Journal of Clinical Neuroscience 20 (2013) 1045–1056

Contents lists available at SciVerse ScienceDirect

Journal of Clinical Neuroscience

journal homepage: www.elsevier.com/locate/jocn

Review

Diagnosis and management of optic nerve sheath meningiomas

J. Shapey [a,*], H.I. Sabin [b,c], H.V. Danesh-Meyer [d,e], A.H. Kaye [e,f]

[a] Department of Neurosurgery, The Royal London Hospital, Whitechapel, London E1 1BB, UK
[b] The London Gamma Knife Centre, St Bartholomew's Hospital, London, UK
[c] Department of Neurosurgery, The National Hospital for Neurology and Neurosurgery, Queen Square, London, UK
[d] Department of Ophthalmology, University of Auckland, Auckland, New Zealand
[e] Department of Surgery, The University of Melbourne, Melbourne, VIC, Australia
[f] Department of Neurosurgery, The Royal Melbourne Hospital, Melbourne, VIC, Australia

ARTICLE INFO

Article history:
Received 20 February 2013
Accepted 9 March 2013

Keywords:
Brain tumour
Optic nerve sheath meningioma
Proton beam therapy
Radiosurgery
Radiotherapy
Surgery

ABSTRACT

Optic nerve sheath meningiomas account for a third of all intrinsic tumours of the optic nerve. Despite their classification as histologically benign tumours they cause progressive visual loss that often leads to blindness if left untreated. Recent therapeutic advances have increased the treatment options available to clinicians but patient management remains controversial. We systematically review the progress made in the diagnosis and management of optic nerve sheath meningiomas, clarify current best practice, and suggest future avenues for research.

1. Introduction

Optic nerve sheath meningiomas (ONSM) are rare tumours. Nevertheless, they are the second most common optic nerve tumour after optic nerve gliomas[1] and represent a third of all intrinsic tumours of the optic nerve.[2] Despite their classification as histologically benign tumours they cause progressive visual loss that often leads to blindness if left untreated. Recent therapeutic advances have increased the treatment options available to clinicians, however the management of these patients remains controversial. We review the progress made in the diagnosis and management of ONSM, clarify current best practice, and suggest future avenues for research.

2. Illustrative patients

2.1. Patient 1

A 69-year-old woman presented with slowly progressive right-sided unilateral visual loss over 10 years. Within 6 years her vision had deteriorated to no perception of light, at which time she presented to her local ophthalmologist. MRI demonstrated a discrete unilateral lesion confined to the optic nerve with no intracranial extension, consistent with an ONSM. She was managed conservatively with regular clinical and radiological surveillance. Apart from well-controlled hypertension she had no other past medical history of note.

After 5 years of follow-up she developed worsening proptosis in her right eye. Vision in her left eye remained normal and she had no other associated symptoms. She had no perception to light on the right side but visual acuity on the left side was 6/6 with a normal Humphrey visual field test. She had restricted upgaze on the right but all other extra-ocular movements were normal and ocular motility on the left side was not affected. Fundoscopy of the right eye demonstrated marked optic disc atrophy but the left fundus was entirely normal. Subsequent MRI demonstrated that her tumour had increased in size with an associated intracranial component but did not involve the chiasm (Fig. 1).

In view of the progression she underwent Gamma Knife (GK; Elekta Instruments, AB, Stockholm, Sweden) treatment of 15 Gy with a 50% isodose covering 97% of a 4.3 cubic centimetre (cc) lesion (Fig. 2). Her initial postoperative scan 6 months following treatment demonstrated a slight reduction in tumour volume and the extent of intracranial extension, and subsequent MRI continues to show stable radiographic appearances 6 years after treatment (Fig. 3).

2.2. Patient 2

A 62-year-old man presented with a 5 year history of progressive visual failure in his right eye. MRI confirmed an intraorbital

* Corresponding author. Tel.: +44 78 1577 0723.
E-mail address: jshapey@doctors.org.uk (J. Shapey).

0967-5868/$ - see front matter © 2013 Elsevier Ltd. All rights reserved.
http://dx.doi.org/10.1016/j.jocn.2013.03.008

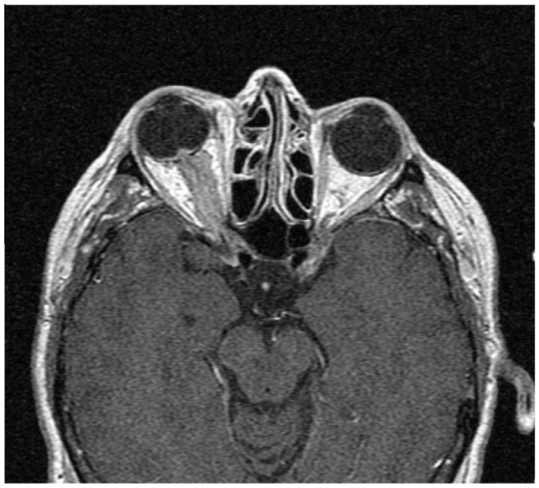

Fig. 1. Axial T1-weighted MRI showing a right-sided optic nerve sheath menignioma.

