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Original Article

Fractionated Stereotactic Conformal Radiotherapy for Optic Nerve Sheath Meningiomas

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Abstract

Aims: To assess visual outcome, tumour control and treatment-related morbidity in patients with optic nerve sheath meningiomas (ONSMs) treated with fractionated stereotactic radiotherapy (FSRT).

Patients and methods: A retrospective analysis of 45 patients (13 men and 32 women, median age 46 years) with ONSMs (51 optic nerves involved) treated in a single institution between 1997 and 2010 was carried out. FSRT was delivered to a dose of 50 Gy in 30 or 33 fractions as primary treatment in 39 patients and after surgery in six patients.

Results: At a median follow-up of 30 months (range 1–13 years), the tumour control in 41 evaluable patients (four were lost to follow-up) was 100% at 5 years with no subsequent local or distant recurrence. Of the 46 evaluable optic nerves treated, 41 had residual vision (38 with impaired vision) before radiotherapy and five were blind in one eye. There was no recovery of vision in any of the blind eyes. Of 41 optic nerves with residual vision, 13 had improvement, 24 remained stable and four deteriorated; two patients (4%) developed radiation retinopathy. One patient developed a central retinal artery occlusion in the untreated eye 10 years after treatment.

Conclusion: FSRT is highly effective at controlling the growth of ONSMs with improvement or stabilisation of visual deficit in 89% of the optic nerves retaining some vision, albeit with a small risk of radiation-induced retinopathy. The results support the use of FSRT as an effective approach in the management of ONSM. The lack of functional benefit in patients with severe visual impairment would argue for earlier institution of treatment before complete visual loss is established.

Key words: Meningioma; optic nerve; stereotactic radiotherapy

Introduction

Optic nerve sheath meningioma (ONSM) is a rare tumour accounting for 1–2% of all intracranial meningiomas and the second most common tumour affecting the optic nerve after optic gliomas. As with other intracranial meningiomas, the most commonly affected group are women in middle age, with a peak of incidence at 41 years (range 2.5–78 years) [1].

ONSMs are mostly unilateral tumours, with about 5% involving both optic nerves, particularly seen in young patients with neurofibromatosis type 2 [2]. They wrap around the optic nerve through the subdural and subarachnoid spaces, along all paths of least resistance, such as vessels and dural septa [1,3]. Circumferential constriction of the optic nerve may impair the vascular supply and interfere with the axonal transport [4].

The presenting features include progressive loss of visual acuity and visual fields, proptosis, optic disc oedema, restricted eye movements, pain and lower eyelid oedema [5,6]. The combination of clinical features with magnetic resonance imaging (MRI) appearance of a thickened optic nerve on fat-suppressed T1-weighted sequences is diagnostic of ONSM, obviating the need for obtaining a tissue sample [7]. Although described in a small proportion of patients, the clinical triad of visual loss, optic atrophy and optociliary shunt veins is considered pathognomonic of ONSM. Despite indolent growth, untreated ONSMs cause progressive visual deterioration leading to blindness.

ONSMs are managed with a combination of surveillance and irradiation with occasional recourse to surgical

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intervention. Surveillance is considered the appropriate approach in patients with stable vision without detectable tumour growth on imaging. Surgery is rarely carried out due to the risk to vision. Debulking surgery is of value as a cosmetic procedure in patients with disfiguring proptosis and biopsy may be necessary in cases of diagnostic difficulty [4,8,9].

Fractionated radiotherapy has been widely adopted in the treatment of ONSMs, with the aim of controlling tumour growth and preserving or improving visual function [8–39]. Fractionated stereotactic radiotherapy (FSRT) as a high precision refinement of conformal radiotherapy provides a more localised delivery of radiation, minimising the volume and the dose of radiation to the uninvolved optic apparatus and the adjacent structures, with the hope of reducing the risk of treatment-related side effects.

We report a single institution experience of 45 patients treated with FSRT for ONSMs, assessing medium-term tumour control, visual outcome and treatment-related morbidity.

Materials and Methods

Patients

Between January 1997 and December 2010, 45 patients (51 optic nerves) with ONSM were treated with FSRT at the Royal Marsden Hospital. Indications for treatment included visual loss at presentation, progressive visual deterioration on surveillance, imaging evidence of tumour growth and a combination of these.

