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CASE REPORT

Recovery of visual field in early treated optic nerve sheath meningioma: A case report

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Abstract

We report the diagnosis and treatment process of a patient who underwent CyberKnife radiotherapy with diagnosis of optic nerve sheath meningioma (ONSM) to emphasize visual field recovery with an early treatment. A 46-year-old woman presented with puffiness, discomfort, and enlargement in right eye. Right sided proptosis and optic disc edema were detected as accompanying findings. Visual acuity was 20/20 in both eyes. Blind spot enlargement and mild peripheral constriction were found on the right side of visual field test. The diagnosis was made on the basis of clinical and radiological findings. Magnetic resonance imaging demonstrated an ONSM. The lesion was treated by three fraction stereotactic CyberKnife radiation therapy with dose of 19.5 Gy. Visual acuity was preserved and visual field was completely recovered after 6 weeks of follow-up. Due to its typical clinical and radiological findings, ONSM can be diagnosed without tissue biopsy. In progressive cases, optic nerve functions may recover by CyberKnife radiation therapy performed before development of significant visual loss. However, patients should be observed for radiation complications.

Keywords: Optic nerve sheath meningioma; radiation therapy; visual field.

Optic nerve sheath meningiomas (ONSM) are rare benign tumors of the central nervous system. ONSM usually arise from the intraorbital part of the optic nerve sheath and account for approximately 2% of all orbital tumors and 1–2% of all meningiomas.[1] Most of the lesions are unilateral. The rare bilateral ONSM tend to occur in patients with neurofibromatosis Type 2.[2] Typically, these tumors affect middle-aged women with an average age of 41 years.[3] They also tend to increase in size throughout pregnancy and during the menstrual cycle.

The pathognomonic sign for the clinical presentation of ONSMs; known as Hoyt-Spencer triad, is composed of progressive visual loss, optic nerve atrophy, and the presence of opticociliary shunt vessels. However, only a minority of patients has this classical presentation.[4,5] The diagnosis of ONSM relies usually on clinical and imaging findings.[5] Meningiomas typically display intense homogeneous enhancement with gadolinium-enhanced fat-suppression T1-weighted pulse sequences in magnetic resonance imaging (MRI).[6] There is no consensus of management of

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such slow-growing tumor. Visual outcome indeed seemed better in radiotherapy than neurosurgery with less late side effects.[7–9] Thus, radiotherapy is becoming the first-choice procedure in this indication.[7–10] Nevertheless, the perfect time of management remains unclear and some practitioner tend to observe small tumors without life-threatening evolution and a preserved functional visual acuity. [4] We reported the diagnosis and treatment process of a 46-year-old woman, who underwent CyberKnife radiation therapy with diagnosis of ONSM. The aim of this report is to emphasize the benefit of this treatment on optic nerve function, performed before reduction in visual acuity.

Case Report

A 46-year-old woman presented with history of puffiness in the right upper eye lid and enlargement, discomfort, and transient visual obscuration in the right eye for about 1 year (Fig. 1). The patient has remained undiagnosed although she visited several ophthalmologists. She was treated with non-steroidal anti-inflammatory drugs but no improvement observed. Visual acuity was 20/20 in both eyes. She had a right relative afferent pupillary defect. Anterior segment examination was within normal limits and her ocular motility was full. Fundoscopic examination revealed right optic disc edema with indistinct borders (Fig. 2). The left eye examination was normal. Hertel exophthalmometric measurements were 15 mm in the right eye and 12 mm in the left eye. The result was defined as a right-sided proptosis. On account of this, the patient was evaluated with MRI and visual field test. Meanwhile, thyroid related hormones were measured within normal limits. Humphrey visual field 30–2 test showed a blind spot enlargement and mild peripheral constriction (Fig. 3). In MRI study, a lesion surrounding the right optic nerve detected in intraconal area, measuring 28×26×20 mm. The lesion appeared as isointense to brain and optic nerve tissue on T1-weighted images (Fig. 4a) and slightly hyperintense on T2-weighted images (Fig. 4b). On T1-weighted images with fat saturation after intravenous administration of paramagnetic substance (gadolinium), the mass presented a homogenous intense enhancement suggesting in appearance a "tram-track" around the hypointense optic nerve (Fig. 4c).[6] On the coronal images, this is seen as a "doughnut sign" (Fig. 4d).[11]

