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An accidental diagnosis of optic nerve meningioma in a patient affected by Thyroid Eye Disease

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Abstract

A 42-year-old woman presented to our hospital owing to a history of right-sided proptosis of 4 months duration, reporting no previous trauma, fever, or recent sinusitis. Her medical record included a diagnosis of Graves' disease. The best corrected visual acuity (BCVA) was 20/20 and a 30.2 visual field test was normal. A 3-Tesla magnetic resonance imaging (MRI) revealed an orbital apex meningioma approaching the walls of the sulcus chiasmaticus. A subsequent 60.4 perimetry test showed bilateral nasal visual field defects, thus confirming the involvement of the optic nerve. The reported case focuses on the differential diagnosis with Thyroid Eye Disease (TED) and the subsequent follow-up.

Keywords: optic nerve, meningioma, thyroid eye disease, Graves' disease

Introduction

The most common cause of both bilateral and unilateral exophthalmos in the adult population is Thyroid Eye Disease (TED) [1]. The rate of ophthalmopathy cases among hyperthyroid patients reaches up to 90%, with a much lower value among hypothyroid patients ranging between 0.2 and 8.6% [2].

Despite being usually associated with TED, unilateral exophthalmos has a broader inventory of diagnostic possibilities to be excluded compared to bilateral exophthalmos. The pathological conditions more extensively regarded in the differential diagnosis comprise: traumatic causes, congenital abnormalities, infectious diseases, granulomatous, inflammatory and vascular disorders, and orbital neoplasms [1].

We report the case of a 42-year-old woman with unilateral proptosis with particular stress on the difficulty entailed in formulating a correct differential diagnosis with TED and the importance of the valuable information added by proper use of diagnostic devices.

Case Presentation

A 42-year-old female presented to the hospital complaining of right-sided proptosis without pulsation or bruit which developed gradually over 4 months, reporting no history of trauma, fever, or recent sinusitis. The patient denied diplopia. Her past medical record was relevant for thyroid disease. In fact, Grave's thyroiditis had been diagnosed 2 years before and propylthiouracil (PTU) 3 x 100 mg/day prescribed. The patient underwent visual acuity assessment, slit-lamp examination, intraocular pressure evaluation with Icare®, fundoscopy and visual field testing (Humphrey 30.2 program) [3,4]. We used a Hertel exophthalmometer to assess the severity of proptosis. An eyelid activity score (EAS), where 0 = normal, 1 = upper lid margin just above the upper limbus (showing the "whites" of the eye), 2 = limbus to eyelid distance ≥1.5mm but <2mm and 3 = limbus to eyelid distance ≥2mm, was estimated for each eye [5]. The Clinical Activity Score (CAS) and the NOSPECS score were used in the evaluation of disease activity and severity, respectively [6]. An endocrinological examination and thyroid function tests were required. We performed a 3T MRI (Discovery–MR750) to evaluate the extraocular muscles involvement. A visual field testing (Humphrey 60.4 program) was also completed for a thorough diagnostic evaluation.

On the first examination, the patient had a best corrected visual acuity (BCVA) of 20/20 in both eyes. The von Graefe's sign was not detected and the EAS score was 0 in both eyes. She didn't have lagophthalmos. The pupillary response, intraocular pressure, and fundus examination were normal. Values of Hertel exophthalmometry were 18 mm in the right eye and 16 mm in the left eye. The CAS score was 1 and the NOSPECS score was 1 as well. Thyroid function tests showed a good control of the disease.

The endocrinologist confirmed the diagnosis of Grave's disease and the systemic treatment previously prescribed. The 30.2 visual field test was normal **(Figure 1a).** We performed a 3T MRI with Gadolinium-DTPA enhancement which unveiled a small soft tissue mass at the apex of the right orbit within the enhancing meningioma in the optic canal with growth along the walls of the sulcus chiasmaticus **(Figure 2).**

A diagnosis of unilateral proptosis associated with optic nerve meningioma was carried out. A 60.4 visual field test

showed bilateral nasal defects **(Figure 1b).** After two months, the patient's BCVA was 20/20. An increase in the severity of proptosis with a value of 19 mm in the right eye was noted and the MRI showed an expanded lesion involving the ophthalmic vein **(Figure 3).**

A neurosurgical assessment ruled out either surgery or radiotherapy and a conservative approach with close supervision of any visual field and visual acuity deterioration was advised. The follow-up visits carried out every 6 months do not show significant modifications in the neuroradiological and ocular outcomes.

