Haemangiopericytoma of Orbit Treated Using Intensity-modulated Radiotherapy

Amit Bahl, DN Sharma, Vellaiyan Subramani, Jyoti Basu, GK Rath
Department of Radiation Oncology, Institute Rotary Cancer Hospital, All India Institute of Medical Sciences, New Delhi, India

Orbital haemangiopericytomas are rare tumours. Surgery alone or followed by radiotherapy forms the mainstay of treatment. Late recurrences have been reported to occur with these tumours. Newer radiotherapy techniques such as intensity-modulated radiotherapy can improve treatment outcome in such patients.

Key words: *Hemangiopericytoma, Orbit, Radiotherapy*

Asian J Ophthalmol. 2006;8:254-5

Introduction

Haemangiopericytomas commonly occur in the retroperitoneum and lower extremity; tumours presenting intraconally in the orbit are rare. These latter tumours may present insidiously. Surgery is the initial modality of treatment but complete excision may not always be achieved.

Even with apparently complete excision, recurrences are well documented. We present a case of intraconal haemangiopericytoma of left orbit treated by surgical excision followed by radiotherapy using intensity-modulated radiotherapy (IMRT) technique. By use of this state-of-the-art radiotherapy technique, high doses can be given to the tumour-bearing area while sparing surrounding tissues.

Case Report

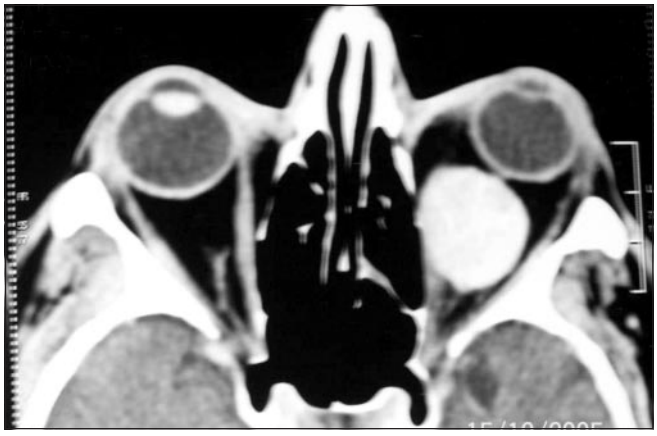
A 53-year-old male patient presented to our clinic with complaints of swelling of the left periorbital tissues for the last 1 year. This change was associated with progressive loss of vision in the left eye and intermittent diplopia.

Ophthalmic examination revealed proptosis in the left eye along with chemosis. Corneal sensations were normal. The visual acuity was 6/18 in the left eye and 6/6 in the right eye. The near vision in both eyes was N6 with a correction of +2.25 D spherical lens. Direct ophthalmoscopy revealed papilloedema of the left optic nerve head. An ultrasound examination revealed an intraconal mass compressing the globe. Routine blood examination was normal. Chest X-ray evaluation showed no abnormality. Magnetic resonance imaging showed proptosis of the left orbit. The globe was pushed anteriorly and outward by a large circumscribed intraconal soft tissue intensity mass lesion, measuring 30 x 28 x 25 mm (Figure 1). The mass showed variegated enhancement following contrast administration and it compressed the medial and inferior rectus muscle. There was no intracranial extension of the mass and optic chaism was normal. The patient underwent wide surgical excision of the tumour using a lateral orbitotomy approach.

Postoperative histopathology examination showed the tumour to be a borderline malignant haemangiopericytoma. The tumour was composed of round elongated cells with moderate pleomorphism and 2-3 mitotic figures per 10 high-power fields. Occasional mast cells were also identified. Immunohistochemistry for CD34 and vimentin was diffusely positive but negative for epithelial membrane antigen. A postoperative computed tomography (CT) scan evaluation showed residual disease.

In view of the incomplete excision, the patient was planned for postoperative radiotherapy with IMRT using an Eclipse radiotherapy

Figure 1. Preoperative magnetic resonance imaging scan of patient showing intraconal mass lesion.



Correspondence: Dr Amit Bahl, 334, AIIMS Doctors Hostel, Masjid Moth, South Extension-II, New Delhi 110 049, India.
Tel: (91 98) 6808 7950;
E-mail: dramitbahl@yahoo.com

planning system and a CLINAC 2300CD linear accelerator (Varian Medical Systems Inc., USA). Three IMRT treatment plans were generated and analysed. In the plan selected for treatment, the 95% isodose surface covered 97.39% of planning treatment volume (PTV). The plan was normalised at isocenter and 5 coplanar beams were used to produce the isodose distribution. The conformity index achieved was 1.11 and the uniformity index was 7.8%.

