CASE REPORT

Total recovery of optic nerve sheath meningioma

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SUMMARY

We reported the case of a 43-year-old woman with a rapid progressive visual loss. The diagnosis of primary optic nerve sheath meningioma was made thanks to atypia evolution, clinical and radiological findings. The patient was treated with an intensity-modulated radiotherapy of 50.4 Gy in 28 fractions 2 months after the diagnosis. Visual acuity and visual field were completely recovered after 2 years of follow-up. No late side effects of irradiation were recorded.

BACKGROUND

Primary optic nerve sheath meningioma (ONSM) is a rare benign tumour of anterior visual pathways whose delayed diagnosis can lead to progressive visual loss and blindness in advanced cases.^{1 2} There is no consensus of management of such slow-growing tumour. Visual outcome indeed seemed better in radiotherapy management than neurosurgery with less late side effects.^{3–5} Thus, radiotherapy is now becoming the first-choice procedure in this indication.^{3 6} However, the perfect time of management remains unclear, and some practitioners tend to observe small tumours without life-threatening evolution, and with a preserved functional visual acuity.³ We report here the case of a woman aged 43 years with ONSM who was treated by radiotherapy 2 months after the diagnosis.

CASE PRESENTATION

A 43-year-old woman was referred to our university hospital for the management of a progressive visual loss. Her ophthalmologist recently diagnosed a unilateral low vision of the right eye (20/200), with afferent pupillary defect and normal fundus examination discovered by routine examination. The medical history included obesity (BMI=32), blood hypertension and depression status under therapy. Contraceptive medication (cyproterone acetate) and oestradiol were taken regularly. No personal and family medical history of migraine or optic inflammation neuropathy was recorded. The

left eye was normal. Initial clinical assessment was in favour of an optic neuropathy.

INVESTIGATIONS AND DIFFERENTIAL DIAGNOSIS

A diffuse scotoma with foveal threshold lower than 10 dB was observed on the visual field ([figure 1](#)). Prolongation of latency and decrease in amplitude were recorded on visual evoked potential. Retinal nerve fibre layer optical coherence tomography (RNFL OCT) analyses were normal ([figure 2](#)). A hypersignal in T2 sequences and an iso-signal in T1-weighted lesion were detected on the right optic nerve using MRI. The lesion enhanced after gadolinium injection without lesion of the white matter. The diagnosis of idiopathic retrobulbar optic neuritis was established because no other causes were found by biological and radiological investigations. Thus, the patient received steroids according to the Optic Neuritis Treatment Trial. Since visual acuity and pain did not evolve despite systemic steroid therapy after 1 month, the diagnosis was re-evaluated with radiologists using additional MRI sequences. The final diagnosis was a cone-shaped ONSM of the right eye in the pre-chiasmatic portion measuring 10×4×6 mm ([figures 3–5](#)).

TREATMENT

RNFL OCT analyses showed atrophy in temporal nerve fibre layers ranging from 65 to 44 µm during the first months of follow-up ([figure 6](#)). Thus, we decided to treat the lesion by radiation therapy. An intensity-modulated radiotherapy (IMRT) of 50.4 Gy in 28 fractions was performed over 6 weeks (28 daily fraction of 1.8 Gy).

OUTCOME AND FOLLOW-UP

Two years later, the visual recovery was unexpected. The best-corrected visual acuity has increased from 20/200 to 20/20, and foveal threshold increased to 34 dB with total vision field recovery. No late side effects were recorded. RNFL OCT analyses remain stable ([figures 7 and 8](#)). The right

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Test de seuil central 30-2

Contrôle de fixation: Suivi regard/T.A.

Stimulus: III, Blanc

Diamètre de la pupille:

Date: 06-09-2013

Cible de fixation: Central

Fond: 31.5 ASB

Acuité visuelle:

Heure: 14:13

Pertes de fixation: 2/17

Stratégie: SITA-Standard

RX: -2.00 DS DC X

L'âge: 40

Erreurs faux pos.: 2 %

Erreurs faux nég.: Sans objet

Durée du test: 08:34

Fovéa: 10 dB ■

Déviation individuelle
non indiquée pour les
champs en chute
considérable. Voir
Déviation totale.

THG
Hors limites normales

VFI 7%

MD -29.86 dB P < 0.5%

PSD 7.27 dB P < 0.5%

Déviation Totale

Déviation individuelle

Déviation individuelle
non indiquée pour les
champs en chute
considérable. Voir
Déviation totale.

:: < 5%

⊗ < 2%

⊠ < 1%

■ < 0.5%

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Figure 1 Visual field at the beginning of management.

