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The irreversible consequences of late diagnosis of pituitary macroadenoma in a young patient

As consequências irreversíveis do diagnóstico tardio de macroadenoma de hipófise em paciente jovem

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Abstract

Pituitary tumors represent approximately 15% of all brain tumors and depending on size, pressure optic chiasma, resulting in impaired visual func-tion that manifests itself as defective in the visual field, decreased acuity visual and color vision. The ob-jetive of the present study was to report a case of pitui-tary macroadenoma with compression of optical chiasma and visual field de-fect, initially treated as glaucoma, leading to a late diagnosis and treatment.

Keywords: *Pituitary gland; Pituitary diseases; Pituitary neoplasms*

Resumo

Os tumores de hipófise representam aproximadamente 15% de todos os tumores cerebrais e dependendo do tamanho, pressionam o quiasma óptico, resultando em comprometimento da função visual que se manifesta como defeitos no campo visual, diminuição da acuidade visual e da visão das cores. O objetivo do presente estudo foi relatar um caso de macroadenoma de hipófise com compressão do quiasma óptico e defeito no campo visual, tratado inicialmente como glaucoma, levando a um diagnóstico e tratamento tardio.

Descritores: Hipófise; Doenças da hipófise; Neoplasias hipofisárias

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Introduction

Pituitary tumors stand for approximately 15% of all brain tumors. Pituitary adenoma and meningioma are the most common causes of compressive optic neuropathy, whose cases often show visual symptoms and cephalea. Growing tumors press the optic chiasm and it results in impaired visual function manifested as defects in the visual field, decreased visual acuity and decreased color vision.[1]

Vision parameter comparison suggest that patients presenting early diagnosis and treatment have better prognosis. They show improved visual acuity, midline deviation and visual field after diagnosis and early surgery. Diagnosis delay significantly compromises preoperative and postoperative visual acuity, therefore, the early detection of visual pathway dysfunction causes can change medical treatments and reduce the incidence of irreversible damage to the optic nerve.[2,3]

The aim of the current study was to report a pituitary macroadenoma case with compression of the optic chiasma and visual field defect. It was initially treated as glaucoma, which led to late diagnosis and treatment.

Case Report

Man, aged 30 years, white, married, glazier, has complained of low visual acuity (LVA) in both eyes for 2 years. He refers to cephalea with progressive LVA. Patient reports to have been previously followed up in another service, where he was diagnosed and treated for glaucoma for 1 year and a half. Personal and ophthalmologic background: head trauma and blunt trauma to the left eye (LE) 3 years ago.

The ophthalmologic exam showed visual acuity with right eye correction (RE) 1.0 and LE without Luminous Perception. RE biomicroscopy showed no changes, LE presented fixed mydriasis. RE ocular reflex showed no changes, LE reduced photomotor and consensual reflex presented relative afferent pupillary defect. Extrinsic eye movement was preserved. Applanation Tonometry at 10am: 12 and 14 mmHg. Fundoscopy/retinography showed RE medium papilla with 0.3/0.3 excavation with nasal pallor, OE medium papilla with 0.3/0.3 excavation with pallor (Figure 1). Visual field exam, optical coherence tomography (OCT) and magnetic resonance imaging of the skull were requested.

Visual field exam reported RE temporal scotoma respecting the midline and LE absolute scotoma (Figure 2). Magnetic resonance imaging of the skull with emphasis on sella turcica: intrasellar and suprasellar lesion (4.0 x 3.0 x 2.5 cm), it encompassed the intracavernous segment of the internal carotid arteries, which caused compression and cephalic displacement of the optical chiasm. Cystic lesion in the left middle fossa (8.0 x 5.0 x 4.8cm), with mass effect, and slight displacement of the midline to the right associated with hypoplasia of the left temporal lobe (Figure 3). Optical coherence tomography (OCT) of RE showed optical nerve with nerve fiber layer loss in the nasal and temporal region, preserved lower fibers and borderline in the upper region, LE with diffuse loss of the nerve fiber layer (Figure 4). The patient was referred to neurologist and neurosurgeon for treatment and follow-up, he was recommended to surgery for tumor removal. Ophthalmologic examination after 6 months reported that visual acuity and field loss remained, there was no prognosis improvement.

