Posterior Scleritis and Myelodisplasia in Relapsing Polychondritis: Case Report and Literature Review

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Abstract

Relapsing Polychondritis (RP) is a rare, recurrent and autoimmune multisystem disorder affecting cartilaginous structures, such as auricles, joints, nasal septum, larynx and tracheobronchial tree. Ocular manifestations may be observed in 42,3-65% of cases. Episcleritis and scleritis are the most common findings. However, RP is frequently misdiagnosed, leading to potentially severe, debilitating and, sometimes, fatal disease.

There is no established standardized therapeutic protocol for RP. Current medical therapy is largely empiric and based on case reports.

The aim of this paper is to document one case of ocular involvement in RP disease, emphasizing clinical and imaging findings that can help to establish an early diagnosis.

Keywords: Relapsing polychondritis; autoimmune disease; myelodysplastic syndrome; scleritis/treatment.

Introduction

RP is an autoimmune disease affecting cartilaginous and proteoglycan-rich structures (eyes, heart, inner ear, kidneys and the vessels). It may be associated with significant morbidity and mortality, caused especially by infection secondary to corticosteroid treatment, respiratory involvement, systemic vasculitis and renal disease. Presentation of anemia is also associated with poor prognosis in RP patients of all ages. Ocular manifestations may be observed in 42,3-65% of cases.1,2

RP affects 3,5 to 4 people per million, predominantly in the sixth decade of life and presents a male: female ratio between 1:1 and 1:3.3 It was first described in 1923 by Jaksh-Wartenhorst as polychondropathia in a patient with an 18-month course of progressive degeneration of multiple peripheral joints, nasal septum, external ears, external auditory canals, inner ears and epiglottitis. In 1960, the term “Relapsing Polychondritis” was introduced by McPearson.

The etiology of this disease is unknown, but an autoimmune mechanism is suspected. This is supported by the presence of lymphocytic and neutrophilic infiltration of the affected tissues, with humoral and cellular responses against collagen type II and other collagen antigens, the high concomitant prevalence of other autoimmune diseases and response to immunosuppressive agents.

There is no specific laboratory test diagnostic for RP, except increased levels of acute phase reactants, which indicate the presence of systemic inflammation.

McAdam et al had suggested clinical criteria for the diagnosis of RP, which included bilateral auricles chondritis, non-erosive inflammatory polyarthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis, cochlear or vestibular damage.4 However, the symptoms and signs rarely appear simultaneously, which difficult and delays the correct diagnosis.

Case Report

A 75-year-old man with a history of prostatic adenocarcinoma and dyslipidemia was referred to the Department of Ophthalmology with severe headache, diminished visual acuity, pain and inflammatory signs in the external ear (Figure 1) with 5 days of evolution. Patient had also recent history of right posterior scleritis (PS) of unknown etiology (Figure 2), which regressed with oral corticotherapy.

He presented with best-corrected visual acuity of 20/400 in both eyes, bilateral conjunctival hyperemia, posterior and superior synechiae and 1+ flare in the anterior chamber of the right eye. Both eyes revealed extensive inferior chorioretinal detachment, confirmed by ocular ultrasound B mode. The patient is being followed by other specialists (rheumatologists and otorhinolaringologists) in reason of a recent migratory, non-erosive and seronegative polyarthritis, affecting lower limbs and knees, plus a cochlear dysfunction.

Macular SD-OCT of right eye revealed cystoid macular edema with serous retinal detachment, extending to peripapillary zone, chorioretinal folds and a foveal thickness of 720μm (Figure 3-panel a,b). Macular SD-OCT of the left eye showed also a macular epiretinal membrane, cystoid macular edema with foveal thickness of 370μm (Figure 3-panel c).

This second inflammatory episode was associated with a decreasing hemoglobin (84 g/L) and increasing erythrocyte sedimentation rate (ESR) (>120 mm), high reactive protein C of 13,7mg/dL and an elevation of β-globulin fraction of the serum proteins. Urine Bence Jones proteins were negative and myelogram revealed a myelodysplasia. Other serological and autoimmune laboratory exams were negative.

Keywords: Relapsing polychondritis; autoimmune disease; myelodysplastic syndrome; scleritis/treatment.

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Retinal fluorescence angiography showed bilateral optic disc extravasation hyperfluorescence, pooling effect in posterior pole and peripheral retina due to hyperfluorescence, pooling effect in both eyes due to pooling effect (white arrow) and optic disk extravasation hyperfluorescence.

Figure 2: Orbital MRI of patient. (panel a) axial high resolution T1-weighted MRI and (panel b) high resolution FIESTA axial images of the orbit revealing choroidal detachment. (panel c) High resolution FIESTA axial image showing scleral thickening of the right eye.

Retinal fluorescnic angiography shows hyperfluorescence in both eyes due to pooling effect (white arrow) and optic disk extravasation hyperfluorescence.

Oral Prednisolone 60mg/day and topical dexamethasone were reintroduced with a dramatic improvement of ocular hyperemia and pain, headache and regression of all signs of ear inflammation after 48 hours.

After 4 months of corticotherapy, the patient presented with no pain and best-corrected visual acuity of 20/100 and 20/400 in right and left eye, respectively. At corrected visual acuity of 20/100 and 20/400 in right and left eye, respectively. At

Discussion

Posterior scleritis is one of the most misdiagnosed conditions in ophthalmology due to its very low prevalence and, sometimes, absence of external ocular signs. It is also a potential blinding condition, associated with autoimmune disorders (such as RP) in 40% of the cases.

Ocular inflammation is present up to 65% of RP cases. However, the eye is involved in only 20% of the cases at the time of initial presentation. In our case, PS was the first manifestation of RP. Our patient revealed history of previous PS with no other symptoms and no relevant laboratory findings, except for anemia and high ESR. Patient was treated with oral corticotherapy with regression of ocular inflammation and with no etiology found for PS.

After stopping oral corticotherapy, the recurrence of scleritis associated with new symptoms such as bilateral auricular chondritis, non-erosive and seronegative inflammatory polyarthitis and cochlear dysfunction (with accentuation of neurosensory hearing loss, which the patient presented previously) confirmed the diagnosis.

Steroids remain the mainstay during disease flares and continued steroid use is often recommended in long-term follow-up to prevent relapses. If ineffective, immunosuppressants like methotrexate, cyclophosphamide, azathioprine, dapsone, cyclosporine, mycophenolate mofetil could be used. More recently, the efficacy of biologic agents use was evaluated in RP patients, with good results.

Our patient showed total regression of auricular chondritis and gradual regression of ocular inflammation with oral corticotherapy. However, corticotherapy was gradually tapered and replaced by methotrexate, because of the secondary effects of oral corticotherapy (Diabetes, Cushing syndrome, hypertension and osteoporosis) and a partial regression of ocular inflammation.

More than 30% of RP patients have an associated disease, such as systemic vasculitis, rheumatoid arthritis, Sjögren’s syndrome, systemic lupus erythematosus and myelodysplasia. The association of RP with myelodysplasia is described as a paraneoplastic phenomenon, being also the prognosis for RP-myelodysplasia patients more guarded than for patients with RP alone.

In conclusion, RP is a rare, autoimmune and potentially fatal disease, affecting cartilaginous and proteoglycan-rich tissues. Early recognition of the signs of this disease and prompt initiation of immunosuppressive therapy can attenuate symptoms and prevent RP complications.