

Mucocoele and Graves' Orbitopathy as
the Causes of Proptosis in One Patient

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Abstract

Orbitopathy is an inflammatory disorder affecting orbital and per orbital tissue. It can manifest itself with proptosis. The main cause of proptosis in adults is Graves' Orbitopathy (GO). Recognition of GO does not exclude other causes of exophthalmos and differential diagnosis is sometimes required.

The case presents the patient with Graves Disease (GD), admitted to the Department of Endocrinology due to the symptoms of active orbitopathy. The patient had significant proptosis (right eye-30mm, left eye-29mm). Computed tomography showed thickening of oculomotor muscles in both eyes but also a mucous cyst localized in the right frontal sinus. The patient underwent endoscopic, intranasal removal of the lesion. Due to active, moderate to severe orbitopathy, he underwent treatment with 12 weekly pulses of intravenous methylprednisolone, followed by a 3-month prednisone therapy and orbital radiotherapy.

The case describes the patient with two coexisting pathologies. Each of them could manifest itself with proptosis. The main cause of proptosis is GO. Physicians must bear in mind that in case of atypical course of GO and lack of improvement, or worsening during corticosteroid treatment, differential diagnosis should be considered. It is worth emphasizing that one of the reasons of proptosis may be a mucocoele (mucous retention cyst).

Introduction

Orbitopathy is an inflammatory disorder affecting orbital and per orbital tissue. It can manifest itself with proptosis. The main cause of proptosis in adults is Graves' Orbitopathy (GO). However, recognition of GO does not exclude other causes of exophthalmos and sometimes differential diagnosis is needed. We should take into consideration further diagnosis in patients with: unilateral exophthalmos, atypical course of GO, lack of improvement or worsening during corticosteroid treatment. One of the reasons of exophthalmos may be a mucocoele (mucous retention cyst).

Case Presentation

A 55-year-old patient with hyperthyroidism due to Graves' Disease (GD), diagnosed in 2010, treated with antithyroid drug from December 2010, was admitted to the Department of Endocrinology in 2011 due to the symptoms of GO lasting from the beginning of 2011. Physical examination revealed signs of active inflammation assessed in the Clinical Activity Score (CAS) as 5/7 in Right Eye (RE) and in Left Eye (LE). The patient had significant proptosis (RE-30mm, LE-29mm) and lagophthalmos in both eyes (3mm). The patient did not have any clinical signs of Dysthyroid Optic Neuropathy (DON). On admission, we diagnosed active, moderate to severe GO according to European Group on Graves' orbitopathy [1]. We performed Computed Tomography (CT) of the orbits that showed thickening of oculomotor muscles and mucous cyst localized in the right frontal sinus of the size: 21x19x20mm (Figure 1). It was distending the wall of the sinus, penetrating to the upper-medial orbital wall. The cyst was modeling media loculomotor



Figure 1: Computed tomography image showed mucous cyst localized in the right frontal sinus of the size: 21x19x20mm, (white arrow).

muscle and reached nervous-vascular bunch, but without the signs of compression. The lateral wall of the sinus was significantly thin, partially invisible - the possibility of its destruction was described (Figure 2).

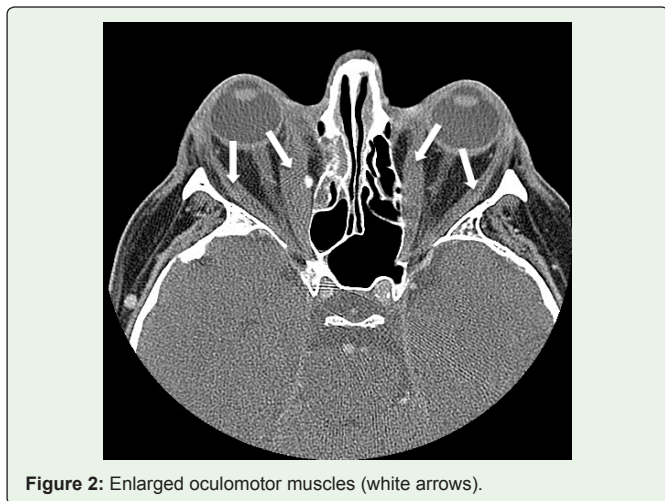


Figure 2: Enlarged oculomotor muscles (white arrows).

The final diagnosis was active, moderate to severe GO and mucocoele. The patient was qualified for endoscopic, intranasal removal of the lesion. The excision was incomplete what was shown in magnetic resonance performed after surgery (Figure 3).