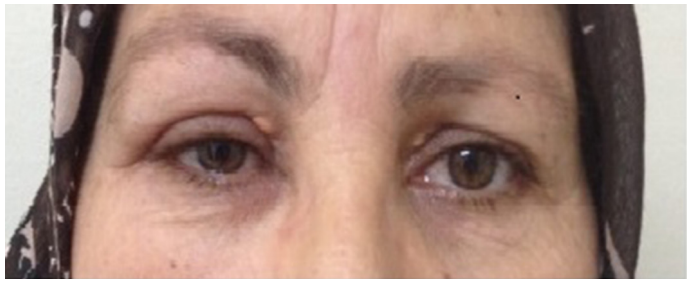

Fig. 1. Appearance of the patient with right mild upper eyelid edema before the treatment.

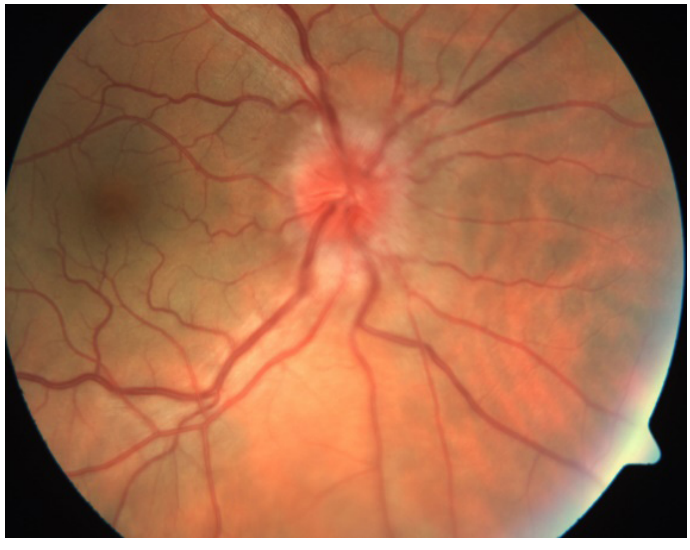

Fig. 2. Right fundus: Indistinct optic disc borders and optic disc edema.

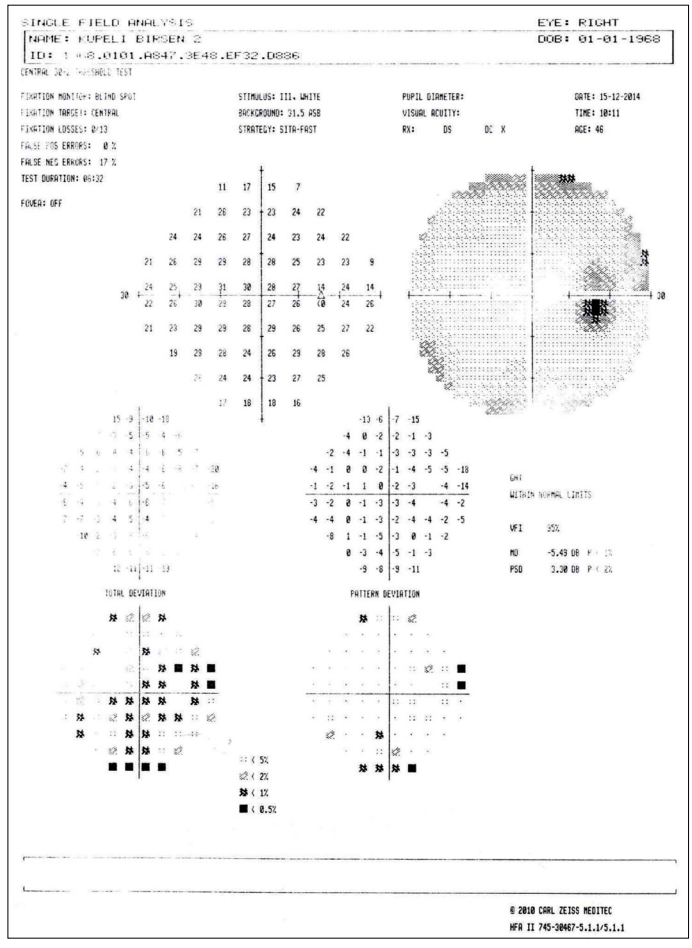

Fig. 3. Visual field test of the right eye before the onset of management.

