Orbital pseudotumor and pituitary lesion with cavernous plexus and internal carotid artery deviation: clinical, MRI and (18F)-FDG PET findings


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Abstract

We present a 43-year-old man with three-month medical history of headache, diplopia, right eye conjunctival chemosis and exophthalmos. Endocrinological investigations revealed high level of serum prolactin (s-PRL). Magnetic resonance imaging (MRI) of the orbits showed right proptosis, homogenous retrobulbar infiltrate with minimal enlargement of the superior rectus muscle. Brain images demonstrated pituitary mass 8/6 mm, extending into the left cavernous sinus. (18F)-FDG PET scans revealed normal cerebral glucose metabolism.

We suggest that this case may represent an orbital pseudotumor and pituitary microadenoma - prolactinoma. The differential diagnosis from other cases of atypical and rare disorders will be discussed: orbital pseudotumor and hypophysitis with hyperprolactinemia, associated with IgG4-related systemic disease, orbital and pituitary lymphoma.

Key words: orbital pseudotumor, pituitary lesion, MRI, (18F)-FDG PET, differential diagnosis

1. Introduction

The most common causes of unilateral proptosis/exophthalmos in adults are thyroid orbitopathy; orbital lymphoma; orbital inflammatory pseudotumor - IgG4-related disease, presented as myositis, dacryoadenitis, anterior, apical, and diffuse orbital process; granulomatosis, orbital tumor: metastases, gliomas/meningiomas, hemangioma; vascular/venous abnormalities disorders: carotid-cavernous fistula, cavernous sinus thrombosis, aneurism. (2, 5)

Differentiation of sellar masses which induce neurological, ophthalmological, endocrine damage still creates diagnostic challenges. Pituitary lesions involve a heterogeneous group of cystic lesions (craniopharyngioma, arachnoid cyst), neoplasms (adenoma, meningioma, gloma, metastases, lymphoma), inflammation/infections (sarcoïdosis, tuberculosis, pituitary abscess, lymphocytic hypophysitis, IgG4 hypophysitis), and empty sella syndrome (4, 7, 9, 11, 12).

A presumptive accurate diagnosis of orbital and pituitary pathology can be made on the basis of clinical course, laboratory findings, and neuroimaging features, but a definitive
diagnosis requires tissue biopsy (3). Evidence exists that magnetic resonance imaging (MRI) and positron emission tomography (PET) using 18F-FDG have been successfully applied in the diagnosis of neurological pathologies, including different vascular, neoplastic, inflammatory, autoimmune and degenerative central nervous system disorders (8, 10, 17). Respectively, we describe a case of right orbital proptosis and left pituitary lesion with cavernous plexus and internal carotid artery deviation, presenting with right eye exophthalmos, diplopia, headache, and hyperprolactinemia. An orbital pseudotumor and pituitary microadenoma/prolactinoma was suspected. A differential diagnosis for appropriate treatment was made.

2. Case report

A 43-year-old man complained of one year headache. Initial laboratory findings and magnetic resonance imaging (MRI) were normal. The patient presented in our neuro-ophthalmological section with three months history of right eye exophthalmos and double vision. Neuro-ophthalmological evaluation revealed right eye proptosis, conjunctival chemosis, diplopia, normal visual acuity and visual field analysis.

MRI of the orbits showed minimal enlargement of the right superior rectus muscle, proptosis, and homogenous enhancing retrobulbar mass (Fig. 1).

Figure 1

![Brain MRI](image1.jpg)

Brain MRI demonstrated pituitary mass 8/6 mm extending into the left cavernous plexus and left internal carotid artery deviation (Fig. 2).

Figure 2

![Brain MRI](image2.jpg)
(18F)-FDG PET revealed normal glucose metabolism of the pituitary gland, right orbital region and whole body (Fig. 3).

Figure 3

Physical (BMI 35 kg/m2), blood pressure 110/80 mmHg, pulse rate 60/min, and neurological examinations were normal. Level of serum prolactin (s-PRL) was significantly increased. A normal serum level of immunoglobulin G4 (IgG4) was observed. A routine blood test revealed normal parameters. Orbital and pituitary biopsy were not performed because the patient refused the examination. An orbital pseudotumor and pituitary microadenoma/prolactinoma were supposed. We initiated treatment with oral prednisone and cabergoline (Dostinex - 0.5 mg/twice weekly). Two months later, clinical improvement was observed – the exophthalmos and double vision decreased. Serum PRL was normalized. MRI revealed that the lesions persisted in the pituitary and the right orbit.

3. Discussion

The differential diagnosis of orbital proptosis and pituitary lesions with hyperprolactinemia always challenges the correct therapeutic behavior, when neurological, ophthalmological and endocrine symptoms occur (2). Approximately 20-40% of patients with biopsy-proven IgG4-related autoimmune disease have normal IgG4 concentration at the time of diagnosis (5, 10). Respectively, despite the normal IgG4 level in our patient, we can discuss the presence of inflammatory orbital pseudotumor and hypophysitis with hyperprolactinemia.
According to the literature, the involvement of ocular, orbital, and pituitary structures is estimated to occur in patients with various neoplasms, including lymphoma. Metastases have also been known to cause a similar type of orbital and pituitary mass. In regards to previous reports, the anatomical and functional neuroimaging techniques are widely used for the detection and differentiation of brain lesions. Although, in our case MRI revealed simultaneous right orbit and sellar region abnormalities, supposing primary or secondary neoplasms, (18F)-FDG PET showed negative results for the presence of any kind orbital and cerebral malignancies. In context of earlier published review articles (1, 15, 18) and aforementioned results, here we also discussed the application of PET in differentiating inflammation from tumor.

In conclusion, this case report supports the notion that the distinction between IgG4 related disease, pituitary adenoma/carcinoma, lymphoma or metastasis is very difficult without a biopsy. Based on the literature review and our own observations, we suggest that brain MRI and (18F)-FDG PET in addition to the clinical and laboratory findings may serve as a useful diagnostic tool in patients with simultaneous orbital and pituitary lesions.
References: