Optic nerve sheath meningioma presenting as progressive visual disturbance during pregnancy: A case report

REINA UTSUGI, YOSHIKO OKITA, NAOKI KAGAWA and HARUHIKO KISHIMA

Department of Neurosurgery, Graduate School of Medicine, Osaka University, Suita, Osaka 565-0871, Japan

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Abstract. Meningiomas are often reported to be sensitive to progesterone, but it is not clear how pregnancy and childbirth affect this. A 41-year-old woman experienced two pregnancies and two deliveries. During the first pregnancy, her right visual acuity was impaired, but it recovered after delivery. However, during the second pregnancy, the right visual acuity was impaired again and did not recover after the second delivery. The magnetic resonance imaging revealed a right optic nerve sheath meningioma (ONSM). Surgical resection of the intracranial extension of the tumor was performed to prevent tumor invasion of the left optic nerve and optic chiasm. Pathological examination diagnosed meningioma with positive immunostaining for progesterone receptor. The present study provided clinical features of ONSM associated with pregnancy. ONSM may present with increased tumor growth and impaired vision with pregnancy.

Introduction

Optic nerve sheath meningiomas (ONSMs) are rare tumors that constitute 1-2% of all meningiomas and ~2% of all orbital tumors (1,2). ONSMs involve the optic nerve and extend to intracranial. In ONSMs having both an intraorbital and intracranial segment, clinical management to preserve visual function of optic apparatus adjacent to intracranial tumor may be challenging. Although meningioma is a benign tumor, it tends to be more aggressive during pregnancy. This is likely the result of tumor response to pregnancy-circulating hormones and homodynamic changes (3-7).

We report a patient with right visual impairment during two pregnancies, who was ultimately diagnosed with ONSM with positive immunostaining for progesterone receptor (PR). This study provides clinical features of ONSM associated with pregnancy. As ONSMs are originally slow-growing benign tumor, treatment strategy, combination of resection and advanced radiotherapy, based on long-term perspective seems to be necessary to prevent further spreading of the tumor not only to contralateral optic nerve but also to other intracranial structures involved in visual function.

Correspondence to: Dr Yoshiko Okita, Department of Neurosurgery, Graduate School of Medicine, Osaka University, 2-2 Yamadaoka, Suita, Osaka 565-0871, Japan
E-mail: yokita4246@gmail.com

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Case report

A 41-year-old woman felt visual impairment in the right eye, late in her first pregnancy. After the delivery, she received an ophthalmic examination and both visual acuity and field were normal. After 2 years, during late pregnancy with her second child, she became aware of slowly progressive visual deterioration in the right eye again. Two months after the second delivery, fundus examination showed swelling of the right optic disc. She was diagnosed with optic neuritis and received steroid pulse therapy. After 1 year, swelling of the right optic disc and right visual impairment had progressed. Her right eye's visual field was impaired, and she had developed lateral lower quadrantanopia. Furthermore, her right visual acuity worsened from 20/20 to 20/200 corrected visual acuity over the course of 1 year. She was admitted at our hospital and underwent magnetic resonance imaging (MRI). MRI revealed an intraconal tumor encasing the right optic nerve (Fig. 1A and B). The tumor extended through the optic canal to the right carotid artery and close to the optic chiasm (Figs. 1C and 2A). Surgical resection of the intracranial extension of the tumor was performed to prevent tumor invasion of the left optic nerve and the optic chiasm (Fig. 2B). After resection, the bilateral optic nerves were confirmed to be intact. The dura of the frontal base was coagulated and incised using a low-power bipolar system and micro dissection needle to prevent tumor invasion of the left optic nerve along the dura matter. The intraconal portion of the tumor encasing the right optic nerve was not resected to prevent compromising visual function. After intracranial tumor resection, her visual acuity maintained the presurgical value of 20/200 corrected visual acuity. The pathological findings suggested angiomatous meningioma, WHO Grade 1 with positive immunostaining for PR and negative for estrogen receptor (ER). Seven months after operation, her visual function did not deteriorate. MRI revealed no tumor size change during the 7-month observative follow-up period. There is a possibility that tumor size may be stable due to reduced influence of progesterone after delivery.

