Case Report
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Orbital myositis secondary to eosinophilic granulomatosis with polyangiitis, about a case

Abstract

Objective: After dysthyroid orbitopathy, idiopathic orbital myositis is the second most common cause of inflammatory disease of the extraocular muscles. Although the typical presentation of this entity occurs suddenly, painfully, in young women, associated with diplopia, there are chronic, recurrent cases, without response to corticosteroid treatment, in which systemic inflammatory, autoimmune and associated infectious causes must be considered.[1]

Clinical case: An 18-year-old female patient referred to the neurology department attended the consultation, where she attended for the first time, due to diplopia associated with bilateral exophthalmos with restriction of extraocular movements, ocular hypertension and ptosis of the right eye. After a complete ophthalmological examination and exhaustive follow-up with pertinent complementary studies, ruling out thyroid pathology with negative tests and an MRI with diffuse inflammation of bilateral MEO. Immunosuppressive treatment is indicated with lack of response and constant relapses. A wash out of systemic medication is performed, with the aim of performing a diagnostic biopsy, yielding as a result an eosinophilic infiltrate compatible with eosinophilic granulomatosis with polyangiitis.

Conclusion: Eosinophilic granulomatosis with ophthalmic polyangiitis is a rare condition, with little descriptive literature on the subject and, therefore, difficult to suspect, with a prevalence of 0.5 to 3.7 per million people.[2] Both this and other secondary causes should be considered in cases of lack of response to immunosuppressive treatment and relapses in patients with inflammatory pathology of EOM in whom laboratory and complementary studies are negative, and the importance of biopsy for diagnostic confirmation and establishment of adequate treatment, as happened in our clinical case.

Keywords: myositis, orbit, eosinophilic granulomatosis, polyangiitis

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Introduction

Orbital myositis is a relatively rare disease, but it is the next most common cause of extraocular muscle disease after thyroid-associated orbitopathy.[3,4] It typically presents as an acute, idiopathic, painful diplopia in young women, with inflammation of a single extraocular muscle, and usually responds to a oral corticosteroid course. However, there are many outliers. These include chronic and recurrent idiopathic orbital myositis, unresponsive to corticosteroids, and cases occurring in the setting of systemic inflammatory, autoimmune, and infectious conditions, and in response to drugs.[1] Within the autoimmune inflammatory conditions are idiopathic orbital myositis, that associated with IgG4, inflammatory bowel disease (Crohn's/ Ulcerative Colitis), sarcoidosis, SLE and LED, giant cell myocarditis, post-streptococcal pharyngitis, Rheumatoid Arthritis, Behçet's disease, Psoriatic arthritis/psoriasis and, as in our case, eosinophilic granulomatosis with polyangiitis (Churg-Strauss Syndrome).[1]

Eosinophilic granulomatosis with polyangiitis (EGPA) was first described in 1951 by Churg and Strauss as a rare disease characterized by disseminated necrotizing vasculitis with extravascular granulomas.[5] Epidemiological data are limited due to its rarity, but reflect an annual prevalence of 0.5 to 3.7 per million. The presentation occurs, in general, in adults, and very rarely in adolescents, in which cases its involvement predominates in women.[6] In a systematic review found on the ophthalmic presentation of EGPA, of 46 people reviewed, 25 of them presented an ischemic vasculitic form and 20 an idiopathic orbital inflammatory form, 1 person presented a mixed form.[2]

Although EGPA has classically been described as a disease that goes through 3 stages (asthmatic and allergic prodromal, an eosinophilic phase with peripheral eosinophilic infiltration to multiple organs, and the final vasculitic phase, 8 to 10 years later, with production of extravascular granulomas[7] the idiopathic orbital inflammatory type typically presents an average of 6.9 years after the initial diagnosis of asthma and its manifestations may be conjunctival nodules, orbital myositis[8–11] orbital inflammatory syndrome[12–14] dacryoadenitis, and cranial nerve palsy. Cases of marginal keratitis, episcleritis, and orbital apex syndrome have also been described. In contrast, presentations of ischemic vasculitis include retinal artery and vein occlusions, ischemic optic neuropathy, and retinal vasculitis or edema. The differential diagnosis of EGPA in the orbit and eye includes hypereosinophilic syndrome, granulomatosis with polyangiitis (Wegener's), microscopic polyangiitis, and parasitic infection. Although for EGPA the history of asthma, rhinitis and ANCA-P is used for diagnosis, only 30-40% are positive, and as in the case report by Takanashi T,[8] there may be a late presentation of asthma and nasal polyposis (30 years of age).[15]

Clinical case

An 18-year-old female patient who attended the clinic due to an acute picture of binocular diplopia with exophthalmos, restricted eye movements and ocular hypertension in both eyes with ptosis of both eyes although more marked in the right eye (Figure 2). At the time of consultation, he did not report any systemic clinical history of interest. A complete ophthalmological examination is performed, in which the aforementioned alterations associated with uncorrected visual

acuity 20/20 are evident, both eyes, with normal color test, preserved ocular reflexes. Biomicroscopy showed no particularities, however intraocular pressure measurement was 26 mmHg in the right eye and 25 mmHg in the left eye, with a normal fundus in both eyes.

A 24.2 computerized visual field was performed without altered sensitivity in both eyes (Figure 1). An optical coherence tomography of the nerve fiber layer was also performed, which showed a borderline alteration of the upper NFL of the right eye (Figure 2), and a nuclear magnetic resonance where a diffuse thickening of the muscles can be observed. extraocular of both eyes without affectation of the tendon insertions (Figure 3). A basic laboratory was performed (complete blood count, coagulogram, glycemia, urea, creatinine, ionogram, erythrocyte sedimentation rate, PCR) to which was added thyroid profile with TRAB and ATPO antibodies, electrophoretic proteinogram, total antibodies, complete rheumatological profile, IgG4, serologies, radiography. chest and PPD with negative results. Having also ruled out possible myasthenia gravis. The only positive result was an IgE in values that oscillated in each control at 1600 U/ml, remaining high in each laboratory control (Figure 4 & 5).

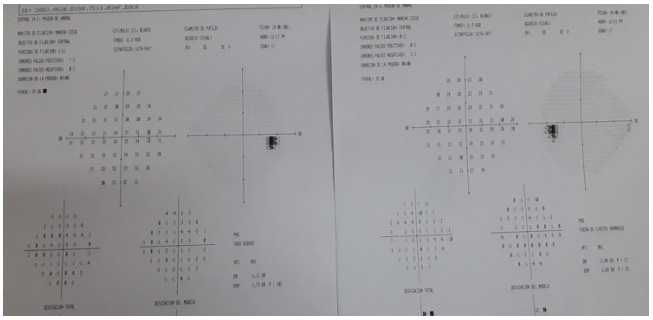

Figure 1 Initial CVC 24.2, without visual field compromise, ruling out optic nerve compression.

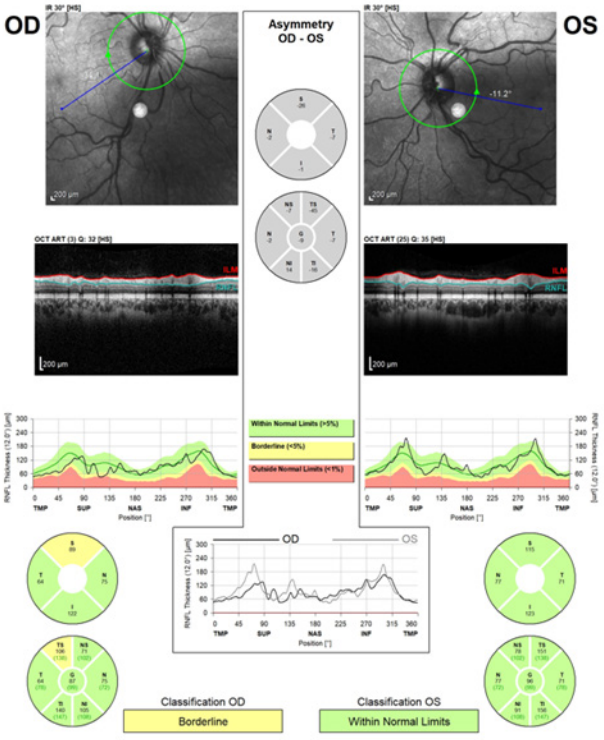

Figure 2 OCT of the nerve fiber layer with a borderline decrease in the upper RFNL of RE.

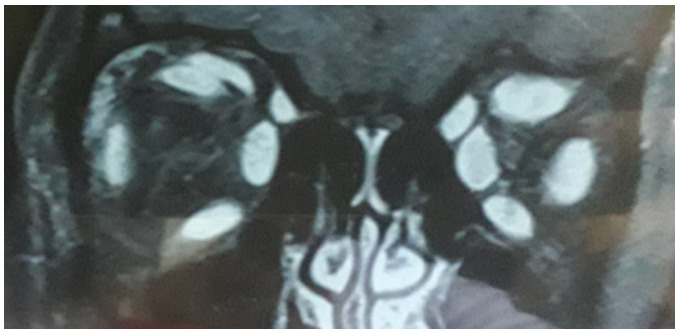

Figure 3 MRI without contrast: Diffuse thickening of the muscular bodies of the recti of both eyes is observed, including the levator palpebrae superioris muscle of the RE, causing the clinically evidenced ptosis.

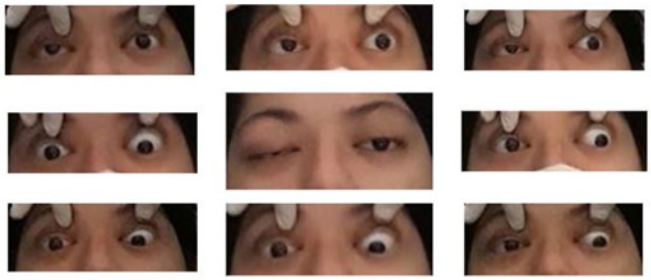

Figure 4 Note the alteration of ocular motility in gaze positions and the ptosis of the right eye.

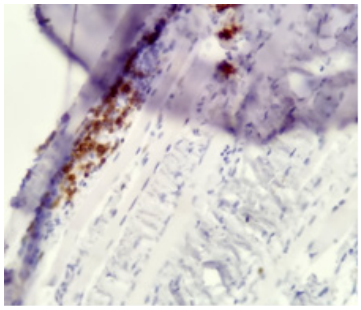

Figure 5 Pathological anatomy obtained from a biopsy of the medial rectus muscle of the right eye.

Immuno suppressive treatment with oral corticosteroids was indicated, initially responding with a decrease in symptoms. The problem began with the decrease in corticosteroids that favored reactivation. Therefore, treatment with azathioprine at maximum dose was started. After the stabilization of the symptoms 8 months later, tricytopenia appeared as an adverse effect of said immunosuppressant, for which it was decided to suspend it, which caused a new reactivation of the symptoms. Therefore, a biopsy of the medial rectus muscle of the right eye was performed. The anatomopathological result is received, showing, in addition to a slight lymphocytic and macrophage infiltrate, an eosinophilic perivascular infiltrate. After ruling out parasitic infectious causes and reviewing the literature, we observed that although criteria such as asthma and allergies are used for the diagnosis of eosinophilic granulomatosis with polyangiitis, there are case reports of manifestations and diagnosis of asthma, allergies, and nasal polyposis in adulthood. . Therefore, we could be faced with an idiopathic orbital inflammatory form of eosinophilic granulomatosis with polyangiitis as the first manifestation of a future systemic disease.

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Discussion

Little is known about the ocular manifestations of eosinophilic granulomatosis with polyangiitis, since there are very few cases described due to the low frequency of this entity. In the case of our study patient, with no prior systemic history of interest, she debuted with recurrent bilateral orbital myositis with each decrease in corticosteroids and with a lack of response to immunosuppressive treatment, whose only positive data was a constantly elevated IgE and a biopsy in which finally shows an eosinophilic infiltration. Ruled out infectious causes (Herpes zoster ophthalmicus, Lyme disease, Whipple disease, Varicella zoster (chicken pox), Coxsackie virus, Cysticercosis, trichinosis), medication (ipilimumab, alemtuzumab, bisphosphonates, statins, interferon, rivabirine and influenza vaccine), and other autoimmune causes (IgG4-related disease, Inflammatory bowel disease, Sarcoidosis, systemic lupus erythematosus, Giant-cell myocarditis, Poststreptococcal pharyngitis, Rheumatoid arthritis, Behçet disease, GPA, Psoriatic arthritis/psoriasis), as well as hypereosinophilia syndrome, it is very likely that we are before a case of eosinophilic granulomatosis with polyangiitis (EGPA) in its orbital inflammatory form, and that this is the first manifestation of this chronic systemic inflammatory disease, since, as observed in According to the report by Takanashi T, the patient began to present pulmonary symptoms and otorhinolaryngology from the age of 30.

Conclusion

In conclusion, we must bear in mind the secondary causes of orbital inflammatory disease in a patient with negative laboratory tests for dysthyroid orbitopathy, with a lack of response to immunosuppressive treatment, as well as perform a biopsy for diagnosis. Also, keep in mind that we are dealing with a very rare case of presentation in an adolescent who has not yet manifested systemic symptoms of a disease diagnosed through biopsy. Therefore, it is important to carry out long-term follow-up, paying special attention to the development of asthmatic/allergic pulmonary manifestations, and otorhinolaryngological manifestations such as the development of nasal polyps and to carry out the appropriate treatment to avoid systemic inflammatory relapses and those of the extraocular muscles.

Acknowledgments

None.

Conflicts of interest

The authors declares that there are no conflicts of interest.

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