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Case Report
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Graves' Ophthalmopathy Associated to Idiopathic Orbital Inflammatory Pseudotumor

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Abstract

Introduction: Association of orbital inflammatory pseudotumor and Graves' ophthalmopathy remains exceptional and unusual with only a few sporadic cases reported in the world literature. This situation represents a real challenge for health practitioners. We report the original case of idiopathic orbital inflammatory pseudotumor associated with Graves' ophthalmopathy in elderly patient.

Case Presentation: A 62-year-old woman with a history of right idiopathic dacryoadenitis and good outcome under systemic glucosteroids was explored two years later for acute and painful bilateral exophthalmia. Somatic examination noted asymmetrical, non-reducible, and painful bilateral exophthalmos with bilateral upper lid retraction. These signs were more prominent in the left eye. There was also a homogeneous and painless goiter.

Biology revealed hyperthyroidism: fT4 at 25.4 pmol/l and TSH at 0.01 µIU/ml. Cervical ultrasound demonstrated moderate, homogeneous, micro nodular, and hyper vascularized goiter. The thyroid immunity demonstrated positive anti-rTSH auto antibodies at 2.5 IU/ml. The diagnosis of Graves' disease complicated by thyroid-associated ophthalmopathy was retained. The patient was treated with methimazole (30 mg/day) and prednisone (1 mg/kg/day) with a favorable outcome.

Conclusion: As rare as it is, this unusual association deserves to be known by all health practitioners to avoid diagnostic delays and improve the prognosis that may be reserved, especially for Graves' ophthalmopathy.

Keywords: Graves' ophthalmopathy; Idiopathic orbital inflammatory pseudotumor; Thyroid-associated ophthalmopathy; Graves' disease; Orbital pseudotumor; Dacryoadenitis

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Introduction

Initially described by Birch-Hirschfeld in 1905 [1], non-specific inflammatory pseudotumor of the orbit remains an exceptional and enigmatic pathology, which represents a real diagnostic and therapeutic challenge for clinicians [2,3]. So called orbital pseudotumor, idiopathic orbital inflammation syndrome, nonspecific orbital inflammation, and orbital inflammatory syndrome, the name of "non-specific inflammatory pseudotumor" was adapted in 1954 by Umiker and Iverson [4] to differentiate these tumors from other neoplastic processes and insist on their benign inflammatory nature [4].

Inflammatory pseudotumors of the orbit are included in the group of extra pulmonary inflammatory pseudotumors, which account for less than 5% of all inflammatory pseudotumors [5]. These inflammatory pseudotumors account for approximately 10% of all orbital mass lesions and are particularly common in children and young adults [6]. The forms of the elderly are rare [5,6].

The association in the same patient of an orbital inflammatory pseudotumor and a Graves' ophthalmopathy remains exceptional and unusual situation with only a few sporadic cases reported in the world literature [7,8]. This situation represents a real challenge for health practitioners since the differential diagnosis between the two entities is not easy to make [8,9]. We report the original case of idiopathic orbital inflammatory pseudotumor associated with Graves' ophthalmopathy in elderly patient.

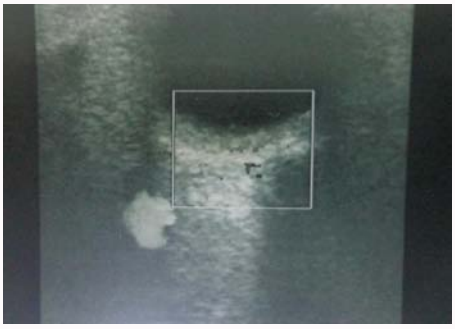

Figure 1: Orbital ultrasound: Dacryoadenitis with upper and lower palpebral oedematous thickening.

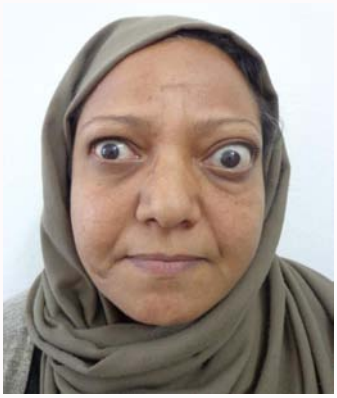

Figure 2: Asymmetrical exophthalmia more marked on the left eye.

Case Presentation

A 62-year-old woman with no significant pathological history was referred to our department for exploration of right insidious ocular pain gradually worsening for two months. The somatic examination was without abnormalities except for a discreet right exophthalmia. Orbital ultrasound revealed upper and lower palpebral oedematous infiltration and homogeneous swelling of the right lacrimal gland (Figure 1). Baseline biology was within normal limits: total blood count, erythrocyte sedimentation rate, C-reactive protein, serum calcium, ionogram, creatinine, transaminases, muscle enzymes, lipid parameters, blood glucose, and serum protein electrophoresis. Thyroid tests did not show abnormalities: total thyroxine (fT4) at 9.2 pmol/l, Thyroid Stimulating Hormone (TSH) at 1.08 µUI/ml, negative anti-thyroglobulin antibody (anti-Tg) at 48 IU/ml (<120), negative anti-thyroperoxidase antibody (anti-TPO) at 30 IU/ml (<120), and negative anti-TSH receptor antibodies (anti-rTSH) at 0.3 IU/ml (<1).

Histopathological examination of the biopsy of the right lacrimal gland resulted in dacryoadenitis. Subsequent investigations had eliminated an underlying systemic disease (connective tissue disease, vasculitis, granulomatosis, hemopathies, and cancers). The diagnosis of idiopathic inflammatory pseudotumor of the orbit/dacryoadenitis type was retained. The patient was treated with systemic glucosteroids with a favorable outcome.