The dose maxima to the PTV was 108.2% and the dose minima was 86.1%. The V95 (volume receiving 95% of prescribed dose) was 97.39% and D95 (dose to 95% of volume) was 60.77 Gy. The maximum dose to left eye ball was 61.67 Gy, the maximum dose to the right eyeball was 9.675 Gy and the mean dose was 1.66 Gy. The right eye lens received 1.37 Gy as a maximum dose, with a mean dose of 0.75 Gy. The dose to the left temporal lobe was 56.53 Gy (maximum dose) and to the right temporal lobe dose was 3 Gy (maximum dose). The maximum midbrain dose was 15.3 Gy and maximum spinal cord dose was 0.67 Gy.

During treatment, the patient developed grade I eye reactions characterised by mild conjunctivitis and increased tearing and grade I skin reaction (Radiation Therapy Oncology Group acute radiation morbidity scoring criteria). Antibiotic eye drops were used to manage the treatment-associated conjunctivitis. The patient tolerated radiotherapy treatment well. His visual acuity in the opposite right eye at 6 months follow-up was 6/6. Visual acuity in the treated eye was 6/24 with a radiological complete response.

Discussion

Haemangiopericytomas account for only 3% of primary tumours of the eye.[1] Arising from pericytes of Zimmerman in the capillaries, they are slow growing tumours and present with proptosis and visual deterioration. These tumours can occur anywhere in the body and are known to occur rarely in the orbit.[2] They occur in the adult age group and both genders are equally affected. The tumours are usually surrounded by a pseudocapsule and are well circumscribed. Haemangiopericytomas behave in a benign manner but presence of mitosis (more than 4 per high-power field), foci of necrosis and increased cellularity are suggestive of malignancy.[3] Local recurrence and distant metastasis are well documented. Recurrences have been reported to occur as late as 10 years.[2,4,5] Surgical excision of the tumour is the usual treatment approach. Local recurrences are known to occur in incomplete excision.

Postoperative radiotherapy has been reported to reduce local recurrences. Tijl et al[4] have reported the use of radiotherapy post-operatively with good results. Setzkorn et al[6] have treated recurrent haemangiopericytoma of the orbit with postoperative radiotherapy to a dose of 64.8 Gy. At seven and a half year follow-up, their patient was disease-free.

Newer radiotherapy techniques offer improved specificity. IMRT is an approach to conformal therapy that conforms a relatively high dose to tumour tissue with minimal delivery to surrounding normal structures.

The IMRT procedure involves initial imaging of the tumour using a CT scan. The CT scan images are used to construct a 3-dimensional image of the tumour and surrounding tissues on the treatment planning system. The tumour-bearing area and organ at risk are then delinated on this 3-dimensional image. Dose constraints are prescribed to the critical structures and organs at risk. Using this inverse planning technique, the treatment plan is generated and transferred to a linear accelerator. A multi-leaf collimator system on the linear accelerator then conforms the radiation dose to the area of interest and at the same time modulates the fluence of radiation within the treatment field. Surrounding critical structures receive minimal or no radiation.[7] IMRT differs from conventional external radiotherapy in delivering a non-homogenous dose to the target area compared with a homogenous dose delivered in conventional treatment. This new modality also allows radiation dose to be escalated to higher levels than those used in conventional treatments.

IMRT has been used successfully in treating malignancies of the head and neck and in carcinoma prostate.[8,9] Its utility in orbital tumours appears promising. Our patient received a postoperative radiotherapy dose of 60 Gy using IMRT with minimal doses to surrounding brain structures and the opposite orbit.

We suggest that such orbital tumours requiring radiotherapy should be treated with newer modalities such as IMRT to prevent long-term sequelae of treatment on surrounding normal tissues and thus help improve the outcome and quality of life of the patient.

References

1. Coden DJ, Hornblass A. Orbital hemangiopericytoma. JAMA. 1990: 264:14:1861.
2. Henderson JW, Farrow GM. Primary orbital hemangiopericytoma. An aggressive and potentially malignant neoplasm. Arch Ophthalmol. 1978; 96:666-73.
3. Enzinger FM, Weiss SW . Hemangiopericytoma. In: Enzinger FM, Weiss SW, editors. Soft tissue tumors. 2nd ed. St.Louis: CV Mosby; 1988.
4. Tijl JW, Koorneef L, Blank LE. Recurrent hemangiopericytoma and brachytherapy. Doc Opthalmol.1992;82:103-7.
5. Gear HC, Kemp EG, Kackpereck A, et al. Treatment of recurrent orbital haemangiopericytoma with surgery and proton beam therapy. Br J Ophthalmol. 2005;8:123-4.
6. Setzkorn RK, Lee DJ, Iliff NT, et al. Hemangiopericytoma of the orbit treated with conservative surgery and radiotherapy. Arch Ophthalmol. 1987;105:1103-5.
7. Williams PC. IMRT: delivery techniques and quality assurance. Br J Radiol. 2003;76:766-76.
8. King A. IMRT for head-and-neck cancer improves quality of life and reduces xerostomia. Nat Clin Pract Oncol. 2006;3:7.
9. Guerrero Urbano MT, Nutting CM. Clinical use of intensity-modulated radiotherapy: Part II. Br J Radiol. 2004;77:177-82.