ID: CZM127240464 Date d'examen: 01/03/2013 01/03/2013 CZM
 Date de naissance: 10/04/1973 Heure de l'examen: 11:33 11:31
 Sexe: Féminin Numéro de série: 4000-4742 4000-4742
 Technicien : Cirrus Operateur, Puissance du signal: 7/10 8/10

RNFL et ONH : Optic Disc Cube 200x200

OD ● OS

Figure 2 RNFL OCT analysis at the beginning of management.

Figure 3 MRI of the lesion of the right optic nerve: hyper-signal on T2 weighted images.

Figure 4 MRI of the lesion enhanced by gadolinium on T1 weighted images.

ONSM decreased in size from 10×4×6 to 5×3.5×7.5 mm and remained stable during follow-up (figures 9 and 10).

Figure 5 MRI of the lesion: T2 weighted images in axial view.

DISCUSSION

ONSMs constitute ~2% of all orbital tumours and 1–2% of all meningiomas.⁷ Typical symptoms in orbital meningiomas are visual acuity loss followed by visual field defect, diplopia, proptosis, and optic disc swelling or atrophy.^{1 2} In practice, these classical signs are rarely seen together and reveal advanced disease.

In our case, we had a small lesion of 10×4×6 mm without external signs, and initial RNFL OCT analyses showed a normal optic disc with a visual acuity of 20/200. During the follow-up, the rapid-onset deterioration in RNFL OCT analysis with unchanged visual functions had led us to initiate therapy. In the last 20 years, radiotherapy has become a first-choice therapy for managing ONSM.⁶ Whatever the procedure used (three-dimensional conformal radiation therapy, IMRT, stereotactic fractionated radiotherapy), the functional outcome is better than surgery. We often use 49–51 Gy given in 1.7 up to 2 Gy fraction to avoid radiation side effects.^{6 8–11} Long-term safety data on this intervention are lacking. Metellus *et al*¹² have demonstrated that fractionated conformal radiotherapy is a safe and effective long-term treatment option. The risk of radiation-induced retinopathy seems to be correlated with the total doses higher than 54 Gy, fractionated doses higher than 1.8 Gy or single dose higher than 6 Gy.^{12 13} ONSMs involving the posterior portion of the globe are at higher risk of developing radiation-induced retinopathy. Thus, an early therapy could be the key to favourable outcomes. In our case, visual acuity and visual field were completely recovered after 2 years of follow-up. No late side effects of irradiation were recorded. The prognostic value of RNFL OCT analysis should be considered for management of ONSM.^{14 15}

ID: CZM1127240464 Date d'examen: 12/12/2013 12/12/2013 CZM
 Date de naissance: 10/04/1973 Heure de l'examen: 11:33 11:31
 Sexe: Féminin Numéro de série: 4000-4742 4000-4742
 Technicien : Cirrus Operateur, Puissance du signal: 7/10 8/10

RNFL et ONH :Optic Disc Cube 200x200

OD OS

Figure 6 Worsening of RNFL OCT analysis during follow-up. RNFL OCT, retinal nerve fibre layer optical coherence tomography.

Test de seuil central 30-2

Contrôle de fixation: Suivi regard/T.A.

Stimulus: III, Blanc

Diamètre de la pupille:

Date: 30-03-2015

Cible de fixation: Central

Fond: 31.5 ASB

Acuité visuelle:

Heure: 14:56

Pertes de fixation: 0/18

Stratégie: SITA-Standard

RX: -2.00 DS DC X

L'âge: 41

Erreurs faux pos.: 0 %

Erreurs faux nég.: 0 %

Durée du test: 06:26

Fovéa: 34 dB

Figure 7 Visual field 1 year after radiation therapy.

ID: CZM127240464 Date d'examen: 02/10/2015 02/10/2015 CZM
 Date de naissance: 10/04/1973 Heure de l'examen: 14:28 14:29
 Sexe: Féminin Numéro de série: 4000-4742 4000-4742
 Technicien: Cirrus Operateur, Puissance du signal: 6/10 7/10

RNFL et ONH :Optic Disc Cube 200x200

OD ● ● OS

Figure 8 RNFL OCT analysis 1 year after radiation therapy.

Figure 9 MRI more than one year after radiotherapy. T2 spin weighted images.

Figure 10 MRI more than one year after radiotherapy: T1 fat sat weighted images.

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Competing interests None declared.

Patient consent Obtained.

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Learning points

- ▶ Optic nerve sheath meningioma (ONSM) should be kept in mind when facing atypical presentations of optic neuritis. These two pathologies indeed may have a similar presentation.
- ▶ Radiation therapy is now becoming the first choice for managing primary ONSM.
- ▶ The prognostic value of retinal nerve fibre layer optical coherence tomography analysis may help during follow-up and for initiating therapy of primary ONSM.

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