MENINGOCELE, GLIOMA AND OPTIC NERVE 
MENINGIOMA: DIFFERENTIAL DIAGNOSIS AND 
TREATMENT

ABSTRACT

Purpose: After studying 3 clinical cases, we have reviewed the clinical and radiological characteristics of meningocele, meningioma and optic nerve glioma. The differential diagnosis and therapeutic management are also discussed.

Methods: Review of three clinical reports of three patients seen in our unit and a bibliographic search concerning the diagnosis and therapeutic management of these three entities at the present time.

Results: Differential diagnosis has to be based on a wide range of parameters: epidemiologic (age, race, sex, prevalence of the tumors), clinical (visual acuity, perimetry, Hertel exophthalmometry and funduscopy) and radiologic (computed tomography and magnetic resonance). Anatomopathologic study is required only rarely. The therapeutic options are: observation, surgery and radiotherapy.

Conclusion: A correct differential diagnosis is mandatory to be able to individualize the treatment for each entity.

Key words: Meningioma, meningocele, glioma, optic nerve.

RESUMEN

Objetivo: Estudiar a partir de 3 casos las características clínicas y radiológicas del meningocele, meningioma y glioma del nervio óptico (NO) para discutir su diagnóstico diferencial así como su enfoque terapéutico.

Método: Se revisaron las historias clínicas de tres pacientes afectos de las patologías anteriormente mencionadas que fueron estudiados en nuestro servicio, y se realizó una revisión bibliográfica sobre el diagnóstico y tratamiento actual de estas entidades.

Resultados: El diagnóstico diferencial debe basarse en un amplio abanico de aspectos: edad, raza, sexo y frecuencia de la tumoración como factores epidemiológicos a tener en cuenta. Clínicamente los pilares fundamentales son: agudeza visual (AV), campimetría, exoftalmometría Hertel y funduscopy, que nos servirán para controlar la evolución del proceso. Las características radiológicas que nos aportan la resonancia magnética (RM) y la tomografía computerizada (TC) como técnicas de elección nos ayudan a diferenciar un cuadro de otro. En raras ocasiones hay que recurrir al estudio anatomopatológico para confirmar la entidad. Las opciones terapéuticas son observación, cirugía, quimioterapia y radioterapia en función de las características del caso.
INTRODUCTION

The surgical pathology of the optic nerve (ON) exhibits some peculiarities characterized by its anatomic location which hinders accessibility. Intracranial invasion risk and visual deterioration requires us to typify each type of injury. The ON meningocele, also described in the literature as arachnoid cyst or ectasia of ON sheaths, is a normal fibrovascular proliferation which causes a benign distention of the meningeal. These cysts may simulate ON neoplasia, particularly meningiomas. Primary ON meningiomas account for a third of primary ON tumors and are the most frequent neoplasia affecting the ON sheaths. Even though ON gliomas are infrequent, they are the most frequent cause of ON neoplasia.

SUBJECTS, MATERIAL AND METHODS

On the basis of 3 cases we describe the clinical and radiological characteristics of an ON meningocele, meningioma and glioma to discuss their differential diagnostic and treatment utilizing the latest advances found in the literature.

RESULTS

Case 1

A male patient, 39 years old, who attended the emergency service due to proptosis with one year evolution in both eyes (BE), without reduction of visual acuity (VA). He did not exhibit any other symptom or history of interest. In the exploration, the patient exhibited a VA of 0.5 in the right eye (RE) (referred as amblyopic by the patient) and 0.8 in the left eye (LE). Hertel exophthalmometry was of 25 mm in RE and 23 mm in LE. Biomicroscopic and tonometric exploration was normal. The eye fundus study exhibited a papillary edema in the RE. The Farnsworth color test was normal. In the campimetric study, the LE was normal but the RE exhibited an inferonasal defect which was not strictly a quadrantnaopsia. A computerized tomography (TC) was requested and reported as ectasia/thickening of the sheaths of both ON in the context of bilateral exophthalmos, more marked in RE (fig. 1). Suspecting the presence of bilateral meningocele, it was decided to explore the patient regularly without additional diagnostic or therapeutic actions. At present, 16 years later, the patient exhibits a VA of 0.6 in RE and 0.8 in LE. The exophthalmometry and campimetry have remained stable. In the ocular fundus, the papillary aspect has hardly varied. The regular explorations continue.

Conclusión: El diagnóstico diferencial de estas patologías es difícil, y hay que basarse en sus características clínicas y radiológicas para así orientar correctamente su tratamiento, individualizando cada caso y teniendo en cuenta el comportamiento relativamente benigno de estos tumores para evitar una decisión terapéutica precipitada.

Palabras clave: Meningioma, meningocele, glioma, nervio óptico.

