



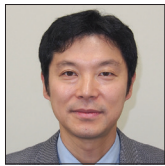
Case Report

Ocular neuromyotonia caused by a recurrent sphenoidal ridge meningioma

Yuto Shingai¹, Hidenori Endo^{1,2}, Toshiki Endo¹, Shin-ichiro Osawa¹, Kuniyasu Nizuma^{1,3,4}, Teiji Tominaga¹

¹Department of Neurosurgery, Graduate School of Medicine, Tohoku University, ²Department of Neurosurgery, Kohnan Hospital, ³Department of Neurosurgical Engineering and Translational Neuroscience, Graduate School of Biomedical Engineering, ⁴Department of Neurosurgical Engineering and Translational Neuroscience, Graduate School of Medicine, Tohoku University, Sendai, Miyagi, Japan.

E-mail: Yuto Shingai - yutos0123@gmail.com, *Hidenori Endo - hideendo@gmail.com, Toshiki Endo - endo@nsg.med.tohoku.ac.jp, Shinichiro Osawa - osawa@nsg.med.tohoku.ac.jp, Kuniyasu Niizuma - niizuma@nsg.med.tohoku.ac.jp, Teiji Tominaga - tomi@nsg.med.tohoku.ac.jp



*Corresponding author:

Hidenori Endo,
Department of Neurosurgery,
Kohnan Hospital, Sendai,
Miyagi, Japan.

hideendo@gmail.com

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ABSTRACT

Background: Ocular neuromyotonia (ONM) is a rare ocular motility disorder characterized by involuntary paroxysmal extraocular muscle contraction and is caused by radiation therapy, vascular compression, and inflammatory disease. This study includes a rare case of ONM caused by a recurrent meningioma.

Case Description: A 56-year-old man presented with diplopia due to the right oculomotor nerve palsy caused by a sphenoidal atypical meningioma, with improved symptoms after initial surgery. During the next 7 years, he underwent local radiation therapy, second surgery, and Gamma Knife radiosurgery to control the tumor's repetitive recurrence around the right anterior clinoid process. After these treatments, residual tumor was controlled for the next 3 years. However, 3 months after his last visit, he started to suffer from the right ONM and visual disturbance. The magnetic resonance imaging results revealed a rapid growth of the posterior part of the residual tumor, involving the right oculomotor nerve. The third tumor resection was performed to prevent further aggravation of the symptoms. Decompression of the right oculomotor nerve was achieved, and ONM disappeared immediately after surgery.

Conclusion: If nerve compression by the tumor is clearly indicated with the neuroradiological assessment, surgical intervention is the treatment of choice to improve ONM.

Keywords: Diplopia, Meningioma, Ocular neuromyotonia, Radiation, Surgery

INTRODUCTION

Neuromyotonia is a condition of spontaneous and continuous muscle fiber activity of the peripheral nerve origin.^[7] Ocular neuromyotonia (ONM) is a rare ocular motility disorder characterized by paroxysmal and involuntary extraocular muscle contractions,^[1] occurring due to radiation therapy of the parasellar region,^[11] inflammatory or autoimmune disease,^[6] or nerve compression by the vascular structure.^[2,3,5,9,12,13] However, no cases have been reported in which ONM occurred due to an intracranial tumor. In this study, we describe a rare case of ONM caused by a recurrent meningioma, which was recovered after tumor resection.

CASE REPORT

A 56-year-old man presented with diplopia due to the right oculomotor nerve palsy. The magnetic resonance imaging (MRI) results revealed a large sphenoidal tumor [Figure 1a]. The patient

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underwent tumor resection, and the pathological diagnosis revealed an atypical meningioma. Diplopia improved after tumor resection and only a slight adduction disturbance of the right eye persisted. During the next 7 years, he underwent local radiation therapy at a total dose of 50 Gy, second tumor resection, and Gamma Knife (GK) radiosurgery to control the tumor's repetitive recurrence around the right anterior

clinoid process. After the GK radiosurgery, residual tumor was controlled for the next 3 years [Figure 1b].

Three months after his last visit, he started to suffer from intermittent diplopia. MRI revealed a rapid growth of the posterior part of the residual tumor [Figure 1c], and heavily T2-weighted imaging showed the involvement of the right oculomotor nerve within the tumor [Figure 1d]. His symptom was intermittent involuntary diplopia, an involuntary inward deviation of his right eye, which resolved within 1 min [Figure 1e, Video 1]. The symptom was mostly triggered by tight eye closing, which frequently occurred every day. The ophthalmological examination of the right eye showed restricted eye movement during upward and medial gaze [Figure 2a]. The ophthalmologist diagnosed this intermittent involuntary symptom as ONM. The results of the ophthalmological examination also revealed worsening of the right vision and nasal hemianopia [Figure 2b].

