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Review

Relapsing polychondritis

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ABSTRACT

Relapsing polychondritis (RP) is a rare disease in which recurrent bouts of inflammation, in some cases followed by destruction, affect the cartilage of the ears, nose, larynx, and tracheobronchial tree. At presentation, however, arthritis is the most common manifestation and more than half the patients have no evidence of chondritis. The subsequent development of chondritis provides the correct diagnosis in patients who present with polyarthritis, ocular inflammation, or skin or audiovestibular manifestations of unknown origin. A concomitant autoimmune disease is present in one-third of patients with RP. The pathogenesis of RP involves an autoimmune response to as yet unidentified cartilage antigens followed by cartilage matrix destruction by proteolytic enzymes. The diagnosis rests on clinical grounds and can benefit from use of Michet's criteria. Anti-collagen type II and anti-matrilin-1 antibodies are neither sensitive nor specific and consequently cannot be used for diagnostic purposes. In addition to the physical evaluation and laboratory tests, useful investigations include dynamic expiratory computed tomography, magnetic resonance imaging, Doppler echocardiography, and lung function tests. Bronchoscopy has been suggested as a helpful investigation but can worsen the respiratory dysfunction. The treatment of RP is not standardized. The drug regimen should be tailored to each individual patient based on disease activity and severity. Glucocorticoid therapy is the cornerstone of the treatment of RP and is used chronically in most patients. Immunosuppressive agents are given to patients with severe respiratory or vascular involvement and to those with steroid-resistant or steroid-dependent disease. Methotrexate is often effective. Cyclophosphamide is used in severe forms.

1. Introduction

Relapsing polychondritis (RP) is a rare connective tissue disease in which recurrent bouts of inflammation, followed in some cases by degeneration and deformation, involve the cartilage of the ears, nose, larynx, and tracheobronchial tree [1–6]. RP is a systemic disease, as shown by the frequent presence of arthritis, ocular inflammation, audiovestibular involvement, skin lesions, heart valve incompetence, and vasculitis. This article reviews the current knowledge about RP with special emphasis on the most recent data.

2. Method

We searched Medline using the following indexing terms: relapsing polychondritis, chondritis, cartilage, associated autoimmune disease, arthritis, respiratory tract, imaging, computed tomography, MRI, PET scan, pathogenesis, and treatment. Reference lists of retrieved publications were searched manually for additional articles.

3. Epidemiology

RP chiefly affects middle-aged adults, with a slight female predominance. Cases have been reported in the very young and very elderly [7]. All ethnic groups are affected. In the US, the estimated annual incidence of RP is $3.5/10^6$ population [3] and the prevalence among Department of Defense beneficiaries is $4.5/10^6$ [6].

4. Clinical manifestations

4.1. Presentation

The presenting manifestations are highly variable. Joint symptoms often inaugurate the disease, and chondritis is absent initially in nearly half the cases, resulting in diagnostic delays of several months or years. The development of chondritis provides

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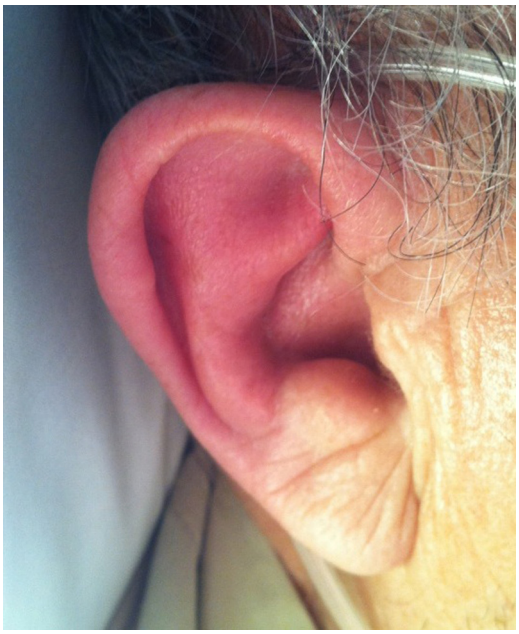

Fig. 1. Acute auricular chondritis in a patient with relapsing polychondritis. The pinna is swollen, red, and painful. Note the sparing of the ear lobe, a structure than contains no cartilage.