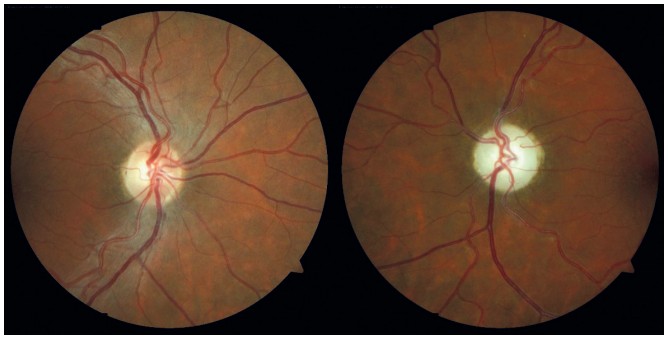

Figure 1. Retinography: RE: medium papilla with 0.3/0.3 excavation with nasal pallor, LE: medium papilla, 0.3/0.3 excavation with pallor.

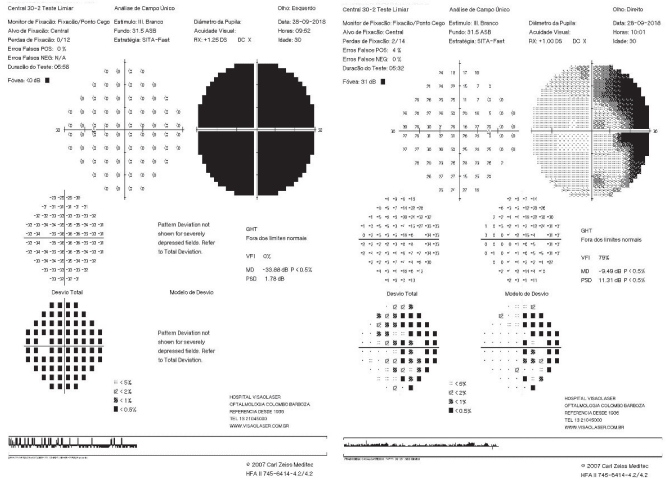

Figure 2. Visual field: RE: temporal scotoma respecting the midline, LE: absolute scotoma

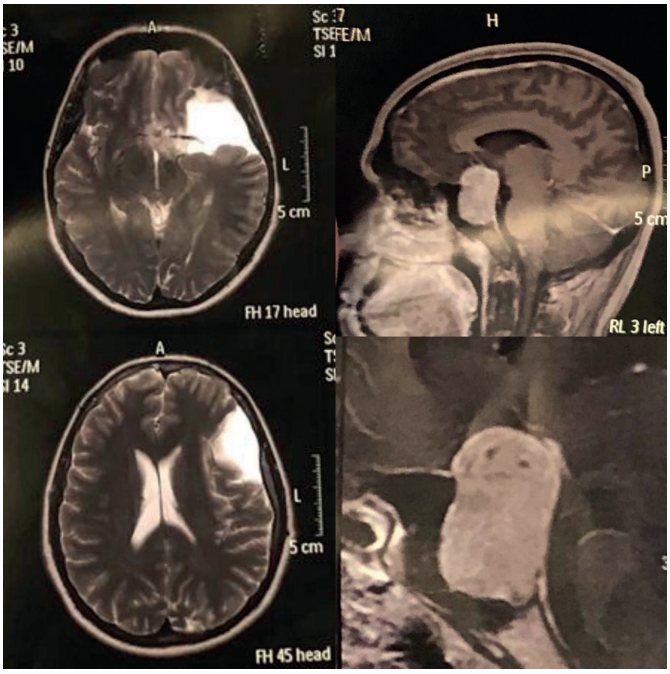

Figure 3. Magnetic resonance imaging of the skull with emphasis on sella turcica: intra and suprasellar lesion (4.0 x 3.0 x 2.5 cm) , it encompasses the intracavernous segment of the internal carotid arteries, compression and cephalic displacement of the optic chiasm. Cystic lesion in the left middle fossa (8.0 x 5.0 x 4.8 cm) with mass effect and slight displacement of the midline to the right, associated with hypoplasia of the left temporal lobe.

