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Original Article

Radiotherapy for Optic Nerve Sheath Meningioma: A Case for Earlier Intervention?

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Abstract

Aims: To assess tumour control, visual outcomes and toxicity after radiotherapy for all patients with optic nerve sheath meningiomas (ONSM) treated by a single radiation oncologist at a single institution over a 15 year period. To explore potential predictors of outcomes.
Materials and methods: All patients underwent ophthalmological and radiological assessments before radiotherapy. These were repeated at regular intervals after treatment. A retrospective analysis of clinical, dosimetric and radiological data was carried out. Patients with useful vision before radiotherapy were divided into two groups – those with maintained or improved vision and those with a deterioration in vision. The groups were compared using the Mann–Whitney *U*-test with regard to eight potential predictors of outcome.
Results: Seventeen patients with 18 ONSM were treated with fractionated radiotherapy (46.8–55.8 Gy in 26–31 fractions). No evaluable tumours grew after treatment: control rate 100% (95% confidence interval 82–100%). Using the most common definition of visual function described in the literature, vision was maintained or improved in 89% (95% confidence interval 67–97%) of cases. In those with useful vision before treatment (13 evaluable eyes), visual acuity was maintained or improved in eight (62%, 95% confidence interval 36–82%). There was a suggestion that the time from the onset of symptoms to radiotherapy may influence outcome. Those with stable or better visual acuity after radiotherapy had been observed for a shorter time compared with those who had worse visual acuity (median of 18 months versus 62 months). Acute and late toxicity from radiotherapy was manageable.
Conclusion: Radiotherapy is an extremely effective modality in arresting the growth of ONSM. A longer time from symptom onset to the start of radiotherapy may predict for poorer outcomes.

Key words: Meningioma; ONSM; optic nerve; optic nerve sheath; radiotherapy

Introduction

Optic nerve sheath meningiomas (ONSM) are rare and account for 2% of all orbital tumours [1]. Like other meningiomas, they are more common in women and are typically slow-growing benign tumours. The usual presentation is with progressive visual loss or proptosis (Figure 1). Historically it was felt that complete visual loss was inevitable and that the only worthwhile intervention was to surgically decompress the nerve in those with useful but deteriorating vision, with the aim of delaying complete visual loss [2]. However, over the last few decades, evidence has emerged that with radiotherapy, not only can the growth of the tumour be stopped or reversed, but also vision can be maintained or improved in most cases [3]. Because of the rarity of ONSM, evidence has come from case series with limited numbers [4–13] that have either combined the experience from several groups [4,5] or have presented single institutional data [6–13]. Various techniques have been used, including conventional three-dimensional conformal radiotherapy (3DCRT) [4,10,12], fractionated stereotactic radiotherapy (FSRT) [4–9,11,13], stereotactic radiosurgery [11] or proton therapy [6]. It is not possible from these small studies to determine if one

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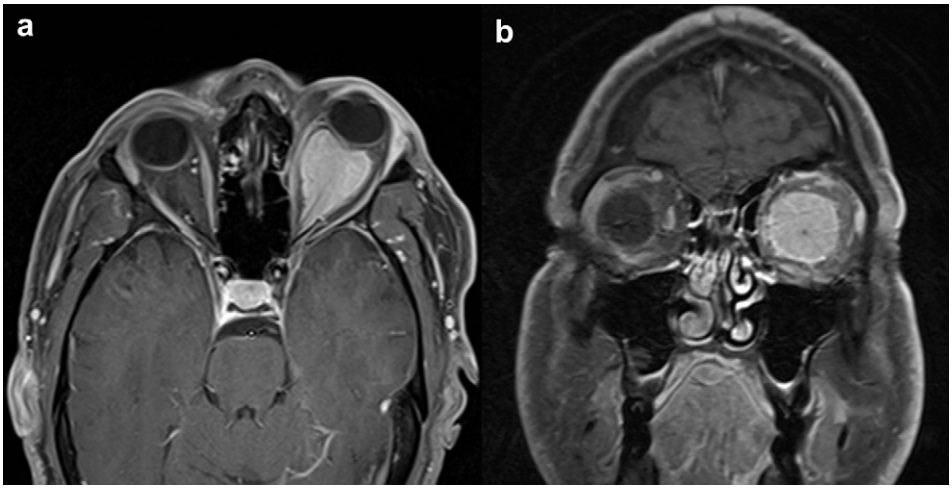

