OPHTHALMIC PRESENTATION OF TWO CASES OF ESTHESIONEUROBLASTOMA

DEBUT OFTALMOLÓGICO DE DOS CASOS DE ESTESIONEUROBLASTOMA


ABSTRACT

Purpose/Method: To report the ophthalmologic presentation of two cases of esthesioneuroblastoma (ENB). A comprehensive ophthalmic assessment was made, including magnetic resonance imaging. A paranasal sinus mass was biopsied in one case and a nasal mass biopsied in the other.

Cases Report/Discussion: Two women, aged 85 and 32 years respectively, presented to the emergency room with ophthalmic symptoms and signs. The first reported orbital pain and eyelid edema and the second, who was 22 weeks pregnant, reported a spontaneous lower eyelid hematoma, orbital pain and rhinorrhea. ENB is a rare malignant tumour and its presentation with ophthalmologic symptoms and signs is very infrequent (Arch Soc Esp Oftalmol 2008; 83: 317-320).

Key words: Esthesioneuroblastoma, eyelid edema, proptosis.

RESUMEN

Objetivo/Método: Presentar el debut de dos casos de estesioneuroblastoma (ENB) con manifestaciones oftalmológicas. Se realizó una exploración oftalmológica completa y estudios de imagen consistentes de Resonancia Magnética (RM). En un caso se realizó biopsia de una masa sinusal y en el otro de una masa nasal.

Caso clínico/Discusión: Dos mujeres de 85 y 32 años, esta última embarazada de 21 semanas, acudieron de urgencia por síntomas y signos oculares. El primer caso refería dolor orbitario y edema de párpado y el segundo hematoma palpebral inferior espontáneo, dolor orbitario y rinorrea. El ENB es un tumor maligno poco frecuente que puede debutar con sintomatología oftalmológica.

Palabras claves: Estesioneuroblastoma, edema palpebral, exoftalmos.
**INTRODUCTION**

Esthesioneuroblastoma (ENB) or olfactory neuroblastoma is an infrequent malignant tumour that occurs in the neuroectodermal cells of the olfactory sensory epithelium. It represents 5% of all malignant tumours of the sinonasal tract. It can appear at any age but normally has a bimodal distribution with two peaks, in the second and sixth decade of life. It is a slow growing tumour but with an aggressive, with a tendency to spread early to the anterior cranial fossa and produce have regional and distant metastasis (1).

**CASE REPORTS CLINICAL CASES**

Case 1

An 85-year-old woman of 85 years with a clinical background of arterial hypertension, diabetes, who had breast cancer in 1994. Cataract phacoemulsification with lens in sac of left eye (LE) with topical anaesthesia without any incidents in 2000. The patient presented returned 39 days after surgery due to pain in the supero-internal angle of the left orbit and nocturnal palpebral edema of LE. The visual acuity (VA) was 0.8 in both eyes (BE). Cober Test 0º and slight ptosis of LE. Increase in pain upon palpation and pressure at the level of the trochlea. The anterior segment (AS) was normal in both eyes and the intraocular pressure was 16 and 19 mm Hg. The back of the eye was normal in both eyes. After a diagnosis of trochleitis, 1cc of triamcinolone was injected into the left trochlear zone. Due to the lack of response a Magnetic Nuclear Resonance (MRI) was requested that showed an intrasinusal solid mass with spreading to the anterior cranial fossa compatible with ENB (fig. 1). The nasal biopsy of the sinusal mass confirmed a esthesioneuroblastoma of high mitotic level and pleomorphism: level III-IV of the Hyams histopathological classification (1988), stage C of the Cadiz classification, stage T4N0M0. The patient rejected any type of treatment but the ENT unit gave her a bimonthly follow up. Six months later she was admitted to Emergencies due to intense cephalalgia, a generally bad state, fever, with double incontinence and cognitive deterioration secondary to neoplastic cerebral disease of two months of duration. Palliative treatment was given and with the patient dying seven days after admittance.

