Case report

Uveitis with retinal occlusive vasculitis and sensorineural hypoacusia as first symptoms of relapsing polychondritis

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ABSTRACT

We herein describe an atypical case of relapsing polychondritis, presenting initially with isolated ocular signs characterised by uveitis and retinal occlusive vasculitis preceded by 10 years of auricular and laringothracheal chondritis. This case highlights the importance of considering connective tissue inflammatory conditions in any retinal vasculitis. A systemic enquiry is invaluable in order to avoid a delayed diagnosis and the subsequent associated complications and mortality.

Introduction

Relapsing polychondritis is a rare connective tissue disorder typically characterised by recurrent inflammation of cartilagineous tissues (1). Auricular chondritis and polyarthritis are the most common signs. The eye is also a frequent target organ. Scleritis is the most frequent ocular manifestation (2), uveitis appears in about 30% while retinal vasculitis is relatively uncommon (3). Ocular signs usually occur in association with systemic disease and correlate with disease activity.

We describe an atypical case of relapsing polychondritis, presenting initially with isolated ocular signs characterised by uveitis and retinal occlusive vasculitis, preceded by a 10-year history of auricular and laringothracheal chonditis. It highlights the importance of considering connective tissue inflammatory conditions in any retinal vasculitis. A systemic enquiry is invaluable in avoiding a delayed diagnosis and the subsequent associated complications and mortality.

Case report

In March 1998, a 66-year-old Caucasian woman presented to the Ocular Immunovirology Service of the Sapienza University of Rome, with a complaint of pain, hyperaemia and blurred vision in both eyes. She had been complaining of bilateral sensorineural hypoacusia since several years and of arthralgia for a few months. At our first observation best Snellen acuity was 20/20 in the right eye and 20/25 in the left eye. Biomicroscopy revealed bilaterally keratic precipitates, mild anterior chamber flare, posterior synechiae, vitreous cells 1+. Fundus oculi evaluation bilaterally revealed pinkish optic disc with blurred margins, retinal haemorrhages, retinal vasculitis with perivascular cuffing and ghost vessels, perivascular cotton-wool spots. Fluorescein angiography revealed dye leakage and vascular staining interesting both arteries and veins (Fig. 1). Venous occlusion in upper-temporal vascular arcade with retinal ischaemia was detected in the right eye.

The patient also presented mildly painful and swollen interphangeal joints in both hands. Investigations were carried out in order to rule out infections, malignancies, and the main rheumatic diseases. ESR and C-reactive protein were normal, rheumatoid factor was absent, antinuclear antibodies were detected on Hep2 by indirect immunofluorescence (titre 1:160), IgG and IgM anti-cardiolipin (aCL) antibodies were detected at a low concentration (aCL= IgG 17.60 GPL, IgM=17.46 MPL with normal value for our lab IgG aCL <15 GPL and IgM aCL <15 MPL) by a commercially available enzyme-linked immunosorbent assay (Diamedix, Miami FL, USA). Lupus anticoagulant and anti-ß2-GPI antibodies were absent, anti-DNA and anti ENA were also negative.

High-frequency audiometry revealed neurosensorial hearing loss. Brain magnetic resonance was negative while x-rays were negative. Hand x-rays showed no erosions.

A diagnosis of bilateral diffuse uveitis complicated by retinal occlusive

vasculitis was than performed and she was treated with methylprednisone 1% drops three times a day and oral prednisone 50 mg tapered in five months. Despite the presence of sensorineural hypoacusia and non-erosive hand arthritis, and ANA, the clinical criteria were not sufficient for a diagnosis of systemic disease associated to the ocular inflammation such as Cogan's syndrome, Vogt-Koyanagi-Harada disease and systemic lupus erythematosus.

Visual acuity gradually worsened bilaterally due to complicated cataracts which were removed by phacoemulsification at the age of 74 and 75 years-old, respectively, in the right and in the left eye, thus obtaining complete recovery of visual function.

In December 2006 the patient consulted a gastroenterologist for persistent diarrhoea. An abdominal ultrasonography revealed an abdominal aortic aneurysm. A vascular prosthesis was applied with good results. In September 2009 the patient presented ankle arthritis. ESR increased to 45 mm in 1st hour and CPR also increased. In January 2010 she presented with a painful swelling of the left auricle, and a diagnosis of auricular chondritis was then made. One month later the patient experienced episodes of mild stridor. Laryngoscopy revealed swelling of vocal cords and collapse of periglottic structure. Coesistence of aortic aneurisma, auricular chondritis, sensorineural hypoacusia, laryngeal chondritis, non-erosive arthritis together with ocular inflammatory involvement led to a conclusive diagnosis of relapsing polycondritis on the basis of the criteria described by McAdam (4). Treatment with prednisone 1 mg/kg was prescribed with complete recovery of auricular and laryngeal chondritis in a few days. She was fitted with an auricular prosthesis for persistent hypoacusia.

