

# Optic nerve sheath meningioma-findings in the contralateral optic nerve tract: A case report

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**Abstract.** The aim of the present study was to observe visual pathway changes on the contralateral side in optic nerve sheath meningioma (ONSM). The authors present a case report of a 43-year-old patient with ONSM on the right side. A complex ophthalmic examination was performed, including an assessment of visual functions, an electrophysiology examination and functional and structural MRI examinations. Visual acuity of the right eye after ONSM remained with no light perception, while that of the left side was normal. The visual field of the left eye was normal as was colour perception. An electrophysiology examination using a pattern electroretinogram revealed low amplitude values in the right eye. In the left eye, the finding was at the lower limit of normal results. The pattern visual evoked potential exhibited a bilateral lesion with a larger decrease in response after stimulation of the right eye. The structural MRI revealed intraorbital atrophy of the optic nerve on the right side throughout the whole course, which was accompanied by atrophy of the right half of the optic chiasm. Functional magnetic resonance imaging revealed zero activity after stimulation of the right eye and decreased activity in the visual centre after stimulation of the left eye. The present study demonstrated that unilateral damage to the optic nerve in ONSM is accompanied by significant changes on the contralateral side of the optic pathway.

## Introduction

Experimental unilateral transection of the optic nerve causes acute retinal glial activation on the contralateral side (1-3).

These experimental observations led us to consider whether bilateral glial activation processes are occurring at the level of ganglionic retinal activation in the human visual pathway. In our experience, glial cells are activated when retinal cells are damaged. In the retina, these include microglia, Muller cells, and astrocytes, while in retinal ganglion cells, it appears that the microglia are primarily activated.

Our recent work focused on traumatic optic neuropathy (TON) (4); however, we also studied damage to the contralateral visual pathways. This case report on optic nerve sheath meningioma is presented mainly as confirmation of our previous focus on TON and also to eliminate the possibility of damage to the contralateral pathway after a contusion. Optic nerve sheath meningioma (ONSM) is a rare benign tumour of the central nervous system. Although the growth of these lesions is slow and progressive, their location is critical. Because of their location, ONSM lesions directly influence the frontal visual pathway and can lead to severe vision problems. The patient in this case was a 43-year-old man who had received a transcranial intervention. As far as we are aware, there are no references in the literature regarding damage in contralateral visual pathways caused by ONSM.

## Case report

A 43-year-old man was diagnosed with right-sided ONSM 12 years prior to this report. At the time of diagnosis, there was an attempt to extirpate the tumour via a frontal transcranial approach. The tumour was inaccessible from this approach, and the surgery was ended. The patient was seen at the first author's workplace in April 2018. The patient was found to be completely blind in his right eye. According to the patient, the blindness had been present since the operation. Repeated MRIs showed a tumour of stationary size; therefore, further surgery was not indicated. In addition to standard ophthalmic examinations, we also examined the visual field of the patient using automated perimetry (Medmont M700; Medmont International Pty Ltd.). Retinal nerve fibre layers (RNFL) and the ganglion cell complex (GCC) were examined using spectral domain optical coherence tomography (SD-OCT) with a RTVue-100. The pattern electroretinogram (PERG) and the pattern visual evoked potential (PVEP; Roland Consult

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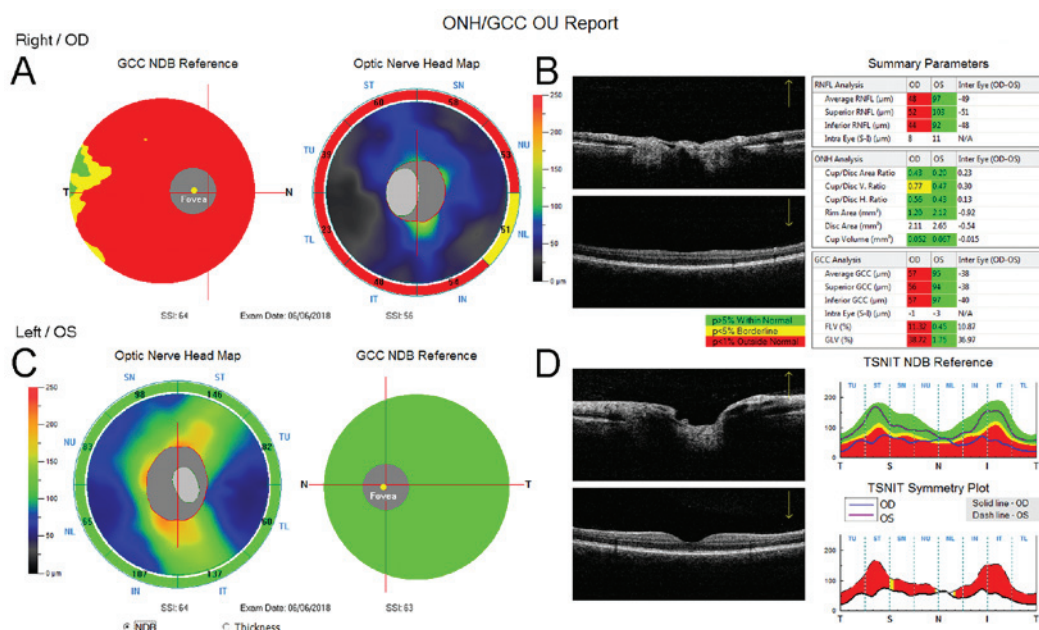


