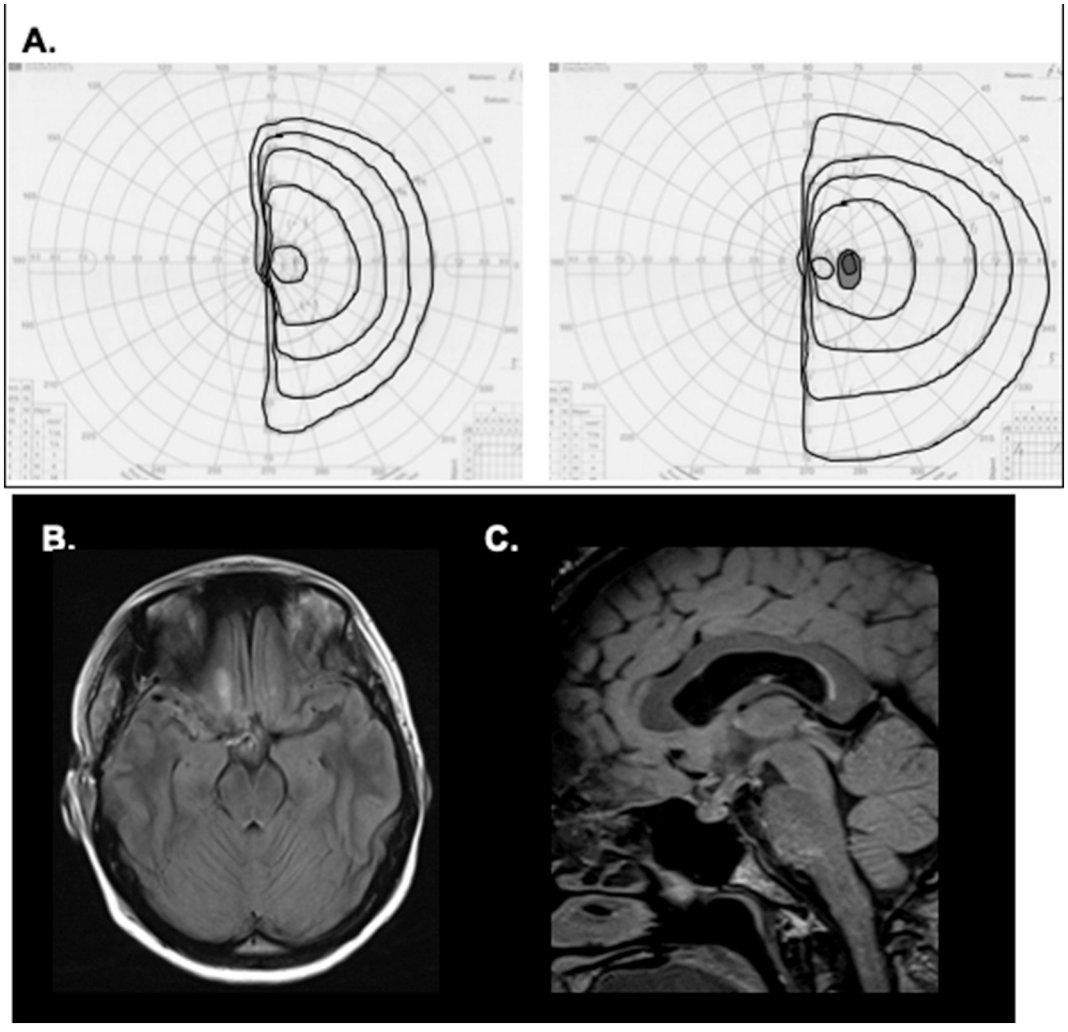

Fig. 5. Postoperative examinations. A: Goldmann's perimetry test shows the improved visual field. B: Axial STIR image shows almost complete removal of the hematoma. C: Sagittal fluid attenuated inversion recovery image shows disappearance of the hyperintense area extending to the hypothalamus and pituitary stalk.

CS delivery. Following the delivery of a healthy > 2500-g baby, her visual symptoms gradually worsened. She then underwent removal of the hematoma and lesion, after which the symptoms disappeared. VEPs were detected only after decompression of the optic chiasm. No permanent complications resulted from the surgical procedure.

An optic chiasm lesion with hemorrhage is rare. Several reports

have indicated that the causes of such hemorrhage are cavernous malformation, optic glioma, craniopharyngioma, meningioma, arteriovenous malformation, venous angioma, thrombosed aneurysm, rupture of anterior communicating artery aneurysm, pituitary apoplexy, and optic neuritis [1,6–9]. It is difficult to identify the cause based on the clinical signs and symptoms [10], so the diagnosis often depends on characteristic MRI findings associated with craniopharyngioma, meningioma, arteriovenous malformation, venous angioma, thrombosed aneurysm, pituitary apoplexy, and optic neuritis [1]. In our case, we could narrow the differential diagnosis using sequential MRI.