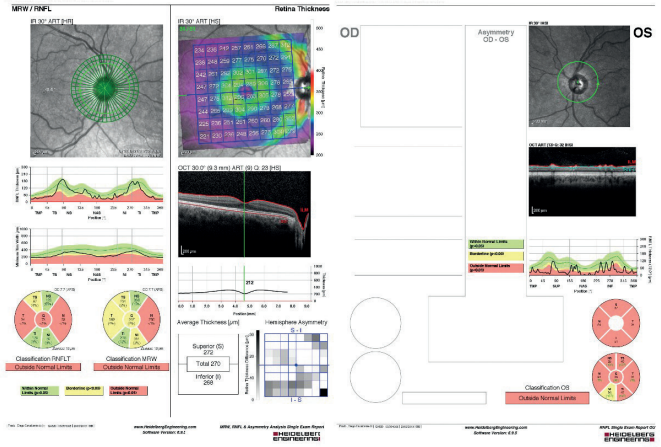

Figure 4. Optic nerve OCT: RE: loss of the nerve fiber layer in the nasal and temporal region, preserved lower fibers and borderline in the upper region, LE: diffuse loss of the nerve fiber layer.

Discussion

Macroadenomas present the following symptoms: visual field defects, cephalea, low visual acuity and progressive vision loss.[4] The mass effect of optical chiasma compression, local ischemia and/or microenvironmental changes due to neoplasia development are the mechanisms causing visual pathway dysfunction.[5]

If adenoma diameter is greater than 10mm (macroadenoma), it can cause neurophthalmological disorders by compressing the adjacent structures. Abnormalities in the fiber layer of the nerve in long-standing injuries are a characteristic feature of pituitary tumors. OCT is one of the tools used to detect such fiber loss in chiasma compression. A prospective study that evaluated the nerve fiber layer (NFL) and the ganglion cell complex (GCC) thickness using spectral domain optical coherence tomography (SD-OCT) concluded that such a damage could be observed even in the absence of compression and with preserved visual acuity. NFL and GCC thickness had 82% and 69% reduction, respectively. The study also showed diffuse loss, particularly in the nasal and temporal aspects of the optic nerve, of patients with long-standing visual field defects. Therefore, SD-OCT can play a key role in the early diagnosis and management of macroadenoma patients since it can early detect the loss of fibers and ganglion cells , even before compression, according to the aforementioned study.[6]

Most patients have the following conditions regarding visual field defect pattern in the most affected eye: temporal hemianopsia, followed by temporal quadrantanopsia with altitudinal defect and generalized depression.[1] Such conditions are explained by the location and crossing of nasal fibers, which carry images of the temporal field.[4] Hemianoptic lesions can be congruent or incongruous. One eye is usually more affected than the other in incongruous defects, as shown in the reported case.[7] Contrast MRI currently allows the early detection of precise anatomical location, which is of great importance for diagnosis and treatment.[8]

The degree of visual impairment is directly related to the quantitative size of the macroadenoma, as well as to the gap between diagnosis and appropriate treatment.[9] Compressive optic neuropathy can be clinically similar to glaucomatous optic neuropathy.[10] Visual field defects in glaucoma usually have arcuate pattern, however intrasellar or parasellar tumors can also cross the meridian.[11] Papilla pallor may have been a confounding factor with increased excavation leading to diagnosis of glaucoma in the current reported case.

The ophthalmologist can be the first doctor to exam a patient with clinical manifestations of intracranial tumors that can cause neurological and ocular complications. The possibility of intracranial tumors must be taken into account in the etiology of visual impairments before the definitive diagnosis is reached.

Lower incidence of pituitary macroadenoma and its similarities to other pathologies can result in confusion at the time of initial examination and lead to diagnosis delay. Visual disorders such as diplopia (by lateral expansion of the adenoma into the cavernous sinus), paralysis of the oculomotor nerves and, more rarely, nystagmus and proptosis, can be observed in pituitary macroadenoma, which contributes to initial diagnostic delay.[12] Optic neuritis is another example of diagnosis differential. It causes reduced vision, an outcome that can worsen in hours or days, is usually unilateral and causes visual field loss (it ranges from slight reduction to absence of light perception). Clinical history and ocular examination - associated with magnetic resonance - help elucidating the diagnosis.

The patient papillary pallor was mistaken for increased excavation at the initial moment. Such fact highlights the importance of adequate fundoscopic propaedeutic by taking into account the deflection of vessels and the non-staining of the papilla to quantify excavation associated with complete neurophthalmological assessment.

The assessed case shows the consequences of late pituitary macroadenoma diagnosis and treatment. It is necessary looking for methods that allow pituitary tumors diagnoses before nerve fiber loss in the retina, since it can leading to irreversible changes in the visual field.[5] In addition, basic propaedeutic with complete anamnesis, physical and complementary exams can directly interfere in prognosis and lead to the best hypothesis and help avoiding irreversible sequelae.

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