Figure 1. The GCC consists of an inner plexiform layer, a ganglion cell layer and the retinal nerve fiber layer (RNFL). The thickness of the GCC (57  $\mu\text{m}$ ) and RNFL (48  $\mu\text{m}$ ) were significantly reduced in the right eye. (A) ONH map of the right eye. (B) Summary table presenting RNFL, GCC and ONH parameters of the right (OD) and left eye (OS). (C) GCC and ONH map of left eye with normal values. (D) RNFL thickness of the right optic nerve (blue color) and left optic nerve (violet color). RNFL, retinal nerve fiber layer; GCC, the retinal ganglion cell complex; ONH, optic nerve head.

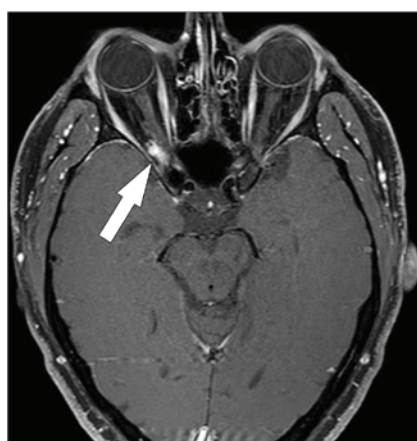


Figure 2. MRI image of the axial plane showing T1 mDIXON with fat signal suppression, as obtained using IV contrast application. The meningeoma residue in the orbital apex can be observed on the right with enhancement (white arrow).

Stasche & Finger GmbH) were obtained while using standard ISCEV methods. The potential for exophthalmos was assessed using a Hertel exophthalmometer, intraocular pressure (IOP) was assessed using an Ocular Response Analyzer, and colored perception was assessed using the Ishihara test for color blindness. The ophthalmologic examination was always performed by the same physician.

Additionally, structural and functional MRI images of the brain were obtained. MRI examinations were carried out using a Philips Achieva 3T TX MR system (Philips Healthcare) with a magnetic field strength of 3 Tesla. The functional MRI (fMRI) used blood oxygen level-dependent (BOLD) contrast. A standard 32-channel head coil was used, and each measurement was performed with a gradient-echo

echo-planar imaging sequence (TR/TE=3,000/30 ms, spatial resolution of  $2 \times 2 \times 2 \text{ mm}^3$ ). Optical stimulation was performed using a black and white checkerboard pattern alternated with its negative image at a frequency of 2 Hz. The size of the black and white checkerboard was  $25.8 \times 16.2$  degrees. Measurements consisted of a sequence of five 30 sec active phase periods and five resting periods of the same length (for each of 10 dynamic scans). During the resting phase, the subject was instructed to maintain view fixation on a static crosshair projected in the centre of the visual field. In total, each measurement included 100 dynamic scans and took 5 min. Each eye was examined in a separate fMRI measurement sequence (LE, RE).

Evaluation of the fMRI data was performed using the SPM 12 software package. Data pre-processing was composed of realignment (motion correction), slice timing (time shift between slice acquisitions), normalization to standard MNI-152 space, and spatial smoothing (FWHM=3x3x3  $\text{mm}^3$ ). A general linear model was used to analyse the data, and the significance threshold was set at  $P=0.05$  with family-wise error (FWE) correction applied to the final t-maps.

Structural magnetic resonance imaging was performed in the standard planes using the sequences as follows: T1-weighted mDIXON, TR of 500 ms, TE of 47 ms, 10 ml Gd-DTPA IV; T2-weighted mDIXON in coronal orientation, slice thickness of 2.5 mm, TR of 3,000 ms, TE of 56 ms; T2-weighted in transverse orientation, slice thickness of 4 mm, TR of 3,000 ms, TE of 56 ms, flip angle of  $57^\circ$ ; FLAIR with slice thickness of 4 mm, TR of 11,000 ms, TE of 125 ms; VenBold, TR of 15 ms, TE of 50 ms, spatial resolution of  $1 \times 1 \times 1 \text{ mm}^3$ ; DWI with slice thickness of 4 mm, TR of 3,443 ms, TE of 76 ms, b-factor 0-800  $\text{s/mm}^2$ , 30 slices.

Structural and functional MRI examinations were carried out in June 2018.

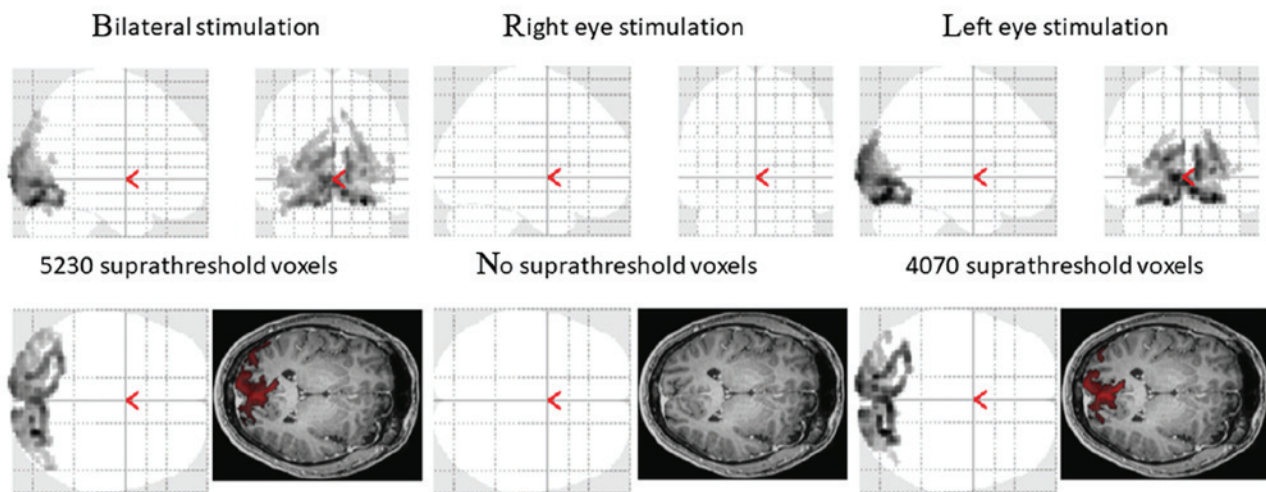


Figure 3. Functional MRI after stimulation of both eyes, right and left eye stimulation.

## Results

Exophthalmos was detected on the right side (up to 2 mm more than the left eye) with 5° of divergence and free mobility. Anisocoria was present, and the relative afferent pupillary defect (RAPD) was positive. Except for this pupilar disorder and simple atrophy of the optic nerve, the ocular findings were normal. On the left side, the findings were normal; the pupil reacted only to a direct light impulse. Vision was with NLP and normal in the right and left eye, respectively. The intraocular pressure (IOP) was 19/16 mm Hg. Colour perception in the left eye remained normal (intact). The visual field on the left side exhibited no decrease insensitivity. The RNFL and GCC were significantly changed on the right side. The average value of RNFL was 48  $\mu$ m on the right and 97  $\mu$ m on the left. The average GCC value on the right was 57  $\mu$ m and 95  $\mu$ m on the left (Fig. 1).

After stimulation of the right eye, the PERG showed decreased amplitude limit values (10  $\mu$ V) and a prolongation of the N95 component to 100 ms. On the left side, the amplitude values were at the standard limit (13  $\mu$ V), and the N95 latency was not prolonged.

The PVEP amplitude values were affected bilaterally. The response of the right side was minimal (0.5  $\mu$ V); the left side response was significantly decreased (6.5  $\mu$ V), and the latency of the P100 component was prolonged up to 118 ms.

The normal value for the PERG amplitudes for P50-N95 components is  $14.80 \pm 2.51$   $\mu$ V; the normal value for PVEP amplitudes for N70-P100 components is  $12.22 \pm 3.22$   $\mu$ V (5).

A structural MRI performed in June 2018 showed a stationary, enhanced meningioma residue in the region of the orbital apex area on the right side without intracranial ingrowth. Intraorbital atrophy of the optic nerve on the right, together with atrophy of the optic chiasma, was visible (Fig. 2). There was no infiltration of the optic nerve chiasma.

A functional magnetic resonance scan was performed two months after the first ophthalmic examination (Fig. 3).

The average healthy population values of responsive values, according to our method, are as follows: When stimulating on the right,  $7,508 \pm 2,018$  voxels; when stimulating on the left,  $7,340 \pm 2,775$  voxels; and after bilateral stimulation,  $7,898 \pm 2,579$  voxels.

