Relapsing polychondritis is a rare autoimmune disorder defined by recurrent episodes of inflammation of cartilaginous structures (especially the ears, nose and tracheobronchial tree). We present a case of polychondritis with an onset during the seventh decade of life, which simultaneously involved both auricular pavilions. A 65-year-old woman referred to our department for intensely painful, erythematous infiltrated plaques, with symmetrical disposition at the cartilaginous part of the ear, sparing the lobule and the presence of erosion at the right antihelix level. The disease onset was 3 months ago. Physical examination and laboratory investigations revealed the coexistence of aortic insufficiency, dorsal spondylosis and bilateral coxarthrosis; the histopathological examination pleaded for the diagnosis of polychondritis. The patient was started on systemic corticosteroids. Given the frequent association of relapsing polychondritis with other autoimmune diseases, the patient will be closely monitored to early detect other pathological conditions that can worsen the vital prognosis.
Introduction

Relapsing polychondritis (RP), also known as chronic atrophic polychondritis is a rare autoimmune disorder defined by recurrent episodes of inflammation of cartilaginous structures (especially the ears, nose and tracheobronchial tree) (1).

Clinical case

A 65-year-old woman, urban area, was referred to the Dermatology Clinic of Craiova, in 2014, for 2 intensely painful, erythematous infiltrated plaques, with symmetrical disposition at the cartilaginous part of the ear, sparing the lobule, which had been present for approximately 3 months. About two months before the presentation to the hospital she affirms the appearance of an erosion located at the right antihelix, 0.5 cm in diameter, well defined, covered by an adherent hematic crust (Figure 1, 2).

Personal medical history: Nodular goiter (2008), stage 2 primary hypertension (2004), osteoporosis (with a 4-month history), total hysterectomy with bilateral anexectomy for uterine fibromas (2005).

The patient was under chronic treatment with: Euthyrox 50 µg/day, Lacidipine 2 mg /day, Nebivolol 5 mg/day, Preductal 40 mg/day, ALPHA D3 0.25 µg /day.

Clinical examination revealed: good general status, afebrile, bilateral hip pain (mechanical nature), normal pulmonary stethacoustic parameters, liver was palpable 1.5 cm below the right costal margin. The patient had tinnitus without response to specific therapy and also hardness in leg flexion with marked difficulty in getting up from a chair.

Abdominal ultrasonography was normal.

Thyroid ultrasound - isthmus 5.8 mm, right lobe 15/18/43 mm, diffuse inhomogeneity, macronodular appearance. Left lobe 16/19/41mm presents the same inhomogeneous and macronodular structure, with hyperechoic formations 4.5 mm in diameter.

Chest radiography: no active lung pathology, no pleural effusion; prominent aortic arch; moderate thoracic spondylosis.

Radiography of the pelvis: osteophytosis of the greater trochanter on infero-external contour and slight osteophytosis in the rim of the acetabulum.

Cardiac ultrasound - Left ventricular hypertrophy, diastolic heart failure by impaired relaxation, mild aortic regurgitation, secondary pulmonary hypertension, left atrial enlargement.

Routine laboratory investigations were normal, except for raised erythrocyte sedimentation rate (18mm/h). Serum protein electrophoresis, antinuclear antibodies, rheumatoid factor (14UI/mL), circulant immune complexes (2RU/mL), PPD and VDRL test were normal.

The histopathological exam (right ear antihelix) showed: ulcerated epidermis with areas of acanthosis, papillomatosis, underlying chronic inflammatory cell infiltrate, predominantly lymphoplasmacytic and diffuse interstitial infiltrate (Figure 3).

Based on the clinical and paraclinical data, we established the following diagnoses: relapsing polychondritis; stage 2 primary hypertension; aortic insufficiency; nodular goiter; dorsal spondylosis; bilateral coxarthrosis.

Recommendations at hospital discharge:

- Low dietary sodium and fat intake;
- Treatment with Prednisone 30mg/day as a single dose in the morning, Omeprazole 20 mg/day, Aspacardin 1t/ day, Piascledine 300 mg / day;
- Continue the monitoring program and the treatment for cardiovascular and endocrine diseases.
Discussions

Relapsing polychondritis was first described in 1923 by the Austrian internist Rudolf von Jaksch-Wartenhorst under the term of “polychondropathia” and in 1960, Pearson and his colleagues called it “relapsing polychondritis” (2).

It is more common in Caucasians, in the fifth decade of life (3).

There is a slight predilection for females (sex ratio F/B: 3.1/1) (4).

It is a rare disorder with an incidence of 3.5 per million people affected annually (5).

The etiopathogenesis is complex and poorly understood.