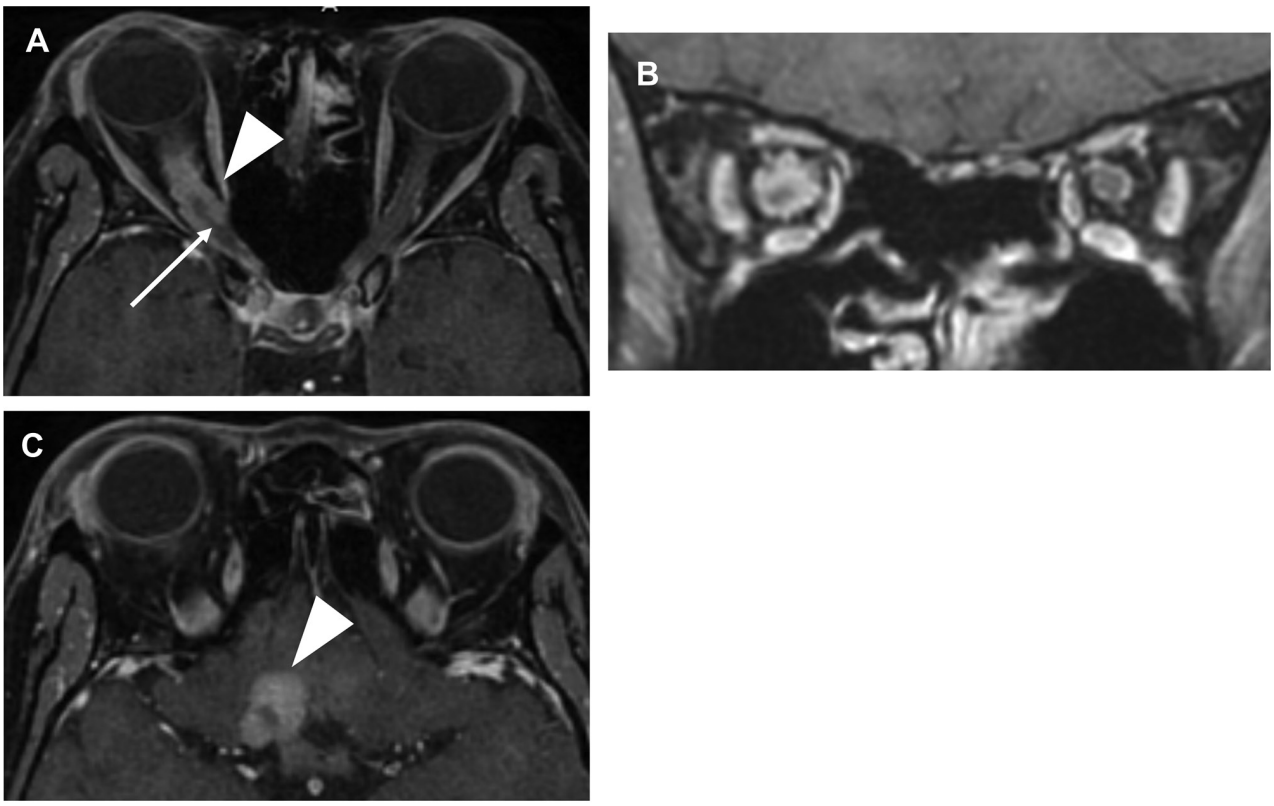

Figure 1. (A) Contrast-enhanced MRI showing thin right optic nerve (arrow) and ONSM encasing the right optic nerve (arrowhead). (B) MRI showing homogeneous enhancement with a 'tram-track sign' around the non-enhancing right optic nerve. (C) ONSM extends to the right carotid artery, close to the optic chiasm (arrowhead indicates meningioma). MRI, magnetic resonance imaging; ONSM, optic nerve sheath meningioma.

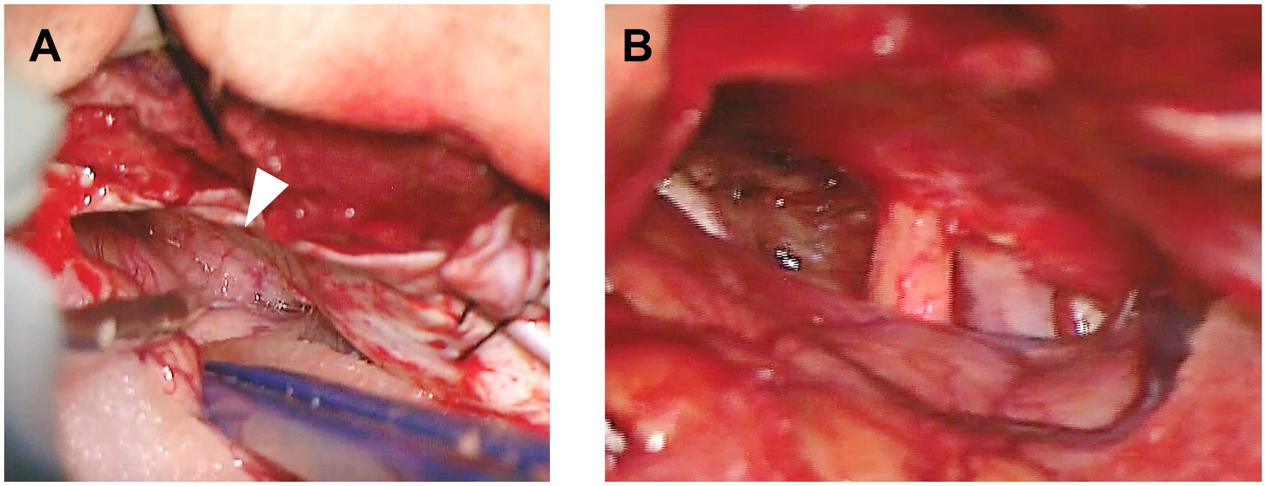

Figure 2. (A) Tumor (arrowhead) extended close to right optic nerve and carotid artery. (B) Intracranial extended tumor close to right optic nerve and carotid artery was resected. Right optic nerve and carotid artery were confirmed to be intact from tumor.

