Recurrence Anterior Uveitis with Hypopyon Revealing Relapsing Polychondritis

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Abstract

Relapsing polychondritis (RP) is a rare disease involving cartilaginous structures, predominantly the ears, nose and laryngotracheobronchial tree. The eyes, cardiovascular system, peripheral joints, skin, and central nervous system may also be affected. Involvement of all ocular structures has been described. Non-granulomatous uveitis is a common feature; however hypopyon is uncommon. We herein present the case of a 42-year-old man who had been diagnosed with bronchial asthma for a long period; and who presented with recurrent hypopyon uveitis as a revealing manifestation of relapsing polychondritis. We emphasize that RP should therefore be considered in the differential diagnosis of sterile hypopyon uveitis.

Key words: relapsing polychondritis, uveitis, hypopyon

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Introduction

Relapsing polychondritis (RP) is a rare, idiopathic systemic autoimmune disease affecting cartilaginous structures. It is characterized by recurrent inflammatory episodes involving predominantly the ears, nose and laryngotracheobronchial tree. Multiple organ systems, including the eyes, cardiovascular system, musculoskeletal system, skin, kidneys and central nervous system may also be affected (1, 2). Ocular manifestations occur in 50% to 70% of patients and may be the presenting symptom of the disease in one third of patients (3-6). The most common ocular symptoms are scleritis and episcleritis; however, all other ocular structures can be affected, including the conjunctiva, cornea, lens, vitreous, retina, and optic nerve. Non-granulomatous anterior uveitis is a common feature; however uveitis with hypopyon is uncommon. In this paper, we report the case of a patient who had been diagnosed with bronchial asthma for a long period of time who presented with recurrent hypopyon uveitis as a revealing manifestation of relapsing polychondritis.

Case Report

A 42-year-old white man was referred to our department in November 2011 for an evaluation of recurrent bilateral anterior uveitis. The patient’s ocular history began in January 2011 when he experienced pain and irritation in his eyes. He did not report any history of ocular trauma. He consulted ophthalmology first when a Slit-lamp examination revealed anterior chamber activity with hypopyon in both eyes. There was no evidence of scleritis or episcleritis. The patient’s visual acuity was 30/100 in the right eye and 40/100 in the left eye. His extraocular movements were full and the applanation pressure was within normal limits. A dilated funduscopic examination demonstrated no vitreous cells. The macula, vessels and periphery of the fundus were normal with no vascular sheathing or signs of retinitis. The patient was started on topical prednisolone acetate 1% and timolol 0.5% in both eyes every two hours with partial improvement.

Upon admission to our Department, the patient complained of marked irritation with photophobia in both eyes. According to his past medical history, he had developed an
acute shortness of breath with expiration in 2002. Rapid aggravation of dyspnea with episodes of asphyxia was observed, resulting in a tracheotomy. Since then, the patient had been treated for bronchial asthma with inhaled steroids and beta-agonists. He also reported a three year history of intermittent swelling and erythema in both auricles that usually resolved spontaneously. There was no history of fever, weight loss or musculoskeletal symptoms. Intense anterior chamber activity with hypopyon was observed on ophthalmological examination (Fig. 1). A physical examination revealed thickening of both external ears with moderate edema and erythema of the auricles (Fig. 2); however, the ear lobules were spared. The patient’s nose was red and tender with a collapsed nasal bridge (Fig. 3). No aphthous lesions were found in the oral or genital mucosa. A thorough physical examination was unremarkable.

The erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) level, full blood count, protein electrophoresis with immunoglobulin, renal and liver function tests proved normal. There was no proteinuria. The remainder of the work-up, including antinuclear antibodies, rheumatoid factor, HLA-B27, HLA-B51, c and p antineutrophilic cytoplasm antibodies; and syphilis serologic results, was negative.

Chest X-ray and electrocardiography demonstrated no abnormalities. Thoracic computed tomography (CT) showed thickening of the airway walls from the trachea to the bilateral principal bronchi with a reduced luminal diameter typical of RP (Fig. 4). CT also showed a depressed height of the nasal cartilage in comparison with the tip of the nasal bone. A biopsy of the auricular cartilage was performed. It showed degenerated and mononuclear cells predominantly infiltrated into the fibrous tissue consistent with RP. A diagnosis of RP was made on the basis of the Michet criteria (6): recurrent chondritis of both auricles, chondritis of the nasal cartilage, chondritis of the respiratory tract and ocular inflammation (uveitis).

The patient was started on oral prednisone at a dose of 40 mg daily and responded well to the therapy. Fifteen days after treatment was initiated, his visual acuity improved and the hypopyon resolved. At present, the patient’s disease process is stable and his airway symptoms are minimal.

Discussion

RP is a rare autoimmune connective tissue disorder affecting primarily cartilaginous tissues (5). It is characterized by recurrent episodes of inflammation involving cartilaginous structures of the whole body (elastic cartilage, hyaline cartilage, fibrocartilage) resulting in tissue damage and destruction (5). It is most common in patients between the ages of 40 and 60; however, it may affect children and the elderly. Both genders are affected equally, and no familial predisposition has been noted (3-6). The disease causes repetitive inflammation, particularly affecting the ears, nose and tracheobronchial tract. Other proteoglycan-rich structures such as the joints, eyes, inner ear, blood vessels, heart and kidneys may be involved (1, 7).