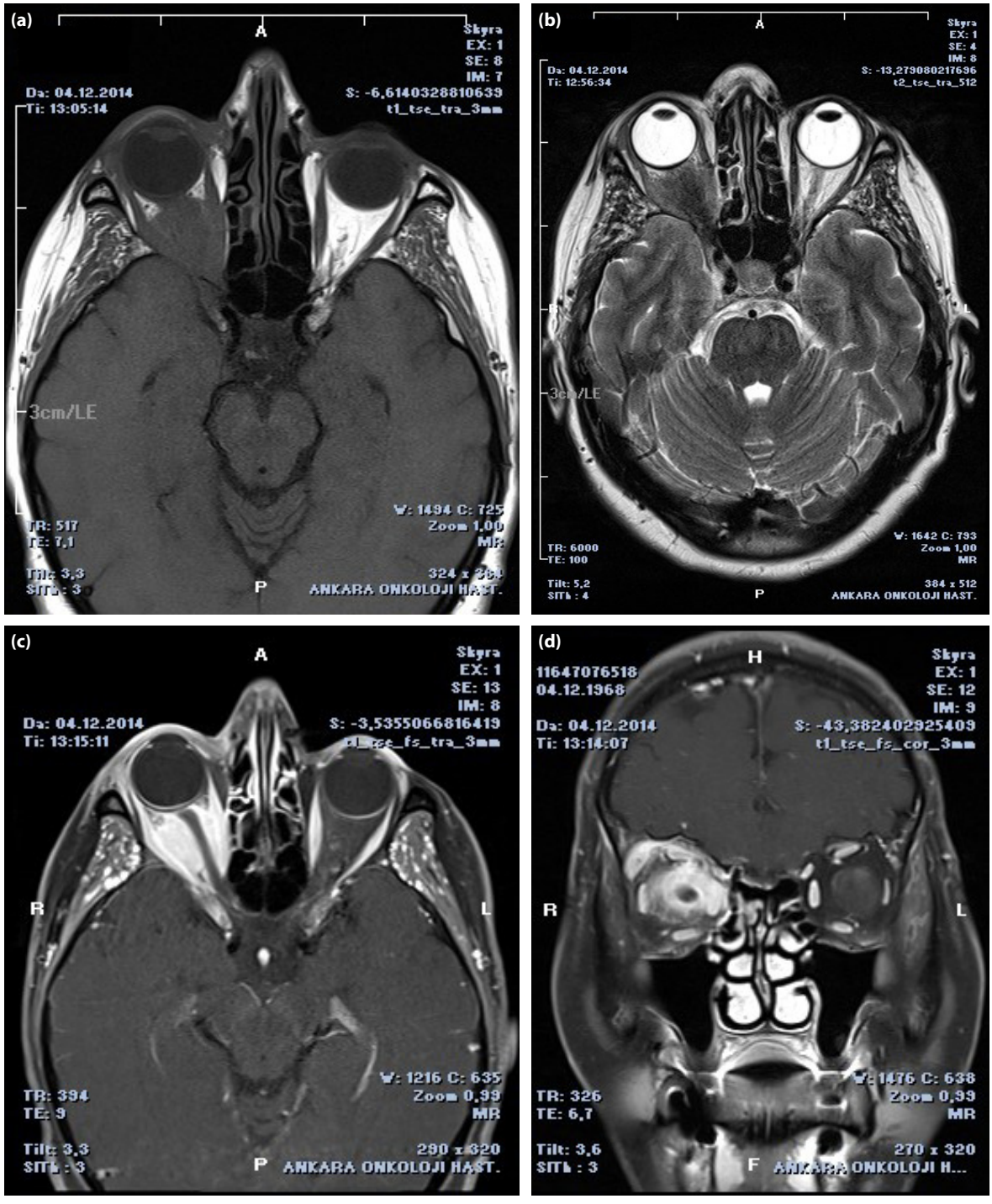

Fig. 4. **(a)** Axial Magnetic resonance imaging (T1 with fat saturation). A lesion with dimensions of 28×26×20 mm, isointense to brain and optic nerve which produces proptosis. **(b)** Axial Magnetic resonance imaging (T2 with fat saturation). The lesion is slightly hyperintense to the optic nerve. **(c)** Axial Magnetic resonance imaging (T1 with fat saturation) after intravenous administration of gadolinium. There is homogenous intense enhancement producing a "tram-track" appearance around the hypointense optic nerve. **(d)** Coronal Magnetic resonance imaging (T1 with fat saturation). "Doughnut sign".

Despite its posterior orbital obliteration, there was no extraorbital/intracranial extension of the lesion. According to characteristic imaging findings, the lesion was diagnosed as an ONSM. Diagnostic biopsy could not be performed because the patient did not accept any surgery. Therefore, the patient was referred to Radiation Oncology Department and treated with three-fraction stereotactic CyberKnife radiation therapy (SRT) with dose of 19.5 Gray (Gy). No acute