The third tumor resection was performed through the right pterional approach to prevent further visual disturbance aggravation. After the removal of the tumor occupying the right retrocarotid space, the oculomotor nerve was found within the tumor and was preserved [Figure 3a]. The oculomotor nerve was tightened by the surrounding tumor. We found no neurovascular contact between the oculomotor nerve and vascular structures. Most of the tumor was removed, except for the part that was tightly adhered to the right optic nerve and optic chiasm, achieving a sufficient neural decompression [Figure 3b]. The tumor was pathologically diagnosed as an atypical meningioma with a MIB-1 labeling index of 12.4%.

ONM disappeared immediately after surgery. A transient worsening of the right oculomotor nerve palsy occurred, which gradually recovered in the preoperative level [Figure 3c, Video 2]. The right visual disturbance and nasal hemianopia continued after surgery. The postoperative

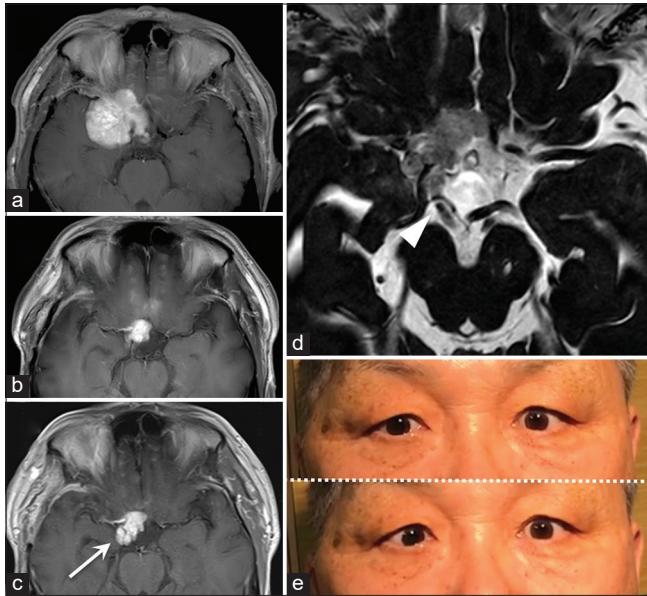


Figure 1: (a) T1-weighted imaging with gadolinium enhancement at the initial presentation showing a large right sphenoidal tumor. (b) T1-weighted imaging after Gamma Knife radiosurgery for recurrent tumor showing a residual tumor around the right anterior clinoid process. (c) T1-weighted imaging during the occurrence of ocular neuromyotonia (ONM) showing a rapid growth of the posterior part of the tumor (arrow). (d) Heavily T2-weighted imaging during the occurrence of ONM showing the right oculomotor nerve involvement (arrowhead). (e) ONM symptom. The position of the eyes in the normal status (upper) and ONM (lower).

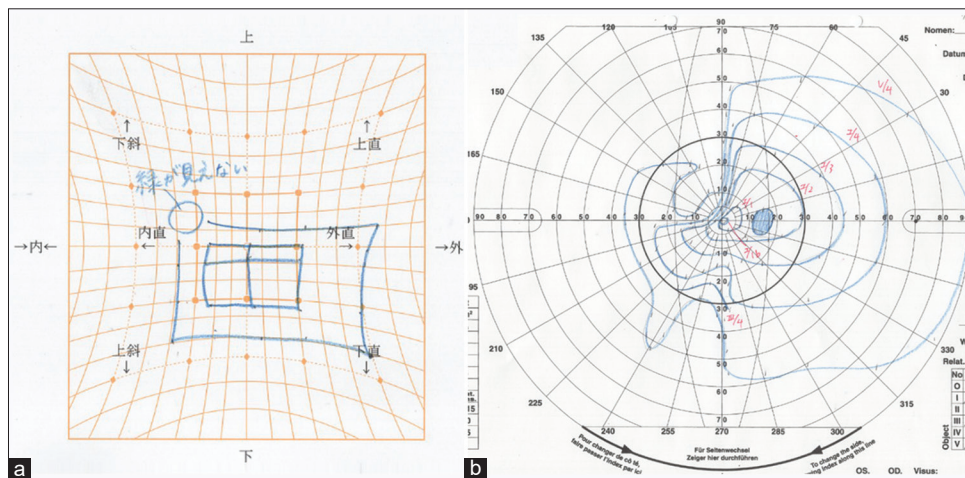


Figure 2: (a) Hess chart showing restricted right eye movement during upward and medial gaze. (b) Visual field examination revealing nasal hemianopia.

course was uneventful. The patient was discharged 15 days after surgery and he remained free from ONM until the last follow-up visit (9 months after surgery).

