
SHORT COMMUNICATION

Unilateral exophthalmos associated with ipsilateral mucosal turbinate hypertrophy: Benign exophthalmos syndrome (BES). A description of a new clinical condition

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PURPOSE. *To report the clinical and radiologic characteristics of a group of patients who experienced unilateral exophthalmos associated with ipsilateral mucosal turbinate hypertrophy. The clinical features of these patients are presented and a hypothesis proposed to explain this condition for which the authors introduce the term benign exophthalmos syndrome (BES).*

METHODS. *Retrospective, noncomparative case series. Participants: Four patients experienced slow progressive unilateral exophthalmos associated with ipsilateral mucosal turbinate hypertrophy, with no evidence of orbital mass or extraocular muscles involvement. Intervention: Main Outcome Measures. Symptomatic outcome and measurement of the degree of relative exophthalmos.*

RESULTS. *The onset of exophthalmos was associated with clinical and radiologic features that resemble BES. In all patients, radiologic examination demonstrated an ipsilateral mucosal turbinate hypertrophy and not the presence of orbital disease or expanding lesions of paranasal sinus. After daily intranasal spray of steroid, in three of the four cases the globe returned to within 1 mm of exophthalmometry of the contralateral eye.*

CONCLUSIONS. *The relationship between the feature of paranasal sinus disease and the development of ipsilateral exophthalmos has been described in the literature. The four cases described herein appear peculiar for the slow progressive onset of the exophthalmos, without inflammatory and mass effect signs. This condition associated in all cases with ipsilateral hypertrophy of the nasal mucosa provides a guide to a hypothetical mechanism for BES. According to these hypothesis, the therapy should be devoted to the nasal disease more than the orbital. (Eur J Ophthalmol 2005; 15: 800-3)*

Key Words. *Exophthalmos, Turbinate hypertrophy*

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INTRODUCTION

Exophthalmos represents the main symptom of orbital diseases, and it usually reveals the presence of Graves' ophthalmopathy, or tumors and inflammation of the orbit. In all these conditions the proptosis is caused by an expanding lesion with some degree of involvement of the orbital structures. Polyps or tumoral affections of the paranasal sinus can also lead to oculo-orbital complica-

tions and to the development of exophthalmos. In these cases proptosis depends on a secondary infiltrating process arising from the paranasal sinuses.

Nevertheless, some cases with very slow progressive unilateral exophthalmos do not have evidence of orbital or paranasal disease and the cause remains uncertain. The careful review of these patients by means of computed tomography (CT) scan and magnetic resonance imaging (MRI) has revealed a paranasal mucosal hypertrophy on

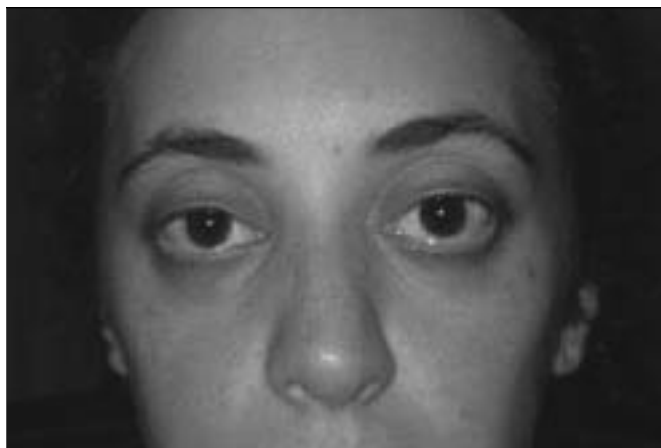


Fig. 1 - Bilateral exophthalmos in a 25-year old lady.



Fig. 2 - Left exophthalmos in a 35-year-old man

the same side as the exophthalmos. This feature could suggest a relationship between the onset of the proptosis and the presence of the ipsilateral exophthalmos. We present four patients with this condition, for which we introduce the term benign exophthalmos syndrome (BES), and describe the characteristic clinical and radiologic features.

MATERIALS AND METHODS

Patients were identified from a series of 4,000 cases with orbital diseases collected between 1980 and 2004 attending the Orbital Clinic of the Department of Ophthalmology of University of Naples Federico II. The presenting symptoms and signs were recorded and the natural history noted. The radiologic changes on computed tomographic scans of the orbit and sinuses were reviewed independently of clinical details by two radiologists.

Patient 1

A 25-year-old woman was referred to the Orbital Clinic, having noted a slow progressive bilateral exophthalmos more evident in her left eye approximately 11 years before the clinic visit. She was asymptomatic and had no past medical problems, injuries, or surgery. She had Snellen acuity of 20/20 in either eye, with no optic neuropathy and no intraocular disease on either side. There was 2 mm of left relative exophthalmos, with a very slight upper lid edema but no detectable limitation of ocular ductions. Thyroid bloodwork was negative for hyperthyroidism. CT and MRI showed a bilateral turbinate mucosa hypertrophy with no evidence of detectable orbital mass or inflamma-

tory disease. With the characteristic history and signs, and the absence of prior orbital and sinus disease, a diagnosis of BES was made. This was confirmed on MRI scan with fat suppression procedure. After a course of daily intranasal spray of steroid she improved in terms of proptosis with reduction of about 1 mm. After therapy, there remained 1 mm of left relative exophthalmos, and the patient was satisfied with the result. There has been no recurrence of the symptoms at 2 years after therapy.

Patient 2

A 35-year-old man had noted a progressive exophthalmos of the left eye but both his wife and friends thought his appearance was symmetric. Over the next 6 months, there was progression of the condition, with a worsening of the proptosis and development of an upper scleral show of 1 mm, such that friends were passing spontaneous comments and his wife was encouraging medical investigation. Thyroid bloodwork was normal and he was in good general health. Hertel examination demonstrated a 17 mm exophthalmos in the left eye (versus 15 mm of the right eye). There were no other ocular symptoms. The CT scan revealed an ipsilateral turbinate hypertrophy. The patient underwent a therapy by means of daily intranasal spray of steroid.

Patient 3

A 44-year-old man was aware of a left exophthalmos for several months owing to bumping of his eyelashes against his eyeglasses. The thyroid screening was normal. A CT scan showed ipsilateral turbinate hypertrophy. Clinical signs and radiologic investigations were typical for

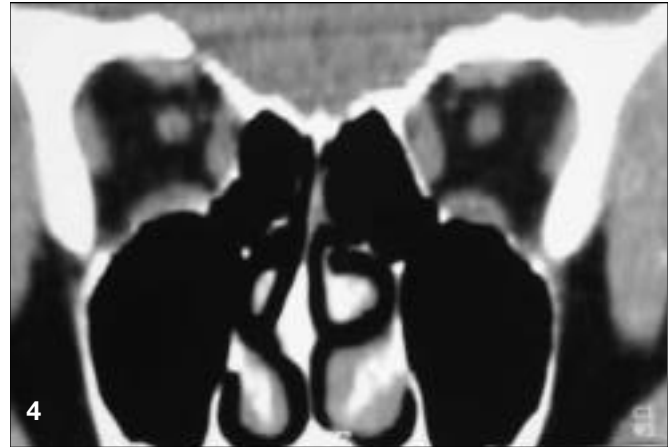


Figure 3, 4 - CT scan showing a ipsilateral mucosal turbinate hypertrophy.

BES, and topical treatment with steroid was administered with a slight improvement of the symptoms.

Patient 4

A 22-year-old woman was made aware of a left exophthalmos by her parents and her ophthalmologist, who sent her to our center. The clinical evaluation revealed a 20 mm left exophthalmos and a scleral show of 1 mm. At the endocrinologic consultation, there was no evidence of thyroid disease. A CT scan showed ipsilateral turbinate hypertrophy. Topical nasal steroid therapy was administered that did not lead to improvement of the proptosis.