Fig. 1: Bilateral meningocele (arrows). Computerized Axial Tomography.
Case 2

A 38-year woman referred from a regional hospital to our centre for assessment of ON tumor. Her history includes a progressive VA reduction in the past year, exacerbated in the last 2-3 months. In July 2005, her VA was of 0.7, in August 0.5, in October 0.1, in November finger counting at one meter and in December 2005, when she was referred to us, she exhibited RE amaurosis and absolute afferent pupillary defect. An exophthalmometry did not reveal asymmetries between both eyes. From the beginning of the study, the patient exhibited RE papilla edema with normality in LE. The CT was reported as asymmetry in optic nerves, with slight thickening of the right one vis-à-vis the contralateral nerve. In December 2005 an orbitary nuclear magnetic resonance (NMR) was made, reported as diffuse and concentric thickening of the dural sheath of the right ON, with absence of enhancement of the nerve and extending throughout the intra-orbitary length in the optic conduit and intra-cranial extension, reaching the anterior right clinoids, suspecting a right ON meningioma (fig. 2). After explaining the therapeutic options, the patient declined any type of action and at present is following regular check-ups.

Case 3

Male, aged 3, under study by neurologists due to suspected neurofibromatosis type 1 because he fulfilled at least one major diagnostic criterion (disseminated darkish spots). The patient exhibited RE axial exophthalmos dated 5 months back. VA with Pigassou test was of 1. A NMR reported a well-defined, homogeneous and isodense thickening of the ON with intact meninges compatible with the diagnostic of ON glioma in its intracanalicular and intraorbitary portion (fig. 3). This confirmed the neurofibromatosis type 1 diagnostic due to fulfilling 2 major criteria. As the patient did not exhibit lagophthalmos and his VA was preserved, we opted for explorations at 3-month intervals the first year and half-yearly controls the second year. Even though the optic glioma progression criteria are not universally defined, in this case four years after diagnostic both the VA and radiological size of the injury remain stable. Accordingly, we are not considering chemotherapy and check the patient every 6 months, with an annual NMR.

DISCUSSION

The differential diagnostic between said pathologies is not always easy; it must be based on a mul-
ti-factorial approach. For the majority of case, epidemiological data, presentation and clinical evolution, as well as the radiological characteristics of the injury, are sufficient data to diagnose these pathologies. Anatomic and pathological studies are necessary only exceptionally to define said entities. From the epidemiological viewpoint, meningoceles are exceptional entities. The only case revision found in the literature (1) refers to 12 arachnoid cysts. Accordingly, it is very difficult to determine its gender distribution and the age group in which these occur with greater frequency. Meningiomas are the most frequent ON sheath tumor, typical among women in the 30-50 age group, and account for 2% of orbital tumors (2). Gliomas are the most frequent ON neoplasia. Most appear in young people under 20 (85%) and particularly in children under 10 (65%) and are typically associated to neurofibromatosis type 1 (Von Recklinghausen disease) in approximately 25% of cases, which are usually multifocal and/or bilateral (3). For this reason, they are considered as one of the six diagnostic criteria of this entity. About 15% of neurofibromatosis type 1 patients exhibit an ON glioma (4). Clinics help us to guide each case, particularly when we focus on the information provided by the VA, the ocular fundus appearance and the proptosis characteristics. In what concerns changes in VA, patients with meningocele do not generally suffer variations thereof as the ON is compressed only slightly (1). Gliomas typically debut with dischromatopsia and slow reduction of VA (5), although this is sometimes difficult to assess because it usually appears at an early age. In meningiomas, the VA reduction is early and evolves faster (2). In what concerns the ocular fundus, the arachnoid cyst usually produced papillary edema which remains in the same severity stage throughout the entire evolution. In meningiomas, where papillary edema also appears, it evolves together with the tumor evolution but the most typical event is the appearance of opto-ciliar shunts. The majority of gliomas exhibit a certain degree of optical atrophy and also associate papillary edema, particularly those limited to the ON and not to the chiasm or higher portions of the optical pathways.

Once the clinical suspicion is raised, it is necessary to request high resolution images, CT or NMR. The CT of an arachnoid cyst can reveal a lobule-like or cystic image, but with the ON preserved and without intra-injury calcifications (1). In a meningioma, contrast capture is usually homogeneous. The «rail track» sign, originally described in these tumors when the ON sheaths are thickened and dense, forming a translucent centre, can also be observed in other pathologies such as lymphomas or even peri-optical orbital pseudo-tumors. The most typical case in CT of meningiomas is the appearance of calcifications, generally surrounding the ON and compressing it, thus reducing its diameter (2), in contrast with the image of gliomas where the ON appears expanded with sinusuous appearance, the meninges intact and rarely exhibiting calcifications (6).