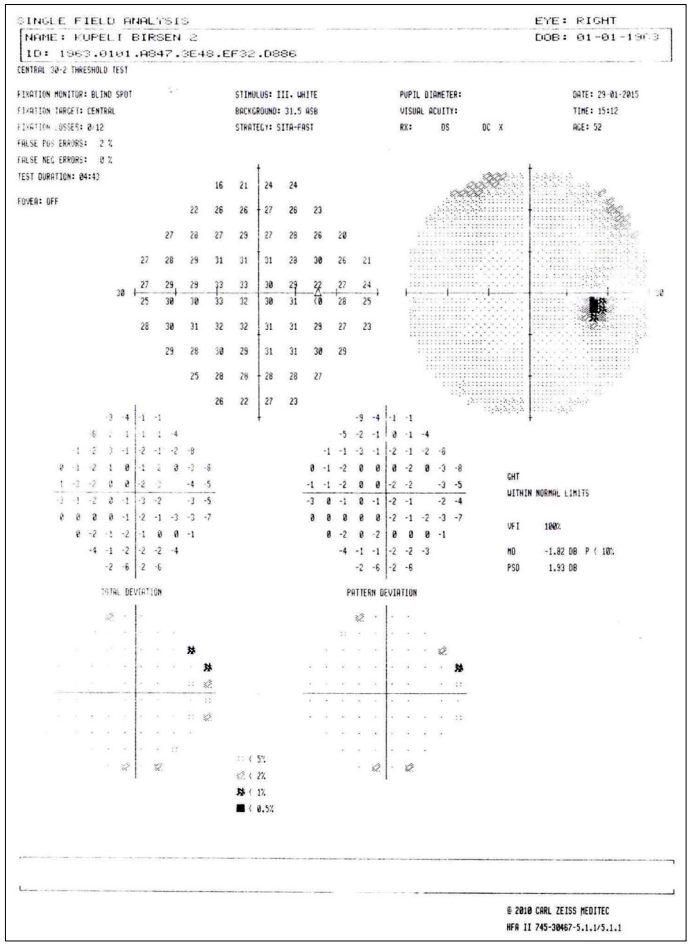

Fig. 5. Recovery of visual field, 6 weeks after radiation therapy.

complications were obtained during the treatment. In follow-up examination performed after 6 weeks, the right ONSM decreased in size from 28×26×20 to 13.8×12.9×11.4 mm, visual acuity was preserved and visual field findings were completely recovered (Fig. 5). After 1 year of treatment puffiness of eyelid, proptosis and optic disc edema were completely recovered and tumor size remained stable. No late side effects of radiation were recorded as well.

Discussion

ONSM is a rare benign tumor of anterior visual pathways, whose delayed diagnosis can lead to progressive visual loss and blindness in advanced cases.[1]

Patients may present in different clinical findings. Common clinical manifestations include ipsilateral slowly painless and progressive visual loss, visual field defect, color vision disturbance, proptosis, optic disc edema, and motility disturbance. Typically, there is relative pupillary defect in the involved eye.