With regard to the blindness in the right eye and with regard to the fact that the tumour has not grown, we decided on a conservative approach with monitoring.

## Discussion

Retinal glial activation on both the ipsilateral and the contralateral sides is well known from the literature on experimental models (1-3). These experimental conclusions made us ask whether the processes of bilateral ganglionic activation are also occurring at the level of ganglionic retinal activation in the human visual pathway. In our recent work, which focused on TON (4), we showed that with unilateral damage to the optic nerve, there are significant changes in the contralateral eye pathways, including functional retinal changes.

In this case, our patient had a normal visual field and colour perception on the contralateral side and did not show any abnormalities relative to the ONSM. This might be because there must be a loss of 25-30% of the ganglionic retinal cells to register any perimetric changes when examining a patient using static automatic perimetry, and the same is with respect to colour perception.

RNFL and GCC examinations showed a loss on the ipsilateral side only. Although these changes can occur on the contralateral side, they cannot be detected by any method currently available. Optic coherence tomography only measures the GCC thickness, which can be affected by glial proliferation. GCC is altered only when structural changes are present. The functional outcome of the PERG and PVEP that was pathological on the contralateral eye is essential.

ONSM is a lesion that is sensitive to gadolinium contrast. On MRI axial images, it presents with a characteristic 'tram-track' sign, which corresponds to enhancement of the outer ONSM encircling the inner non-enhanced optic nerve. On coronal images, this pattern will have the appearance of a 'doughnut.'

One issue related to contralateral eye nerve damage or, rather, damage to the contralateral eye pathway after ONSM, is not mentioned in the human medical literature. Using PERG in a mouse model, Liu *et al* (6) found changes both in the ipsilateral and in the contralateral eye in situations with unilateral



damage to the optic nerve; these findings preceded morphologic changes in the retinal nerve fibre layer. The PERG results in our patient support this finding.

We are not aware of any available PVEP or fMRI findings related to ONSM in published experimental or human studies. Our results show changes to the entire visual pathway not only on the damaged side but also on the contralateral side.

ONSM is not a malignant tumour; nevertheless, the consequences of ONSM can lead to blindness. A patient with ONSM usually visits an ophthalmologist due to a temporary unilateral fogging of eyesight, changes in the visual field, and worsening vision. These visual problems, on closer inspection, are often accompanied by edema of the optic nerve. A slight proptosis of the affected bulb can also be accompanied by a loss in ocular mobility. An afferent pupillary defect is also obvious. The gold standard for diagnosis is magnetic resonance imaging using gadolinium as a contrast substance. Sequences with fat signal reduction are preferable. An MRI examination can show the extent of damage, the degree of optic nerve channel infiltration, and possible extension into the optic chiasma and other intracranial structures. In terms of therapy and management of ONSM, the recommended methods are similar. Monitoring remains a suitable conservative treatment when visual functions remain stable, especially in patients whose central visual acuity is 0.4 or better. Neuro-ophthalmologists should carefully monitor patients and perform a thorough neuro-ophthalmic examination, including regular monitoring of the visual field and the peri-papillary RNFL. Repeated MRI examinations are also justified (7-9).

In cases where visual function deteriorates, fractionated stereotactic radiotherapy has become the preferred treatment since it provides sufficient radiation of the tumour in a very localized way. This method can preserve visual functions in most patients, although risks from radiation retinopathy or optic neuropathy certainly exist (7-9).

Tumour resection is almost impossible without significant loss of vision due to the close proximity of the ONSM to the optic nerve. Nevertheless, surgical resection can be justified in cases of increased proptosis that has significantly reduced visual functions or in cases of intracranial enlargement (7,8).

Our previous work, together with this case study, shows that unilateral damage to the optic nerve associated with ONSM can lead to significant changes in the contralateral visual pathways, including changes in retinal function. Clinical trials with companion ophthalmologic correlative studies of optic nerve pathways and neurotransmissions are required to help inform about mechanism of both side visual pathway affection after one side optic nerve damage.

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## Availability of data and materials

The datasets used and/or analyzed during the present study are available from the corresponding author on reasonable request.

## Authors' contributions

All the authors were involved in conceiving and designing the present study. JL drafted and wrote the manuscript. MK, JL and JT were responsible for collecting and analyzing patient data. PH drafted the manuscript and revised it critically for important intellectual content. All authors have read and approved the final version of the manuscript.

## Ethics approval and consent to participate

Written informed consent was obtained from the patient prior to enrolment.

## Patient consent for publication

The patient gave written consent; however, the authors also made efforts to remove any identifying information to protect the privacy of the patient.

## Competing interests

The authors declare that they have no competing interests.

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