The most common early signs of RP are auricular chondritis, and polyarthritis occurring in over 80% of patients (5, 8). However, patients with RP present with a wide
spectrum of clinical signs and symptoms that often raise major diagnostic dilemmas (8). Ocular manifestations are frequent in the course of the disease. They are found in 50% to 70% of patients and may be the presenting symptom in as many as one third of patients (3-5). They are correlated with disease activity and usually occur in conjunction with systemic disease (9, 10). Common ocular manifestations include episcleritis or scleritis, keratitis and keratoconjunctivitis sicca (11, 12). Uveitis has been reported in 3% to 22% of cases and can compromise visual outcomes (3, 6). In one series, 3.1% of patients with scleritis were diagnosed with RP (13). Another series found that 8.2% of the patients with scleritis-associated uveitis had RP (14). Typically, nongranulomatous anterior uveitis is seen (15), often in conjunction with scleritis. Cyclitic membranes have been reported (16); however, hypopyon uveitis without scleritis is very exceptional (17). To our knowledge, only three reports (four cases) have been published (10, 17, 18). Anderson et al. (10) reported two patients with hypopyon uveitis associated with RP. The first patient was a 70-year-old man who developed hypopyon uveitis with scleritis and migratory polyarthritis as the presenting symptoms of the disease. In the second patient, a 42-year-old woman, the diagnosis of RP had been made 10 years before the occurrence of bilateral hypopyon uveitis. In the other two papers, the authors describe two cases of hypopyon uveitis without scleritis as a manifestation of RP (17, 18). This case represents an unusual and original form of RP in a patient presenting initially with respiratory symptoms treated for asthma who developed recurrent anterior uveitis with hypopyon. The present patient had been treated for bronchial asthma for approximately 10 years. Although the patient exhibited flaring/swelling of the ear auricles and a saddle nose deformity, this was not considered to be related to his respiratory symptoms by his doctor, thus leading to a misdiagnosis of RP. Trentham and Le (8) reported that the mean interval from the occurrence of the first symptom to the diagnosis of RP is three years.

Hypopyon is associated with a variety of ocular disorders. In both intraocular infection and inflammation, hypopyon consists largely of tissue debris, fibrin, inflammatory by-products and leukocytes (19). Ramsay and Lightman (20) classified the causes of hypopyon into infectious agents, non-infectious causes (including uveitis, medical agents and surgical causes), neoplasia or “masquerade” syndromes and cornea-related disorders. Behçet disease, HLA-B27-associated acute anterior uveitis (AAU) and diabetic iridocyclitis are the principal differential diagnoses for non-infectious hypopyon uveitis. A large number of systemic inflammatory disorders can affect the eyes and be complicated by hypopyon uveitis, including several of the HLA-B27-associated autoimmune diseases (such as ankylosing spondylitis (AS), Reiter’s syndrome, psoriatic arthritis and inflammatory bowel disease), Behçet’s disease and mouth and genital ulcers with infected cartilage (MAGIC) syndrome (10). Relapsing polychondritis (RP), varicella zoster virus (VZV) uveitis, ulcerative colitis (UC) and human T-lymphotropic virus (HTLV-I)-associated uveitis (HAU) should all be included in differential diagnosis.

As demonstrated in this case, we emphasize the importance of considering connective tissue inflammatory conditions in any uveitis unresponsive to topical medications. A systemic enquiry is invaluable in avoiding a delayed diagnosis and subsequent associated mortality (11). The patient presented in this report did not meet the clinical criteria for either Behçet’s disease or any of the HLA-B27 related diseases.

As no specific tests are available for RP, the disease must be diagnosed on clinical grounds. Many diagnostic criteria have been established by McAdam et al. (4), Michet et al. (6) and Damiani et al. (21). In our case, auricular chondritis, nasal chondritis and respiratory tract chondritis were noted in addition to cartilage destruction identified on an auricular cartilage biopsy, thus leading to a diagnosis of RP.

The prognosis of patients with RP is variable and it depends on organ involvement and response to treatment. Mild forms of the disease are treated with anti-inflammatory and antineutrophilic agents (dapsone, non-steroidal anti-inflammatory drugs, colchicine and/or low doses of oral corticosteroids). Advanced cases, including those involving...
acute airway obstruction, multiple relapses and cardiovascular disease, may require high doses of prednisone (1 mg/kg per day) or even intravenous pulse methylprednisolone (1 g per day for three days) (1, 4, 5, 8). For patients with signs of systemic vasculitis or who develop resistance to steroids, a variety of immunosuppressive agents, including cyclophosphamide, azathioprine, mycophenolate mofetil and methotrexate, are reported to be useful (8). Tracheostomy may be required in the setting of respiratory distress and RP localized around the glottic area (8). In our patient, recurrent hypopyon uveitis was treated successfully with oral steroids and RP responded well to treatment with systemic steroids; however in one patient (10), relapse of resistant scleritis required treatment with azathioprine.

In summary, RP is a rare and difficult to diagnose condition with a variety of non-specific manifestations. It represents a rare and uncommon etiology of hypopyon uveitis. To our knowledge, this case represents the fifth case of hypopyon uveitis caused by RP. This report highlights the importance of considering connective tissue inflammatory conditions such RP in the differential diagnosis of hypopyon uveitis.

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References