RESULTS

All the patients had a similar clinical history in terms of onset, progression, and concomitant symptoms except for one patient (no. 1) who experienced a subacute development of proptosis with slight signs of inflammation. Radiologic examination demonstrated the association in each patient of the proptosis and turbinate hypertrophy. Therapy based on prescription of a middle course of topical nasal steroid in three of four cases resulted in a reduction of about 1 mm of proptosis, stable for 6 to 8 months.

DISCUSSION

A relation between inflammatory disorders of the nose and paranasal sinuses with inflammation of the orbital contents has been proven. Duke-Elder (1) discussed this

condition, although he focused the discussion on cases with acute inflammation of the orbit associated with classical inflammatory signs, such as eyelid edema, conjunctival chemosis, extraocular impairment, and proptosis due to sinusial diseases. Some authors (2) have described the incidence of sinusitis in patients with idiopathic orbital inflammatory pseudotumor (IOIP) and the relationship between sinusitis and IOIP, concluding that sinusitis may be a cause in the etiology of IOIP. In a combined review (3) of ophthalmologic and otorhinolaryngoiatric clinical series, the relationship between oculo-orbital affections and sinusial lesions was established and the authors concluded that patients with inflammatory or tumoral affections of the paranasal sinus with oculo-orbital complications refer to an ophthalmologist as first consultation.

All the mentioned articles discuss the acute presentation (4) of exophthalmos with inflammatory signs or subacute dramatic progression of the proptosis due to an existing tumor. Nevertheless, some cases may be clinically very puzzling: proptosis appears without other symptoms and probably subsides with no or indecisive treatment. Frequently no evident cause can be found and management of these patients could become very frustrating. This prompts us in reviewing our large clinical series of cases with orbital diseases to identify this kind of patients. At the beginning the number of cases found was more than the four presented here, but we excluded cases with a long-standing exophthalmos probably present from birth and both radiologically and clinically due to a slight facial asymmetry. Other patients were otherwise excluded for the presence of monolateral myopia even if not very high in grade.

The common clinical feature of the patients presented

in this article is the slow progression of the exophthalmos: two patients (Cases 1 and 2) were worried about monolateral proptosis for a long period (Figs. 1 and 2). Another patient (Case 3) was aware of a left exophthalmos since he noted bumping of his eyelashes against his eyeglasses several months before the visit.

The last patient (Case 4) was made aware of a right exophthalmos by her parents. No one had pain or evidence of other clinically relevant features. Therefore we can state that in patients with BES the clinical onset is subtle without signs of orbital inflammation. Only in Case 1 a moderate lid edema was found.

As we reported in Results, all the radiologic examinations were negative for the presence of expanding lesion of the orbit and paranasal sinus as well as for signs of orbital inflammation. In all the patients, ipsilateral hypertrophy of the turbinate was clearly detectable (Figs. 3 and 4). The mucosa of the nose appeared thickened and congested at the same side where the exophthalmos developed. Nevertheless, these patients did not complain of nasal disturbance. In their anamnesis there was no history of recurrent cold or sore throat or of allergic episodes. Furthermore, we have a minimum of 2 years of follow-up of these patients and no one showed evidence of hyperthyroidism or alteration of the rheumatoid index during the periodic bloodwork.

Rhinologic examination to rule out the presence of

orbital edema excited by an infected paranasal sinus was performed.

Because all the clinical parameters examined in terms of orbital and sinus diseases considered were negative, and considering the isolated presence of ipsilateral hypertrophy of the turbinate mucosa, the possible pathogenesis of BES could be a decrease of the venous outflow from the orbit to the nose that leads to an edematous congestion of the orbital tissue.

The incomplete reduction of the proptosis that three patients had with use of intranasal steroid spray and the lack of response of the other patients support the hypothesis of the chronic "congestion mechanism" more than the inflammatory mechanism, although further proof is necessary.

In the presence of a slight and slow progressive exophthalmos in absence of orbital tumor or Graves ophthalmopathy or other expanding lesions, the ophthalmologist should be driven to evaluate a paranasal sinus condition.

The authors have no proprietary interest in any aspect of the article.

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