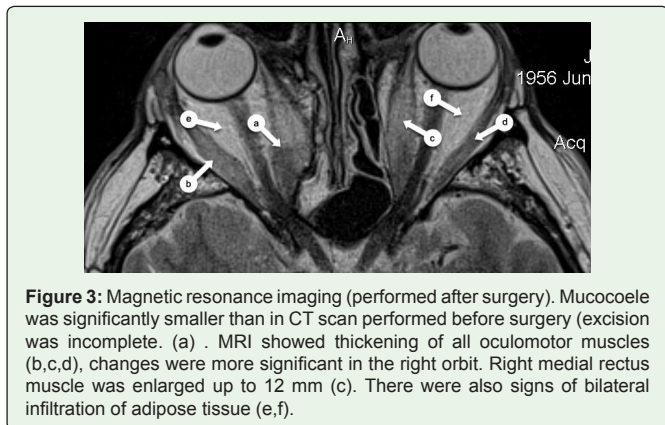


Figure 3: Magnetic resonance imaging (performed after surgery). Mucocoele was significantly smaller than in CT scan performed before surgery (excision was incomplete. (a) . MRI showed thickening of all oculomotor muscles (b,c,d), changes were more significant in the right orbit. Right medial rectus muscle was enlarged up to 12 mm (c). There were also signs of bilateral infiltration of adipose tissue (e,f).

Due to active, moderate to severe GO, the patient underwent a treatment with weekly pulses of intravenous methylprednisolone (ivMP) (6x500mg, 6x250mg), followed by a 3-month prednisone therapy and orbital radiotherapy.

Unfortunately, in the beginning of 2013, the patient noticed a deterioration of visual acuity and colour vision. On ophthalmologist examination, visual acuity was assessed as 0.9 and 1.0 for RE and LE respectively. The patient did not recognize all Ishihara plates with both eyes. The diagnosis of bilateral DON was established. The patient received ivMP pulses (3x1000mg) on three consecutive days. Due to lack of improvement the patient was qualified for endoscopic bilateral intranasal orbital decompression. As a continuation of implemented treatment the patient received another course of 12 ivMP pulses followed by a 3-month prednisone therapy. In the final assessment, we

did not find any clinical signs of DON. On physical examination there are, however, still signs of active inflammation- assessed in CAS scale as 3/7 in both eyes. Lack of satisfactory treatment effects is probably caused by smoking cigarettes throughout the whole treatment period.

In May 2013, due to a three-year period of antithyroid drug treatment, complete normalization of serum TSH receptor antibodies and stabilization of euthyroidism, the treatment was discontinued. The patient remains in clinical and hormonal euthyroidism.

Discussion

There are many diseases that can manifest themselves with proptosis. Most of them are listed in the Table 1 [2].

The differential diagnosis of proptosis is broad and includes a wide range of mass lesions that derive from orbits, the cranium, sinuses, and paranasal space. The main cause of proptosis is GO. Physicians must be aware of the possibility of coexistence of several pathologies.

The patient described above had two coexisting diseases, and each of them could manifest with proptosis. In case of atypical course of GO or lack of improvement or worsening during corticosteroid treatment it is necessary to verify the diagnosis. It is worth remembering that one of the reasons of exophthalmos may be a mucocoele.

Table 1: The differential diagnosis of proptosis.

Infections	<ul style="list-style-type: none"> Orbital and preseptal cellulitis. Bacterial, fungal, parasite infections Idiopathic orbital inflammatory disease
Vascular malformations	<ul style="list-style-type: none"> Varices, lymphangiomas, capillary haemangioma, cavernous haemangioma, arteriovenous malformations, carotid-cavernous fistula
Cystic lesions	<ul style="list-style-type: none"> Frontal or ethmoidal mucocoeles
Tumours	<ul style="list-style-type: none"> Pleomorphic lacrimal gland adenoma Lacrimal gland carcinoma Optic nerve glioma Optic nerve sheath meningioma Neurofibroma Lymphomas Rhabdomyosarcoma Childhood metastatic tumours Adult metastatic tumours
Other causes	<ul style="list-style-type: none"> Trauma with orbital or facial fractures or with retrobulbar haemorrhages which, if large enough, can push the globe forward Iatrogenic, post-surgical Sarcoidosis Cushing's disease Liver cirrhosis Wegener's granulomatosis Histiocytosis

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