Even patients, who do not have significant visual loss, often have disturbances of color vision and visual field defects.[3] The most common visual field defect is peripheral constriction. Other field defects such as blind spot enlargement, altitudinal field defects, and central scotomas have been also described.[1,3,12] In the presenting case, although there was no loss of vision, blind spot enlargement and mild peripheral constriction were detected in right sided visual field.

Less common symptoms include transient visual obscurations, pain or discomfort, and double vision.[3] The obscurations of vision are almost always associated with optic disc swelling, as observed in our case. Chronic optic disc swelling occurs when the lesion surrounds or compresses the intraorbital part of the optic nerve,[4,5] reminding a presentation of optic neuritis. For this reason, ONSM should be kept in mind, when facing atypical presentation of optic neuritis. These two pathologies indeed may have a similar presentation.

The diagnosis of ONSM relies heavily on imagining findings.[6] It is confirmed with MRI, especially with gadolinium-enhanced fat suppression sequences. ONSM is a sensitive lesion to gadolinium contrast. On MRI axial images, it presents with the characteristic "tram-track" sign, which corresponds to enhancing outer ONSM encircling the inner non-enhancing optic nerve. The patient that we presented was misdiagnosed because of prominent non-specific and external symptoms of the eye and preserved visual acuity. We were able to make diagnosis thanks to complete eye examination which revealed right-sided proptosis, of which reason was confirmed with MRI study.

The goal of treatment for cases of ONSM is tumor control and improvement of vision and optic nerve functions. Conventionally, orbital meningiomas have been observed. Observation remains an acceptable conservative measure in cases, with small and non-progressive tumor and high functional vision, especially in patients who maintain a central visual acuity of 20/50 or better.[11] Tumor resection is almost impossible without incurring a severe visual loss, due to the relationship of the ONSM to the optic nerve. However, surgical resection can be justified in cases of intracranial extension or in cases of disfiguring proptosis, where the visual has significantly decreased. Given its technical difficulties, trend toward post-operative blindness and risk of recurrence after incomplete excision; surgery for ONSM has largely been replaced by radiation therapy. In the past 20 years, radiotherapy has become a first choice therapy[10] and is accepted as the appropriate vision-preserving therapy for the management of ONSM. Whatever the procedure used (IMRT-Intensity-Modulated Radiation Therapy, three-dimensional conformal radiation therapy, stereotactic fractionated radiotherapy-SRT), the functional outcome is better than surgery.

Fractionated radiation therapy is currently the technique most likely to achieve long-term preservation of visual function for most patients.[13]

Metellus et al.[14] have reported that fractionated conformal radiotherapy is safe and effective long-term treatment option. The risk of radiation-induced retinopathy seems to be correlated with total doses higher than 54 Gy, fractionated doses higher than 1.8 Gy or single dose higher than 6 Gy.[14,15] Charpentier et al.[16] have reported usage of 49–51 Gy given in 1.7 up to 2 Gy fraction to avoid radiation side effects.

Our case was treated by three-fraction SRT with dose of 19.5 Gy. As a result of treatment, visual acuity was preserved, and visual field was completely recovered after 6 weeks of follow-up.

Conclusion

ONSM have typical and characteristic radiological findings. The diagnosis is confirmed with MRI, especially with gadolinium-enhanced fat-suppression sequences. The MRI has become the gold standard for the diagnosis and has obviated the need for tissue biopsy. In progressive cases, optic nerve functions may recover with CyberKnife radiation therapy which is performed before development of significant visual loss. However, patients should be observed for radiation complications.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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Authorship Contributions: Concept: S.S.G., H.G.; Design: S.S.G., H.G.; Supervision: S.S.G., H.G.; Resource: S.S.G.; Materials: H.G.; Data Collection and/or Processing: H.G.; Analysis and/or Interpretation: S.S.G.; Literature Search: S.S.G.; Writing: S.S.G.; Critical Reviews: S.S.G.

Conflict of Interest: None declared.

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