DISCUSSION

Paroxysmal ocular movement abnormalities can be caused by the damage of the cranial nerves, such as the oculomotor, trochlear, and abducens nerves. Among them, the symptom associated with the trochlear and oculomotor nerves or the abducens nerve is known as superior oblique myokymia^[11] and ONM, respectively.^[2,9]

The exact mechanism of ONM remains unclear. Several mechanisms have been proposed in recent literatures. One of the hypotheses is that nerve injury leads to segmental demyelination, resulting in local ephaptic transmission.^[12,13] Segmental demyelination can be triggered by radiation therapy, inflammatory or autoimmune disease, or physical compression by the vascular structures. Ephaptic

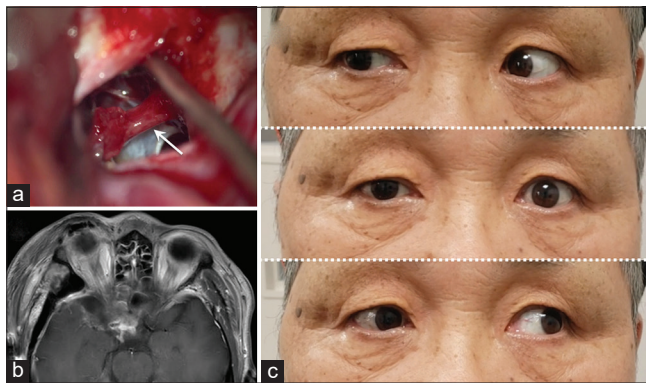


Figure 3: (a) An intraoperative view of the right oculomotor nerve after tumor removal (arrow). (b) T1-weighted imaging with gadolinium enhancement after surgery showing that most of the tumor was removed. (c) Eye movement after surgery showing a resolution of the right ONM with only a slight disturbance of the inward movement of the right eye.

transmission by neurovascular compression is a widely accepted etiology in the trigeminal neuralgia, hemifacial spasm, and superior oblique myokymia.^[3,4]

In this case, previous radiation therapy might have potentially impaired the right oculomotor nerve. Subsequently, nerve compression by the tumor recurrence overlapped. Considering the improvement of ONM after surgery, the tumor compression of the right oculomotor nerve was the main cause of his symptom, which can be explained by the improvement of trigeminal neuralgia or hemifacial spasm after microvascular decompression.

ONM has been treated with medication or surgery. Seven cases with ONM have been reported in the English literature, in which the relationship between the symptom and nerve compression was clearly indicated [Table 1]. Five of the eight cases were treated with medication (carbamazepine), out of which only two cases showed improvement. Moreover, three of the eight cases underwent surgical intervention, out of which two underwent open surgery and one underwent endovascular treatment, all of which showed resolution of symptoms. Inoue *et al.* reported a case of ONM, in which the oculomotor nerve was pinched by the posterior cerebral artery and superior cerebellar artery.^[3] The patient was administered carbamazepine, but suffered from side effects. Finally, they performed microvascular decompression and achieved complete resolution of the symptoms. Park *et al.* described a case of abducens ONM caused by a large aneurysm in the cavernous portion of the left internal carotid artery.^[9] They performed coil embolization of the aneurysm and achieved resolution of ONM 4 months after the treatment. The delayed resolution of the symptoms might be due to the reduced pulsation stress after the coil embolization, frequently seen in the endovascular treatment for internal carotid artery aneurysm with oculomotor nerve palsy.^[8] The resolution of ONM after surgical intervention was an offshoot of the treatment for the recurrent tumor or cerebral aneurysm in two of the three cases, including our case. However, these

Table 1: The cases of ocular neuromyotonia caused by nerve compression.

| Author, year | Age/sex | Affected nerve | Compression structure | Radiation | Treatment | Outcome |
|--------------------------------|---------|----------------|-----------------------|-----------|------------|---------------|
| Hashimoto <i>et al.</i> , 2016 | 40/F | Oculomotor N. | Vessel (SCA) | None | AED | Not effective |
| Cruz <i>et al.</i> , 2013 | 75/F | Oculomotor N. | Vessel (Pcoma) | None | AED | Effective |
| Inoue <i>et al.</i> , 2012 | 62/F | Oculomotor N. | Vessel (PCA and SCA) | None | MVD | Effective |
| Versino <i>et al.</i> , 2005 | 45/F | Oculomotor N. | Vessel (PCA) | None | AED | Effective |
| Tilikete <i>et al.</i> , 2000 | 67/M | Oculomotor N. | Vessel (BA) | None | AED | Not effective |
| Ezra <i>et al.</i> , 1996 | 60/M | Oculomotor N. | Aneurysm (ICA) | None | AED | Not effective |
| Park <i>et al.</i> , 2008 | 73/F | Abducens N. | Aneurysm (ICA) | None | Coil + AED | Effective |
| This case | 65/M | Oculomotor N. | Tumor (meningioma) | 50 Gy | Removal | Effective |

N: Nerve, SCA: Superior cerebellar artery, Pcoma: Posterior communicating artery, PCA: Posterior cerebral artery, BA: Basilar artery, ICA: Internal carotid artery, AED: Antiepileptic drug, MVD: Microvascular decompression

results suggest surgical intervention as the treatment of choice if nerve compression by the tumor or vascular component is clearly indicated with the neuroradiological assessment.^[10]

CONCLUSION

ONM is a rare ocular motility disorder caused due to nerve compression by an intracranial tumor. If nerve compression is clearly indicated with the neuroradiological assessment, then surgical intervention is the treatment of choice.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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