Two years later, the patient returns to our consultation because of acute and painful bilateral exophthalmia associated with a decrease in visual acuity.

Somatic examination noted asymmetrical, non-reducible, and painful bilateral exophthalmos with bilateral upper lid retraction. These signs were more prominent in the left eye (Figures 2-4). There was also a homogeneous and painless goiter.

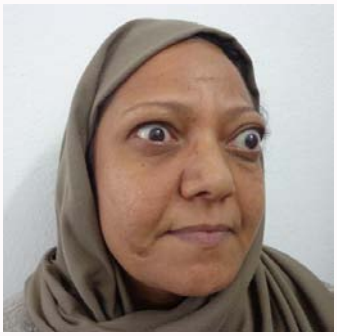

Figure 3: Bilateral upper lid retraction.

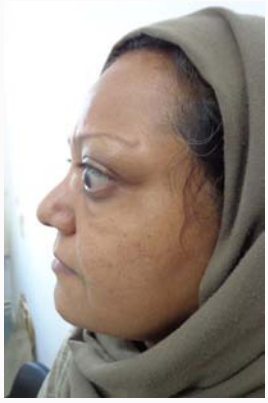

Figure 4: Left Exophthalmos in side view.

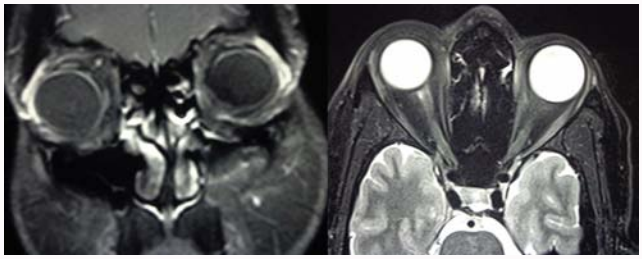

Figure 5: Normal orbital MRI at six months of treatment (coronal and axial views).

Biology revealed hyperthyroidism: fT4 at 25.4 pmol/l and TSH at 0.01 µIU/ml. Cervical ultrasound demonstrated moderate, homogeneous, micro nodular, and hyper vascularized goiter. The thyroid immunity demonstrated positive anti-rTSH auto antibodies at 2.5 IU/ml. Anti-Tg and anti-TPO antibodies were always negative (15 IU/ml and 35 IU/ml). The diagnosis of Graves' disease complicated by thyroid-associated ophthalmopathy was retained. The patient was treated with methimazole (30 mg/day) and prednisone (1 mg/kg/day) with a favorable outcome: Fast disappearance of functional complaints, normalization of thyroid tests at two months, and normal appearance of orbital Magnetic Resonance Imaging (MRI) at six months (Figure 5).

Discussion

The association of idiopathic orbital inflammatory pseudotumor with a thyroid-associated ophthalmopathy is exceptional and unusual [7,8]. In fact, Yan et al. [10] in their large series of 209 cases of orbital inflammatory pseudotumors, found no association with a thyroid eye disease; and the systematic review of the literature made in 2015 by Shen et al. [8] found only 7 cases of this association.

This association was described with different types of idiopathic orbital inflammatory syndromes: myositis, sclerosing orbital inflammation, IgG4-related lymphoplasmacytic infiltrative disorder, dacryoadenitis, and lymphatic inflammatory pseudotumor [7,8,11-14].

The chronology of these two diseases in the same patient is variable; most often they occur specially in time [11,14]. More rarely, a concomitant diagnosis of the inflammatory pseudotumor of the orbit and Graves' ophthalmopathy has been reported [7,8,12]. The diagnosis of this association represents a real challenge for clinicians for several reasons:

- The inflammatory pseudotumor of the orbit can mimic a Graves 'ophthalmopathy, especially in the so-called "euthyroid graves' disease" forms [9,15]. The bilateral forms of the orbital inflammatory pseudotumors, observed in 25% of cases, can also pose the differential diagnosis with thyroid-associated ophthalmopathy [6],
- Graves' ophthalmopathy can sometimes present as unilateral ocular involvement and mimic an idiopathic orbital inflammatory pseudotumor [16].
- Thyroid-associated ophthalmopathy can cause enlarged lacrimal glands or true "thyroid eye disease-related autoimmune dacryoadenitis" as well as an orbital tissue enhancement secondary to inflammation, inducing confusion with authentic idiopathic orbital inflammatory pseudotumor [17,18].

These similarities in the clinical and radiological presentations of these two diseases explain the diagnostic difficulties, particularly in cases occurring simultaneously [7,9,17,18].

The exact pathophysiology of this association is not yet well understood [8]. A common dysimmunitary mechanism at the origin of both entities is often mentioned as a promoting factor [8,11,12]. This theory is comforted by the association with other autoimmune diseases in three of the four cases of Bijlsma and Kalmann [11]. In addition, a significant association of these two diseases with anti-55-kD auto antibodies protein of eye muscle membrane antigens in porcine models has been objectified [19]. These cases classically respond favorably to systemic corticosteroids [8].

Conclusion

As rare as it is, this unusual association deserves to be known by all health practitioners to avoid diagnostic delays and improve the prognosis that may be reserved, especially for Graves' ophthalmopathy.

Our observation is characterized by its occurrence in an "older adult" patient (over 60 years). At this age orbital space-occupying lesions are often malignant whereas inflammatory pseudotumors remain rare.

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