We have planned adding high-precision radiotherapy when the tumor grows to achieve our goals of tumor control and preservation of visual function.

Discussion

ONSMs are rare and slow-growing benign tumors. Meningiomas may grow during pregnancy because of hormone receptor expression and homodynamic changes (3-7). Progesterone is thought to be a significant factor for meningioma growth during pregnancy. It has been shown that meningioma growth is enhanced in the progesterone-dominated luteal phase of the menstrual cycle (8). Sex steroid of a meningioma was associated with pregnancy, and showed significant expression of PR; whereas no significant expression of ER was observed (9). Surgery after pregnancy was found to be more frequent in patients with PR positive meningioma (3). Our patient presented with transient visual impairment during

her first pregnancy, resolved after delivery, which reoccurred and gradually worsened with the second pregnancy. PR positive meningioma appears to be exacerbated by hormones circulating during pregnancy. However, it is controversial whether the growth of meningiomas during pregnancy is only due to the expression of PR. A study of the pathology of meningiomas during pregnancy found that the frequency of PR positivity during pregnancy was similar with the control group (6,10). Meningioma growth during pregnancy have the possibility to be affected by not only hormone receptor expression but also tumor location (6,11). Tumors adjacent to the optic apparatus may cause visual impairment during pregnancy. Pituitary adenoma, meningioma in skull base, and orbital schwannoma showed tumor growth and presented with visual impairment during pregnancy (11-15). In our case, the right optic nerve was encased by intraconal tumor and seemed to be vulnerable to visual impairment for tumor growth. On the other hand, in a few meningioma cases, symptoms subside and tumor size regresses after delivery (4,16,17). In our case, visual function and tumor size did not change during the 7-month observative follow-up period after delivery. We have planned adding high-precision radiotherapy when the tumor grows.

The primary treatment is complicated by the need to balance tumor curability and preservation of visual function. Conservative management is usually advocated when there is no significant visual dysfunction or progression of visual loss (18). Total resection is strongly associated with visual impairment because of pial vascular plexus damage, as well as incomplete resection of the tumor with local recurrence (1,19). As surgical intervention for ONSM has technical difficulty and postoperative deterioration of visual function, surgery for ONSM has largely been disregarded in favor of radiotherapy (20). Surgery is recommended for intracranial tumor extension that could cause loss of vision in the contralateral eye (1,18,19). In our case, the patient's right visual function gradually worsened during pregnancy but was still preserved. Our treatment goal was to not exacerbate right visual impairment and to preserve left visual function. Therefore, we not only resected the intracranial tumor segment, but also coagulated and incised the frontal base dura to prevent tumor invasion of the contralateral optic nerve.

Conventional radiotherapy has been widely used for ONSMs (21). Radiotherapy for ONSMs has the potential for late toxicity of the optic nerve and adjacent tissues, such as the optic chiasm, if included in the irradiation field. Recently intensity-modulated radiation therapy (IMRT) and stereotactic radiosurgery can result in significant visual improvements in selected cases, especially in cases with preservation of vision (22-24). In our case, the tumor had extended intracranially, close to the optic chiasm. Thus, surgery combined with new radiation modalities such as IMRT and stereotactic radiosurgery can be effective to avoid late radiotherapy toxicity of the optic nerve and the optic chiasm. In order to preserve contralateral visual function, our treatment plan involved resecting the portion of the tumor close to the optic chiasm and excluding the optic chiasm from the irradiation field. As ONSMs are originally slow-growing benign tumor, a careful treatment strategy based on long-term perspective, combining surgical resection and targeted radiotherapy, was necessary to prevent further spread of the tumor to the contralateral optic nerve and other intracranial structures.

In conclusion, this report describes a patient with ONSM who presented with progressive visual impairment during two pregnancies. ONSMs should be considered in cases of visual impairment that develop during pregnancy. Although ONSMs are typically slow-growing benign tumors, the treatment strategy should be based on preventing further spread of the tumor and preserving contralateral visual function. In ONSM patients who developed visual impairment, surgery may serve as an important but restricted adjuvant to radiotherapy.

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Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Authors' contributions

YO conceived and designed the study. RU acquired the data. RU, YO, NK and HK analyzed and interpreted the data and drafted the manuscript. YO and HK confirm the authenticity of all the raw data. All authors critically revised the manuscript for important intellectual content. All authors read and approved the final manuscript.

Ethics approval and consent to participate

The study was conducted according to the guidelines of the Declaration of Helsinki and approved by the Ethics Committee of Osaka University Hospital. Written informed consent was obtained from the patient.

Patient consent for publication

Written informed consent was obtained from the patient for publication of the data and images in this case report.

Competing interest

The authors declare that they have no competing interests.

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