Fig 1. T1-weighted post-contrast magnetic resonance imaging of a neglected 35 × 27 × 26 mm left optic nerve sheath tumour causing blindness in a patient initially observed, then lost to follow-up before seeking treatment for cosmetic reasons. (a) Axial image showing severe left proptosis and 'ghost' of the atrophic optic nerve within the enhancing tumour posteriorly; (b) coronal image through the anterior aspect of the tumour showing the compressed left optic nerve (central dark spot), and the posterior aspect of the normal right globe (dark).

modality is superior to others. However, given that radiation tolerance of the optic nerve and chiasm may be improved by using conventionally fractionated (1.8–2 Gy per fraction) radiotherapy, and the limited toxicity data on hypofractionated radiotherapy on these structures [14], it is preferable to use conventionally fractionated radiotherapy when attempting to maintain vision.

The timing of when radiotherapy should be administered is less certain. Due to concern regarding late effects on the optic apparatus, a period of observation is often advocated while there is no clinical or radiological deterioration [3,4,15]. Yet, the question arises as to whether prolonged observation may compromise preservation of vision as the tumour continues to grow-albeit slowly.

The aim of this study was to assess tumour control, visual outcomes and treatment-related toxicity for all patients with ONSM treated with radiotherapy at the Royal Adelaide Hospital, South Australia, and to compare our experience with previously published series. In addition, we wanted to explore the influence on these outcomes of factors such as the prescribed dose of radiotherapy, the dose to critical structures, the tumour size or the timing of radiotherapy.

Materials and Methods

All patients who received radiotherapy for ONSM between 1996 and 2011 were identified using the departmental electronic database. The diagnosis was based on the presence of typical clinical features and characteristic magnetic resonance imaging (MRI) findings [3,15]. Where there was sufficient doubt about the diagnosis, a biopsy was carried out.

Relevant information was obtained from hospital records, radiotherapy treatment plans and serial imaging (baseline and follow-up MRI scans). For patients referred from elsewhere, their ophthalmologist was contacted directly for further information when required.

Ophthalmological Signs/Symptoms

All patients had a complete assessment at first presentation to an ophthalmologist. This included best corrected visual acuity (Snellen chart), colour vision, visual fields, eye movements, optic discs, proptosis, relative afferent pupillary defect and pain. At the time of referral for radiotherapy, any changes in these findings from baseline were recorded.

After completing treatment, patients were followed long term by their referring ophthalmologist with regular repeat assessments and (usually) yearly imaging. Cataract formation as a cause of declining visual acuity was identified and corrected where appropriate.

Radiotherapy

In all but one patient, radiotherapy was delivered using conventional conformal techniques using 6 MV photons with immobilisation in a thermoplastic shell. On the planning computed tomography scan, the gross tumour volume was defined as the visible (enhancing) meningioma and was marked with the aid of the diagnostic MRI (fusion images when available). A margin of 5–10 mm was added to create the planning target volume. Organs at risk, namely retina, optic nerve, chiasm and brain stem, were delineated. An optimal conformal plan was obtained with regard to the prescribed dose and tolerances of organs at risk. A typical beam arrangement involved a three- to five-field plan sometimes including a superior–inferior oblique field exiting via the oral cavity to help spare dose to anterior orbital structures. Image guidance protocols followed departmental protocols at the time.

One patient was treated using FSRT, using three non-coplanar 6 MV photon arcs with circular collimators to a total dose of 52.2 Gy in 29 fractions prescribed to the 90% isodose curve (final fraction off retina). The gross