Fractionated Stereotactic Radiotherapy Technique and Dose Prescription

Technical details have been reported previously [40–43]. Briefly, patients were immobilised in a Gill–Thomas–Cosman frame. High-resolution planning computed tomography (2 or 3 mm slice thickness) was used to outline the gross tumour volume (GTV) in five patients treated before 2004. In all other patients, the computed tomography (CT) scan was fused with a planning MRI scan. The GTV was defined as the lesion on CT and fat-suppressed T1-weighted gadolinium-enhanced MRI scans. The three-dimensional volume growing algorithm was used to expand the GTV by 3 mm (initially 5 mm) to generate a planning target volume (PTV). Critical structures including the eyes, optic nerves and optic chiasm were also outlined. No device to reduce eye movement was used.

The PTV was treated with four non-coplanar conformal fixed fields (one patient with three fields) based on the class solution reported previously [44]. Initially, beam shaping was achieved with customised lead blocks (four patients) and subsequently with a 120 multileaf collimator (41 patients). Forty-two patients received 50 Gy in 30 fractions and three patients received 50 Gy in 33 fractions using 6 MV linear accelerator. Doses were prescribed at the isocentre according to ICRU 50 criteria with the PTV covered by the 95% isodose in three dimensions (Figure 1).

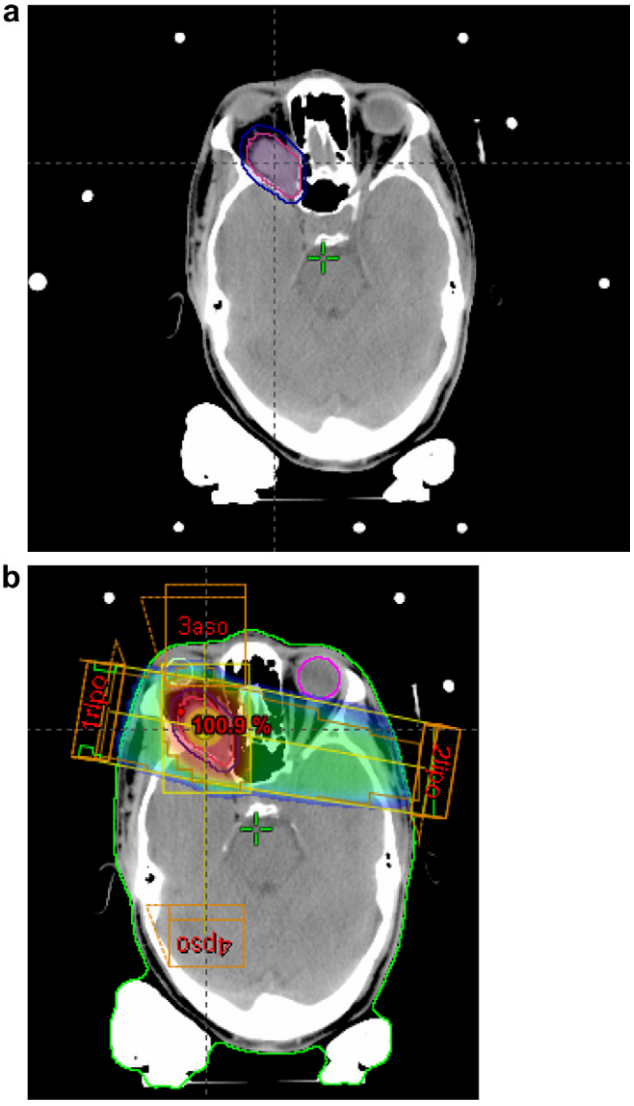

Fig 1. (a) The gross tumour volume (GTV) (purple line) and the planning target volume (PTV) (blue line) outline and (b) isodose distribution of a four-field non-coplanar beam arrangement.

Clinical Assessment and Follow-up

Patients were reviewed weekly during radiotherapy for the assessment of acute toxicity and then 1, 3 and 12 months after the completion of treatment with clinical assessment of neurological status. The first ophthalmological follow-up examination with visual acuity and visual field testing was carried out 3 months after treatment. A baseline MRI scan was carried out at 3 months after the end of radiotherapy. Subsequently, patients were reviewed annually (or more often as clinically indicated) with a repeat MRI scan to assess local control. Formal ophthalmological assessment at the time of diagnosis was not available for all patients, although most presented with impairment in visual acuity. Visual outcome after treatment was defined as improved, deteriorated or stable on the basis of the ophthalmologist's report and of the patient's subjective assessment.

Pituitary function was assessed at annual intervals in an endocrine clinic. Toxicities were documented according to