RADIO TERAPIA FRACCIONADA ESTEREOTÁCTICA EN EL MENINGIOMA DEL NERVIO ÓPTICO

FRACTIONATED STEREOTACTIC RADIOTHERAPY IN OPTIC NERVE SHEATH MENINGIOMA

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ABSTRACT

Case report: A 56-year-old woman with a right optic nerve sheath meningioma and visual loss, was treated with fractionated stereotactic radiotherapy. This resulted in an almost complete recovery of her visual acuity.

Discussion: Currently there are many different alternatives available for the management of an optic nerve sheath meningioma: observation, surgery, conventional radiotherapy and fractionated stereotactic radiotherapy. The last of these treatments has been demonstrated to preserve or improve vision in 2 out of 3 treated patients, with fewer side effects, and has not been associated with recurrent disease or tumor progression (Arch Soc Esp Oftalmol 2008; 83: 441-444).

Key words: Optic nerve, optic nerve sheath, meningioma, conventional radiotherapy, fractionated stereotactic radiotherapy.

INTRODUCTION

The management of optic nerve meningiomas (ONM) is currently a controversial issue. Therapeutic options include: resection, conventional radiotherapy and radiosurgery, and stereotactic fractionated radiotherapy (SFR). All of these pose risks, both in terms of visual prognosis and potential damage to adjacent structures (1,2).

SFR has few side effects. It preserves the functionality of the nerve, and seems to be a promising therapeutic option compared to the ones mentioned above, although long term effects are still to be determined.

RESUMEN

Caso clínico: Mujer de 56 años con meningioma del nervio óptico derecho y pérdida de visión tratada con radioterapia fraccionada estereotáctica, mejorando la visión casi en su totalidad.

Discusión: Actualmente existen diversas alternativas en el manejo del meningioma del nervio óptico: observación, cirugía, radioterapia convencional y radioterapia fraccionada estereotáctica. Esta última es la única técnica que ha demostrado preservar o mejorar la visión en dos de cada tres pacientes tratados, con menor tasa de efectos secundarios y ausencia de recidiva o progresión tumoral.

Palabras clave: Nervio óptico, vaina del nervio óptico, meningioma, radioterapia convencional, radioterapia fraccionada estereotáctica.
CASE REPORT

56 year old female, diagnosed 11 years ago. Primary ONM of the right eye. Examination showed unilateral proptosis with soft palpebral edema (Fig. 1), conjunctival-scleral ingurgitation due to a compromised orbital venous drainage, a restriction in ductions for the right eye, and binocular diplopia on elevation, levo-, and dextroversions, isochoric and normally reactive pupils, and 16 mm Hg intraocular pressure for both eyes. Initial visual acuity (VA) was one. Ocular fundus examination revealed a hyperemic, slightly elevated papilla in the right eye. Image tests identified a lesion compatible with intraconal ONM (Fig. 2), which was later confirmed by biopsy. Given the benign pathology and the VA measured, observation was prescribed, together with an MRI and visual field test every 6 months.

After a follow-up period of 4 years a lower peripheral scotoma was detected in the right eye, with deterioration of VA, and an increased IOP of up to 24-25 mm Hg. These were controlled by topical betablockers and latanoprost. One year later VA for the right eye was 0.3, IOP was 20 mm Hg (with the treatment), and the visual field (Fig. 3) and the ocular fundus (Fig. 4) have worsened significantly. Given this progression, an SFR treatment was agreed (total dose 50 Gy in 30 1.67 Gy fractions). Final VA for the right eye was 0.1.

Visual Field (Fig. 5), the FO (Fig. 6) and AV recovered gradually (38 months after treatment VA for right eye was 1); however, the palpebral edema, the proptosis, and ingurgitation have not improved, binocular diplopia is stable and well tolerated, and IOP is well under control with the treatment applied.

DISCUSSION

Orbital meningiomas may originate at the optic nerve sheath (primary tumors), from ectopic orbital
meningiomas, or by extension of intracranial meningiomas (3).

Primary ONMs account for 2% of all orbital tumors, and 1-2% of all meningiomas. However, they account for a third of optic nerve tumors, where these are the second most frequent cause, after gliomas. The most frequent tumor histology is benign meningotelial and transitional, with no documented reports of malignant occurrences (1-3).

The mechanisms altering the nervous function are: ischemia, distortion, elongation, demyelination, and interruption of the axoplasmic flow. A compression optic neuropathy is developed. This condition starts showing most frequently with a decreased VA. The classic triad (loss of VA, optic atrophy, and ophtcillar vascular shunt) appears only for 30% of all cases. Other signs include: proptosis, chemosis, palpebral edema, and limited motility (1,3). Although visual prognosis is poor, vital prognosis is excellent, with 0% mortality (3).

Neurofibromatosis type II must be discarded for all ONM patients, given the strong association between the two pathologies (4).

The new image techniques are essential for differential diagnosis, which includes: glioma, neuritis, sarcoidosis, syphilis, orbital pseudotumor, meningeal carcinomatosis, arachnoideal cyst, and perineural hematoma (1,3). These techniques also avoid the need for biopsies in most patients, thus preventing iatrogenia. Biopsies are usually required only for radiotherapy treatment on elderly patients with a progressive decrease in VA, or fusiform varieties (difficult differential diagnosis with gliomas) (1,3).

The objective of the treatment is preserving or improving vision, as systemic morbidity is non-existent, and probability of intracranial extension is uncertain. Resection is seldom prescribed, given the rates of recurrence and post-surgery blindness.
Resection is prescribed for: almost zero VA, bilateralization through chiasma or hypothalamus, disfiguring proptosis, or very young patients (more aggressiveness is assumed) (1-3). Observation is preferred for non-progressive meningiomas with a stable VA.

High-dose external conventional radiotherapy produces unilateral blindness by anterior or posterior neuritis 1-4 years after application. Other side effects may include retinopathy, dry eye syndrome, iritis, cataracts, and (only rarely) tumor induction.

SFR seems to improve tumor response to radiation, whilst decreasing normal tissue exposure, thus decreasing side effects. No reports have been made of tumor progression or recurrences. According to this series, this procedure has been proven to preserve or improve vision for 60-90% of patients (1-5). This is therefore the most appropriate therapeutic option when vision starts deteriorating, and no signs are present of intracranial extension.

Prospective studies will be necessary in order to determine the role of SFR as a first therapeutic choice for ONM, as well as its possible longer term side effects.

REFERENCES