If, after a clinical and radiological study, the conditions still is doubtful for a diagnostic, which is rare, we can make a histopathological study of the injuries. Ectasie of the ON sheaths are simple proliferations of meningoepithelial cells which adopt a macropscopic cystic shape which gives no room for doubt (1). When approached, the majority of orbital meningiomas extend through the dura mater. This does not occur with gliomas. In addition, in the anatomopathological study we can observe the proliferation of cells from the external layer of the arachnoids become organized in rings which can have some degree of hyaline degeneration and typically become calcified forming the classical psammoma bodies (7). In gliomas, the primary proliferation is at the expense of astrocytes, which could stimulate a fibrovascular proliferation of arachnoid meningoepithelial cells arranged forming psammoma bodies, but in this case they calcify only exceptionally (8).

After typifying the condition, we must consider the right therapeutic approach for each clinical phase. Meningoceles, that can be congenital, idiopathic or post-traumatic, usually do not require treatment (1,9). However, a close follow-up of the patient is required because the literature (10) describes some cases of papillary edema progression or VA reduction which oblige therapeutic intervention. Initially, we can consider the option of treating the patient with systemic carbonic anhydrase inhibitors based on the possibility that the osmotic gradient between the brain sub-arachnoid and peri-optical space causes a reduction in the ectasia size. In symptomatic meningoceles cases, the role of IV corticoids has not been discussed in existing publications. Puncturing and aspiring these cysts is reserved for post-traumatic cases where the dysfunction of the ON is manifest and the recovery of the papillary edema and VA improvement have better chances.
In meningioma management we must always ask ourselves if the patient needs treatment or he can wait to be treated for the tumor. Even though the percentage of metastasis and mortality is very low, at present it is recommended to treat patients for a number of reasons (2,11-15): the VA reduction occurs in 85% of cases and, in up to 38% of cases with intra-canaliculare involvement, alterations in the contralateral eye have been described. In addition, observing the patient without treating the tumor while its size increases and visual deterioration progresses could diminish the probability of success of a therapeutic intervention in the future. In patients under 20, it is assumed that the behavior of meningiomas is more aggressive and therefore the option of observing without treating should be taken with prudence on a case by case basis.

The cases selected for observations without treatment would be those which maintain a good VA and do not exhibit associated morbidity (such as proptosis, among others), with annual ophthalmological, radiological and neurological check-ups, or every six months if the patient is under 20.

For the cases selected for treating, i.e., the meningiomas which gradually worsen the patient’s VA or cause severe exophthalmos or have an intra-cranial expansion risk, we have two therapeutic weapons: neoplasia surgery and radiotherapy. The surgery for removing a primary meningioma from the ON is associated to a high risk of blindness and, if residual tumor mass remain, there is a high risk of malignization. Accordingly, this option should be reserved only for anteriorly located primary neoplasias with exophytic growth and clear focal infiltration of the dura. Tumor removal benefits patients with visual deterioration only exceptionally, it could even be argued that in some cases this procedure is associated to massive orbit infiltration by the tumor (2), and even though it is possible that, in cases where the tumor is distended by spinal fluid trapped between its sheaths, the removal might halt the visual deterioration, at present it is an abandoned practice (16) as well as biopsies due to their high co-morbidity (17).

Radiotherapy was initially used as adjuvant to surgery, but in 2002, in a retrospective series of 64 cases (14), Turbin demonstrated that stereotactic fractioned radiotherapy as single treatment for primary ON meningiomas is the therapeutic option which yields the best visual results (80% of patients maintain or increase their visual function) and therefore it is considered as the treatment of choice for the meningiomas that require treatment (12,18). Still, some authors (including Turbin himself) defend the usefulness of surgery as an adjuvant measure to radiotherapy in selected cases (19,20).

The most controversial treatment is that of gliomas (3) and will always depend on the localization of the tumor, its growth rate and size, as well as the symptoms it produces. We have yet to reach universal consensus about the criteria to be applied to define whether the glioma is progressing or not, with evidence of increased size in NMR and/or VA loss being the most utilized criteria. When the tumor is restricted to the ON, the vital prognosis is excellent and, if the patient maintains a useful VA, a conservative management and regular follow-up of the disease is indicated. Therefore, gliomas must only be treated when the tumor progression is evident. At present, chemotherapy is the treatment of choice, with a combination of vincristine and carboplatinum being the most widely recommended options (21). Radiotherapy is reserved only for the cases where chemotherapy combinations have failed because it has a high risk of major complications (endocrinopathies and cerebro-vascular accidents). Fortunately, radiation techniques are increasingly selective and act on the core of the tumor with minimum effect on adjacent structures (fractioned stereotactic radiotherapy and gamma knife-guided radiotherapy). However, it remains a second-line therapeutic option because the risk of malign transformation of the glioma after radiotherapy persists.

The surgical option should be considered only in cases of tumors confined to the ON with clean edges in the context of a patient with blindness, severe exophthalmos or when the extension through the intra-cranial segment threatens the chiasm. If the latter is involved, the risk of death due to hypothalamus or third ventricle compromise increases to 28%. In these cases, surgery is associated to a high rate of morbimortality and does not seem to increase survival, whereupon radiotherapy is preferred.

REFERENCES