Discussion

This case strongly upholds the importance of a thorough medical history in the evaluation of proptosis sometimes supported by appropriate instrumental investigations to formulate a definitive diagnosis. One should always keep in mind the likelihood of an orbital neoplasm as the cause of unilateral exophthalmos even in a patient carrying a diagnosis of thyroid disease older than 2 years. The evaluation through a 60.4 visual field test could be a fast-diagnostic tool in cases of unilateral proptosis prior to performing an MRI. In fact, in our

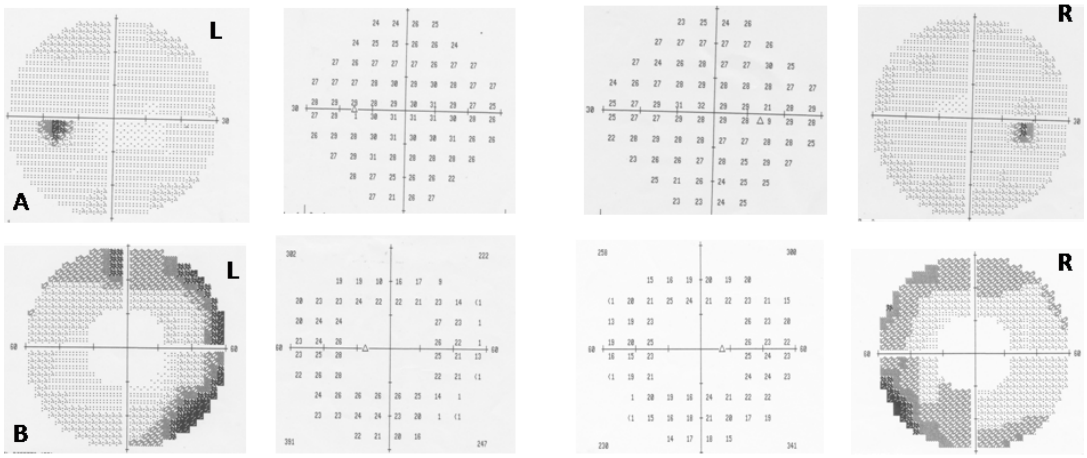

Figure 1. *A) Humphrey automated visual field perimetry (30.2 program) was normal in both eyes. B) Humphrey automated visual field perimetry (60.4 program) showed a bilateral nasal defect.*