Mano et al. reported that cavernous malformation and optic glioma are difficult to differentiate because their clinical symptoms and time courses often overlap [11]. In our case, the patient's visual dysfunction was acute, so a cavernous malformation was suspected. Her symptoms, however, did not change during the 10-day period from her admission, which was atypical of that diagnosis. Clinical courses and MRI findings sometimes suggest cavernous malformation or glioma with hemorrhage, but pathological examination could exclude the possibility of an optic glioma. Miyahara et al. has reported that some types of vascular malformation might be involved in cases of chronic encapsulated intracerebral hematoma even if the cause of bleeding is unknown [12]. Hence, pathological examination in our case suggested the diagnosis of cavernous malformation based on finding a capsulated hematoma.

Cavernous malformation arising in the optic chiasm is rare, accounting for only < 1% of all cavernous malformations [13]. The annual rate of hemorrhage being associated with cavernous malformations is generally 0.7%–3.1% [14,15], although the natural history of cavernous malformation in the optic chiasm is not clear because most reported cases have been treated surgically [1]. Age at onset, female sex, and initial location are reportedly related to symptomatic hemorrhage [16]. Cavernous malformations are more likely to enlarge during pregnancy, and their incidence of bleeding is higher [17,18], although some reports suggested that the risk of hemorrhage during pregnancy is not increased [19,20]. In our case, there was continuous growth of the hematoma before and after delivery, suggesting that the natural history of cavernous malformation of the optic chiasm was different from that of the malformation in the cerebral cortex or brain stem.

Emergent surgery is generally recommended for optic apoplexy to prevent permanent visual impairment. Liu et al. reviewed 65 cases of cavernous malformations of the optic pathway and hypothalamus, among which the consensus was that surgical intervention should be performed within 24 h from the onset of visual loss [1,21]. In contrast, it has been suggested that craniotomy with hematoma removal performed 1 week after presenting with a bi-temporal visual field defect could improve visual function [22]. Moreover, radiographically identified hemorrhage exacerbation may not be associated with fluctuating visual symptoms [23]. Our patient had the visual symptoms for 3 days before admission, and they did not worsen during the 12 days afterward even though the hematoma enlarged. Hence, we decided to treat her conservatively until her general condition improved after the CS delivery. We then scheduled surgery for decompression of the optic chiasm.

Management of a pregnant woman with hemorrhage due to cavernous malformation remains debatable regarding both mother and child. Most pregnant women with cerebral cavernous malformation have safely undergone CS because of the concern of hemorrhage from the cavernous malformation [24,25], whereas there is no benefit of CS over vaginal delivery in a neurologically stable woman with hemorrhage caused by glioma [26]. In our case, however, CS was a necessary precaution, thereby preventing a hemorrhagic event during delivery because it was difficult to differentiate cavernous malformation from glioma before brain surgery. As for treating a cavernous malformation during pregnancy, brain surgery is rarely required except in life-threatening, rapidly progressive clinical situations [27,28]. From the anesthesiologist's point of view, the major physiological changes during pregnancy must be approached as risks during brain surgery [27]. In our case, we safely managed the hematoma and the cavernous malformation following conservative treatment and CS, suggesting that devising a treatment strategy in cooperation with the obstetrician is important.

Despite a critical location of the defect, excellent visual outcome after surgery could be achieved. Among patients with cavernous malformation of the optic pathway, visual acuity and visual field improved in 85% of those who underwent a decompression procedure with hematoma and lesion removal [1,22]. Total resection is the gold standard of surgical treatment to prevent re-bleeding from any residual tissue [21]. Decompression without complete resection also can improve visual function, but this strategy does not eliminate the risk of recurrent hemorrhage [1]. However, there have been no reports describing long-term follow-up data regarding re-bleeding rates or the durability of visual improvement following the surgery. In our case, we might not have achieved total resection of the lesion grossly because the posterior end of the lesion was difficult to visualize and thus remove unless we made an incision in the normal right optic tract. Therefore, the patient would have required a close, prolonged follow-up to ensure her visual function and that the lesion in the optic chiasm had been removed.

VEP monitoring was used to allow preservation of function. Several reports indicated its usefulness during endoscopic surgery to remove pituitary lesions [5,29], although the role of VEPs in the removal of optic nerve lesions remains unclear [30]. VEP data could not be obtained in the presence of severe preexisting visual dysfunction [31]. VEP monitoring has proved reliable for guiding surgical resection of optic lesions, even when visual function is significantly damaged [32]. In this case, VEPs could not be detected before incising the optic nerve because of severe visual impairment, but they appeared after hematoma removal. These findings suggested that VEPs could recognize and signal improved visual function resulting from surgical decompression of the optic tract.

We experienced a case of hemorrhage around the right optic nerve and optic chiasm originating from a cavernous malformation in a pregnant woman. We safely managed the hematoma and cavernous malformation with conservative treatment and CS. VEPs detected the improved visual function following surgical decompression of the optic tract.

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Declaration of Competing Interest

All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; expert testimony or patent-licensing arrangements) or non-financial interest (such as personal or professional relationships, affiliations, knowledge, or beliefs) in the subject matter or materials discussed in this manuscript.

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