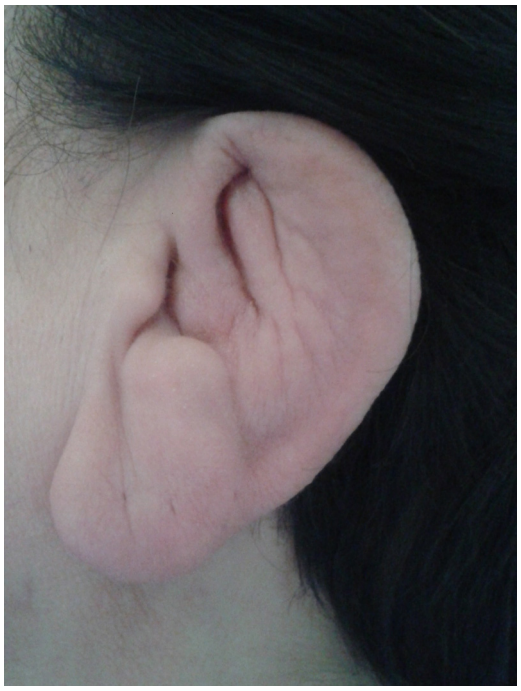

Fig. 2. Appearance after several flares of auricular chondritis in a patient with relapsing polychondritis: the auricular cartilage is floppy and misshapen.

the correct diagnosis in patients evaluated initially for joint, ocular, cutaneous, or audiovestibular abnormalities that were either misinterpreted or unexplained. In patients with chondritis at presentation, the pinna is the most common site of involvement. Unexplained prolonged fever may be the presenting symptom of RP.

4.2. Chondritis

Chondritis is the characteristic abnormality and is required for the diagnosis of RP. Recurrent inflammation of cartilage structures may eventually result in local degeneration and atrophy.

4.2.1. Auricular chondritis

Auricular chondritis is specific of RP once a local disease or infection has been ruled out. Auricular chondritis is present in 20% of patients at presentation and in 90% at some point during the course of the disease. One or both ears may be affected. The entire pinna is swollen, red or less often purplish, warm, and painful to even the slightest touch. The ear lobe, which contains no cartilage, is not affected (Fig. 1). These manifestations last a few days or more rarely a few weeks then resolve spontaneously only to recur at variable intervals. The cartilage collapses, resulting in the cauliflower ear deformity (Fig. 2). The pinna may be either floppy or hardened by calcifications or ossification of the connective scar tissue that replaces the cartilage. Cauliflower ear deformity occurs in about 10% of patients.

4.2.2. Chondritis of the nose

Nasal chondritis is a presenting manifestation in 15% of patients and occurs at some point in 65%. The inflammation involves the bridge of the nose and is often less marked than at the ears. Nasal obstruction is an uncommon feature. Atrophy may develop secondarily or insidiously, resulting in collapse of the nasal septum with saddle-nose deformity, which is painless but irreversible (Fig. 3).

4.2.3. Chondritis of the larynx and tracheobronchial tree

These sites are involved in 10% of patients at presentation and 50% over the course of the disease. Laryngeal and tracheobronchial involvement is more common among females and should be sought routinely, particularly as it causes one-third of all deaths among RP patients [1].

Laryngeal chondritis manifests as pain above the thyroid gland and, more importantly, as dysphonia with a hoarse voice or transient aphonia. Recurrent laryngeal inflammation may result in laryngomalacia or permanent laryngeal stenosis with inspiratory dyspnea that may require emergency tracheotomy as a temporary or permanent measure [8].

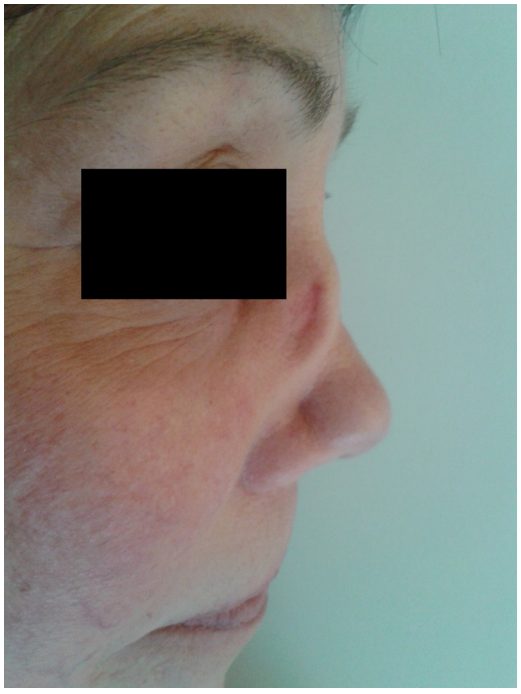

Fig. 3. Saddle-nose deformity due to collapse of the cartilaginous nasal septum.