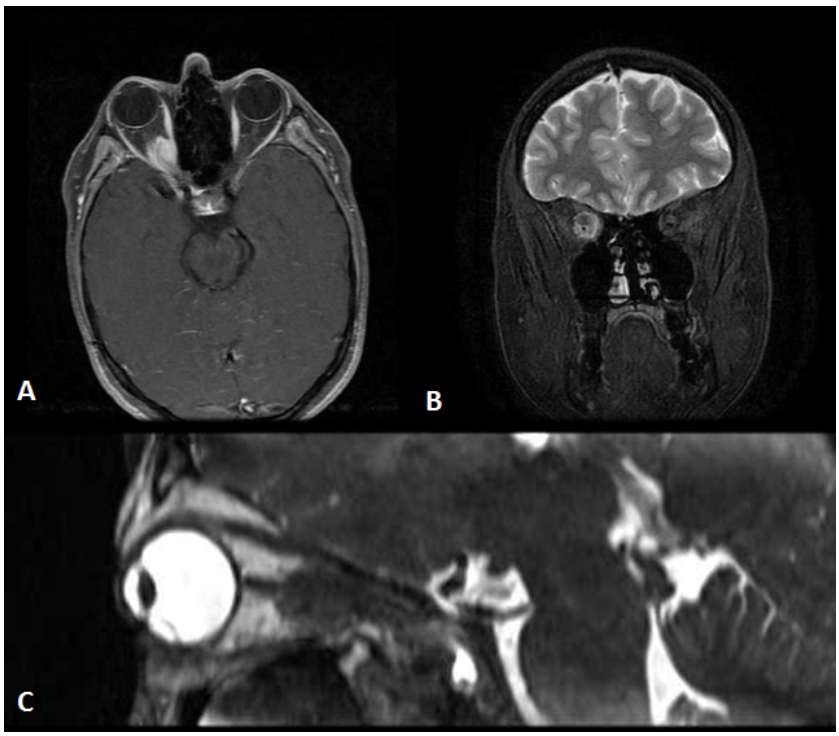

Figure 2. *3T MRI A) Axial T1-weighted sequence showing a hyperintense mass with defined limits localized at the apex of the right orbit and partially in the optic foramen, highly suggestive of meningioma. B) Coronal T2-weighted sequence showing a hyperintense mass inside the orbit encasing the intraorbital portion of the optic nerve. C) Sagittal FIESTA sequence showing an iso-hypointense mass in the intraorbital portion of the optic nerve.*

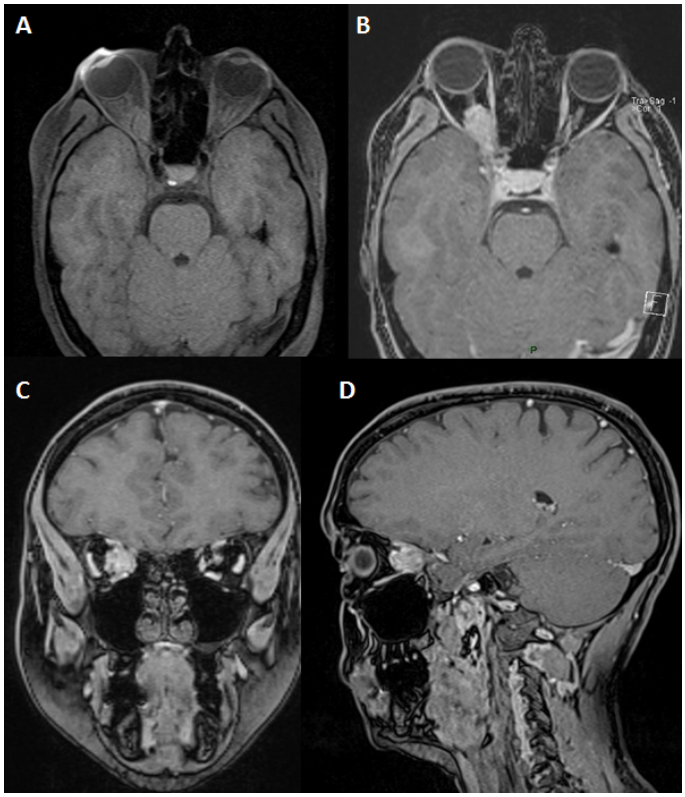

Figure 3. *3T MRI. Axial T1 sequences before (A) and after G-DTPA administration (B) showing an isointense tumor that enhances intensely and homogeneously. The coronal T1-weighted sequence shows a multilobulated tumor with positive enhancement located on the medial aspect of the right orbit close to the ipsilateral medial and inferior rectus muscle .The sagittal T1-weighted sequence shows a hypervascular tumor localized at the apex of the right orbit and around the intraorbital portion of the optic nerve.*

case this test showed a defect in the visual field not associated with TED and it could plausibly aid in the differential diagnosis of controversial cases.

Optic nerve meningiomas are rare tumors of the anterior visual pathway [5] and the lesions can come into close contact with the optic nerve and its vascular supply. The surgical excision may be difficult and can result in postoperative blindness in the affected eye [7-11].

Radiotherapy has been shown to have good efficacy with local tumor control and has been recommended as the best option for most cases of progressive or advanced disease [9-12] but concerns regarding the safety of this treatment are still being discussed.

In conclusion, the preserved visual acuity of the patient described in our case report led to refrain from choosing the treatment described above, rather supporting more conservative measures while awaiting for an eventual deterioration of vision caused by any ensuing tumor expansion.

Conflict of interest

None of the authors has any financial interest in the information presented in this manuscript. The authors alone are responsible for the content and writing of the article.

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