There are arguments that support the involvement of autoimmune mechanisms (6): Antibodies to types II, IX, and XI collagen in the serum of patients (30%-70%);

Immunoglobulin and complement deposits are detected at sites of inflammation;

A significantly higher frequency of HLA-DR4 than in healthy individuals;

Macrophage migration inhibitory factor (MIF) levels were significantly higher than in controls;

Elevated levels of anti-matrilin-1 antibodies (cartilage-specific matrix protein highly expressed in tracheal, nasal, auricular and costal cartilages).

There have been described rare cases of relapsing polychondritis after administration of hormones (human chorionic gonadotropin injections) or after auricular cartilage trauma (ear piercings) (7,8).

The higher prevalence of the disease among patients with connective tissue diseases raises the question of common pathophysiological mechanisms.

McAdam et al have reported that 25%-35% of patients with relapsing polychondritis have an associated autoimmune disease (9).

The autoimmune conditions reported in patients with Relapsing Polychondritis include: Wegener’s granulomatosis, polyarteritis nodosa, Churg-Strauss syndrome, microscopic polyangiitis, systemic lupus erythematosus, Sjogren’s syndrome, Raynaud’s syndrome, systemic sclerosis, Bechet’s disease (MAGIC syndrome), rheumatoid arthritis, endocrinopathies, ulcerative colitis disease Crohn’s disease, type I diabetes, pernicious anemia, psoriasis, primary biliary cirrhosis, etc. (10).

Relapsing polychondritis is a multisystemic disease with the following clinical features (11):

Musculoskeletal—nonerosive seronegative inflammatory polyarthritis, myalgias, back pain and migratory or generalized arthralgias;

Audiovestibular damage: especially nocturnal ear pain, sudden loss of hearing, vertigo and tinnitus;

Respiratory tracts chondritis: dyspnea, wheezing, hoarseness, cough, recurrent pneumonia;

Nasal: saddle-shaped nose, nasal obstruction, rhinorrhea, epistaxis;

Ocular—decreased visual acuity, conjunctivitis, episcleritis, scleritis, diplopia, eyelid edema, cataracts, optic neuritis, oculomotor nerve palsy, retinal vasculitis;

Cardiovascular disease: aortic insufficiency, pericarditis, myocarditis, conduction abnormalities, myocardial infarction;

Renal: edema, hematuria;

Neurological abnormalities: headache, ataxia, confusion, cranial nerve palsies, sensation changes, dementia, seizures.

Auricular chondritis is the most frequent manifestation of relapsing polychondritis (85%-95% of patients), affecting usually both ears. Patients have severe pain, tenderness and swelling of the cartilaginous portion of the ear. The ear lobes are not affected.

The prolonged or recurrent episodes of RP lead to a flaccid appearance of the ear (cauliflower appearance).

The differential diagnosis of RP was made with: skin tumors (squamous cell carcinoma, basal cell carcinoma), skin infection (cellulites, tuberculosis, leprosy, syphilis), contact dermatitis, gout, lupus erythematosus, cystic chondromalacia of the ear.

The episodic character of the disease leads to a delay in diagnosis.

In a study involving 66 patients, Trentham et al. observed that the elapsed time from patient presentation for medical care to the diagnosis of polychondritis was reported to be 2.9 years (12).

The presence of nasal chondritis, systemic vasculitis, laryngotracheal stricture, arthritis and anemia in patients younger than 50 years, contribute to a negative prognosis. Renal involvement is a negative prognostic factor at all ages. The goal
Table 1. Diagnostic criteria for Relapsing polychondritis

<table>
<thead>
<tr>
<th>McAdam et al.</th>
<th>Damiani and Levine</th>
<th>Michet et al.</th>
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<tbody>
<tr>
<td>1. Bilateral auricular chondritis</td>
<td>1. Three McAdam et al criteria</td>
<td>1. Proven inflammation in 2 of 3 of the auricular, nasal, or laryngotraheal cartilages</td>
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<td>2. Nonerosive seronegative inflammatory polyarthritis</td>
<td>2. One McAdam et al criteria plus positive histology results</td>
<td>2. Proven inflammation in 1 of 3 of the auricular, nasal, or laryngotraheal cartilages plus 2 other signs including ocular inflammation, hearing loss, vestibular dysfunction, seronegative inflammatory arthritis</td>
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<tr>
<td>3. Nasal chondritis</td>
<td>2. Two McAdam et al criteria plus therapeutic response to corticosteroid or dapsone therapy</td>
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<td>4. Ocular inflammation</td>
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<td>5. Respiratory tract chondritis</td>
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<td>6. Audiovestibular damage</td>
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3 of 6 clinical features necessary for diagnosis

Conclusions

Relapsing polychondritis is a rare condition that due to its episodic character may result in a delayed diagnosis, with multiple problems of differential diagnosis.

Given the frequent association of relapsing polychondritis with other autoimmune diseases, the patient will be monitored closely to detect early other pathological conditions that can worsen the vital prognosis.

The authors have declared no conflict of interest.

Bibliography