ONSM, but following further visual deterioration he underwent a course of focused radiation therapy, 3 years after his initial presentation (50.4 Gy in 28 treatments prescribed to the 90% isodose). He subsequently had further loss of visual field, and MRI demonstrated intracranial extension of the ONSM involving the chiasm (Fig. 4). Visual acuity had reduced markedly, being 6/36 in the affected eye with only a central field remaining and colour vision being 1:14 on the Ishihara plates. In view of the progressive growth with intracranial extension a craniotomy was performed with excision of the intracranial tumour. At surgery the tumour was noted to be engulfing the right optic nerve and extending to the optic chiasm. The intracranial tumour was resected. Postoperatively there was a deterioration in the patient's vision to perception of light only in the right eye, which transiently improved following steroid administration but subsequently deteriorated to continuing perception of light only. The vision in the left eye was unaffected, with normal visual field.

3. Epidemiology

ONSM account for approximately 2% of all orbital tumours and represent around 1–2% of all meningiomas.[2–5] Most meningiomas that involve the orbit are secondary tumours and represent extensions from intracranial sites whereas true primary ONSM are far less common and may arise from the intraorbital or portion of the optic nerve sheath.[6] In a review of 5000 orbital meningiomas, Dutton reported that 90% were secondary tumours resulting from intracranial extension and 10% were primary orbital tumours, 96% of which originated from the optic nerve sheath and 4% from other ectopic locations within the orbit. Of the primary ONSM reported, 92% had an origin within the intraorbital nerve sheath whilst only 8% were intracanalicular in origin. Most ONSM are unilateral (95%) however 65% of bilateral lesions are intracanalicular.[2] In Dutton's review, about half of the bilateral tumours also showed extension along the planum sphenoidale in continuity with the optic nerve lesions.[2] Subsequent authors have therefore questioned the true origin of bilateral ONSM.[7] It is probable that some of the reported patients with bilateral ONSM that predate MRI are not true ONSM but rather the spread of skull base meningiomas that have been mistakenly presumed to be ONSM.

ONSM typically affect middle-aged women, however the age of presentation of ONSM is slightly younger than for other meningiomas. The average age at presentation for ONSM is approximately

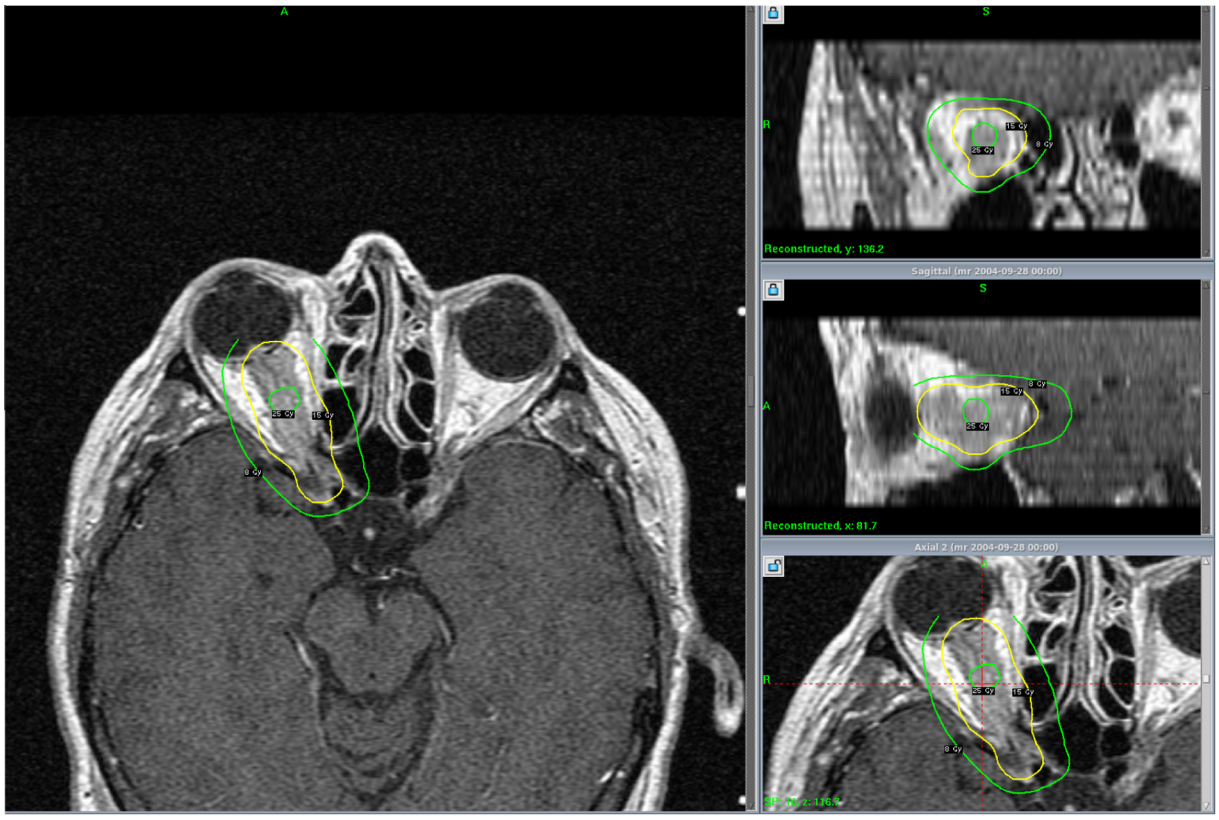

Fig. 2. Treatment planning scan of Patient 1:15 Gy with a 50% isodose covering 97% of a 4.3 cubic centimetre lesion. Inner circle = 25 Gy, middle circle = 15 Gy, outer circle = 8 Gy.