Case 2

A 32-year-old woman of 32 years, 21 weeks pregnant and diagnosed with vasomotor rhinitis and nasal polyps. On arrival she presented spontaneous hematoma in the internal third of the inferior eyelid, pain and blurry unclear vision of in LE. The patient referred to cephalalgia and rhinorrhea rhinorrhea of months of evolution. An AV of 1 in RE and of 0.5 in LE was determined objectified, left afferent pupillary defect, a left exophthalmos of 21mm ( RE 18mm, intercanthus intercanthal distance of 100) (fig. 2) and a complete paralysis of the right VI pair par (fig. 3) with left inervational endotrophy (fig. 4). She also presented ann inferior palpebral inferior hematoma (figs. 3 and 4). The SA of the AO was normal. The papilla of RE was normal and exhibited presented elevation of the superior and inferior poles without any presence of choroidal coroidal folds in the LE. The ultrasound showed an increase of the space of the optic nerve she-
athes and a nasal hypoechogenic image. A diagnosis of left orbital mass was reached with possible intracranial extensions spreading to the right cavernous sinus. The MRI showed an aggressive tumor that was affecting the ethmoidal fossas, left maxililar sinus and an anterior portion of the skull base suspected of ENB (fig. 5). The ENT examination of the nasal fossas objectified a friable and bleeding mass coming out of the left medium meatus surpassing the level of the medium cornet. The morphological and immunohistochemical findings of the nasal biopsy were compatible with esthesioneuroblastoma grade III-IV of the Hyams histopathological classification, stage C of the Cadiz classification, stage T4NOMO. It was considered to be inoperable due to the stage of the tumor, and a chemotherapy protocol for pregnant women was chosen for the pregnant women to reduce the tumor size, followed

Fig. 2: RE Exophthalmos more noticeable looking looking down.

Fig. 3: Limitation of abduction in RE complete paralysis of right IV pair par.

Fig. 4: RE endotrophy endophthy in primary looking gaze position with predominant right eye fixer due to it being the eye with having the best vision.

Fig. 5: MRI T1 with gadolinium: large mass located in nasal fossas. Spreading to the left maxilla sinus invading the left orbit. Spreading to the anterior portion of skull base. Involvement Affectation of inferior orbital fissure and right cavernous sinus.
DISCUSSION

The above ENB cases of ENB presented were diagnosed after consulting the ophthalmologist ophtalmologist because of symptoms and orbital signs. Only the second case presented nasal symptoms: obstruction and rhinorrhea rhinorhea. Although ocular symptoms and signs during ENB are frequent and found in around 53% of the cases (2) it is not common that these precipitate the diagnosis.

The persistent nasal obstruction and epistaxis are the most common symptoms at presentation. Nevertheless, ocular symptoms and signs can be the first to appear (periorbital pain, epiphora, exophthalmous, visual alterations...) (2,3) but using them for diagnosis is not frequent because they appear later than non ocular ones symptoms.

However, treatment is controversial. It seems that the best results are obtained with a combination of surgical resection and radiotherapy, with or without chemotherapy (4). The survival rate at 5 years is between 51-81%. In general, these are tumors that are very advanced when diagnosed diagnosis present are very advanced, as described in our cases, and this leads to poor survival rates.

ENB is a tumor with a tendency to recur locally. The most frequent metastasic lesions are in the cervical ganglions or lungs. The recurrence time may be very long.

In conclusion, ENB is a very infrequent tumor. Nevertheless, it is necessary that for ophthalmologists to be is aware of this serious disease when seeing all patients with maintained nasal obstruction or recurring epistaxis or exophthalmos exophthalmus to face up to the diagnosis and handling of this tumor in a multidisciplinary manner. The essential tests for diagnosis of ENB are image techniques (CT or MRI) and the histopathology of the tumor by biopsy.

Finally, the ophthalmologist should carry out a follow up of these patients treated by radiotherapy due to the ocular and the lacrimal system (glaucoma, epiphora) complications that can arise.

REFERENCES