The patient is now 80 years old. Uveitis showed only mild recurrences while retinal vasculitis did not progress. Visual acuity at follow-up is 20/20.

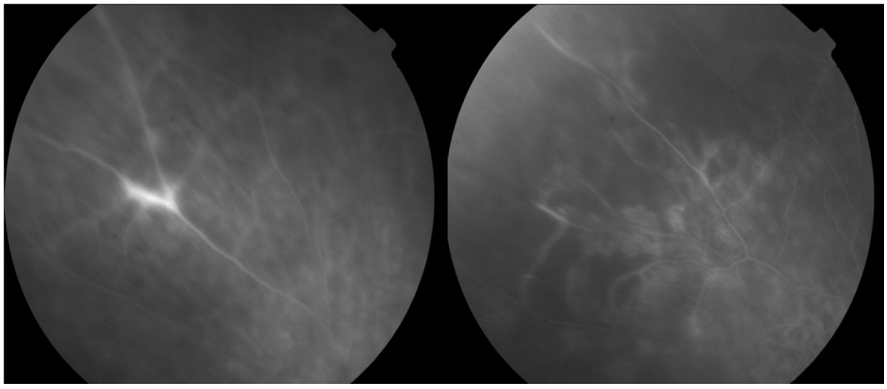

Fig. 1. Fluorescein angiography in the right eye revealed dye leakage and vascular staining in upper-temporal vascular arcade with retinal ischaemia.

Discussion

Relapsing polychondritis is characterised by recurrent flares of inflammation of cartilaginous tissue and of other tissue rich in proteoglycans with progressive lesions of the affected organs (1, 5). Recurrent inflammation of auricular cartilage is the most common manifestation (6). Asymmetrical non-erosive arthritis occurs in 75%: it can affect all synovial joints. Upper respiratory tract cartilage can be involved and the most threatening complication is the involvement of the media of the arteries and heart valve which can lead to fatal outcome (7). Hearing is frequently impaired. Sensorineural hypoacusia may be due to a vasculitis of the vestibular or cochlear branch of the internal auditory artery (4).

Ocular symptoms are a major component of the disease, being present in 60-70%. Most frequent is scleritis (8, 9); uveitis appears in 30% as an anterior uveitis or a sclerouveitis. Retinal vasculitis is relatively uncommon. Isaak (10) described nine patients with retinopathy consisting of microaneurysms, haemorrhages and cotton-wool spots. Two other patients had retinal vasculitis associated with vein occlusions. Venous or arterial thrombosis in relapsing polychondritis may be related to the presence of serum antiphospholipid autoantibodies (11) which were detected also in our patient, although at low titer, and were no longer present subsequently. It is well known that in almost one-third of relapsing polychondiritis patients an overlap with other diseases, such as systemic vasculitis, connective tissue disorders, or myelodysplastic syndromes can be found (12). However, a significant association between relapsing polychondritis and anti-phospholipid antibodies syndrome is still not convincing.

Laboratory abnormalities in relapsing polychondritis are non-specific and simply reflect a chronic inflammatory process. Relapsing polychondritis may go unrecognised for several months. The mean time from the onset of symptoms to diagnosis is 2.9 years (13), although cases of up to 8 years of diagnostic uncertainty have been reported (14). From 19% to 32% of patients had ocular symptoms at the time of presentation and 50%–57% developed ocular manifestations later on (9, 15). In our patient the diagnosis was delayed for many years from her first symptoms: bilateral uveitis with retinal vasculitis, occurred 10 years before the cohort of typical clinical manifestations which led to the diagnosis of relapsing polychondritis preceded only by hypoacusia. By that time other causes of retinal vasculitis were investigated and ruled-out. Even if some cases of severe relapsing polychonditis have been described where the use of biologic agents was required (16, 17), in the present case only corticosteroids were successfully used. Our case, in which the full spectrum of relapsing polychondritis was present ten years from atypical ocular manifestation, reflects the important role of ophthalmologists in diagnosing this rare disease. We stress the importance to continually monitor patients' medical conditions to see whether, over time, all or some can be tied together with one diagnosis bearing in mind that uveitis and retinal vasculitis are not static conditions.

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