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# **Coexisting relapsing polychondritis and sarcoidosis: an unusual association.**

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**Abstract**

Relapsing polychondritis is an episodic and progressive systemic inflammatory disease characterized by auricular chondritis, polyarthritits, nasal and respiratory tract chondritis. About 30% of the patients have additional autoimmune and or hematological diseases, most frequently systemic vasculitis, rheumatoid arthritis, myelodysplastic syndromes or systemic lupus erythematosus. So far, only one case of coexisting relapsing polychondritis and sarcoidosis in a patient with AIDS has been reported. We describe here a case of sarcoidosis and relapsing polychondritis in an immunocompetent patient. Physicians should be aware of this possible association.

**KEY WORDS:** Relapsing polychondritis, sarcoidosis, autoimmunity

## **Introduction**

Relapsing polychondritis (RP) is an episodic and progressive inflammatory disease of the cartilaginous structures, including elastic cartilage of the ear and nose, hyaline cartilage of peripheral joints, fibrocartilage at axial sites, and cartilage of the tracheobronchial tree (1; 2). Inflammation of other proteoglycan-rich structures, such as the eyes, heart, blood vessels, and kidneys may also be present. Diagnostic criteria for RP are listed in Table 1 (1; 2). Although the etiology of RP is unclear, an autoimmune mechanism is considered to be most likely. RP is associated with autoimmune disorders in about one third of the patients, most frequently with connective tissue diseases and vasculitides (3; 4).

Sarcoidosis is a systemic granulomatous disease that most frequently involves the lungs, lymph nodes, skin and eyes (5). An increase in polyclonal gamma-globulin serum level is frequent, with rarely autoantibodies

We report here an unusual observation of coexisting RP and sarcoidosis. To our knowledge, this association in an immunocompetent patient has not been previously reported in the literature.

## Case report

A 42-year-old woman was admitted to our hospital with complaints of swelling and pain of the left ear and arthralgia of the hands for 3 weeks. Her past medical history was significant for pulmonary sarcoidosis of 2 years. The diagnosis of sarcoidosis was based on the clinical picture of chronic cough, mediastinal lymphadenopathies, diffuse micronodular interstitial lung disease at chest imaging, and demonstration of typical granulomas at bronchial biopsy. Infectious causes were ruled out. As pulmonary function was normal and chest CT scan was stable, oral corticosteroids were not considered.

Upon examination the significant findings were: left pinna swelling, tenderness and warmth, sparing the non cartilaginous lobe (Fig. 1) and bilateral symmetrical polyarthritis involving wrists and metacarpophalangeal joints. The patient was afebrile. Heart and lung examinations were normal, and there was no evidence of peripheral lymphadenopathy, hepatosplenomegaly or eye involvement. The anamnesis revealed a first episode of chondritis of the right ear 3 years previously, and an episode of nasal chondritis 1 year before, both of which recovered spontaneously. Laboratory results revealed elevated serum level of C-reactive protein of 25 mg/L, and fibrinogen of 4.6g/L, with inflammatory profile of blood proteins electrophoresis. Blood count, liver enzymes, kidney function tests results and serum angiotensin-converting enzyme level were normal. Repeated blood cultures were sterile. Serologic investigations for HIV, hepatitis C and B viruses, *Parvovirus* B19, syphilis and *Borrelia burgdorferi* were negative. Rheumatoid factor, anti-anticyclic citrullinated cyclic peptides, antinuclear, anti-double-stranded DNA and type-II anticollagen antibodies were not detected. Serum complement levels were normal. The HLA class II DR4 was not present. Bone radiographs of hands showed no erosions. Spirometry and chest CT results remained unchanged. Fiberoptic bronchoscopy showed absence of laryngeal or tracheal abnormality.

Based on the clinical presentation, the diagnosis of RP was established and treatment with nonsteroid anti-inflammatory agents and colchicine was initiated, with clinical improvement within one week. At 6 months follow-up, she was completely asymptomatic, and physical examination was normal. Fibrinogen and C-reactive protein levels were normal. The CT scan of the chest was unchanged.

**Discussion:**

According to Mc Adam et al.(1), 25% to 35% of patients with RP have a concurrent disease (Table 2). Coexisting RP and sarcoidosis is very unusual since it has been described only once. Zandman-Goddard et al. reported a case of coexisting RP and sarcoidosis in a patient with AIDS whose disease improved with antiretroviral drug therapy (6). We report here the first observation in an immunocompetent patient. Sarcoidosis can lead to sclerouveitis, middle ear involvement, polyarthritis and saddle nose deformity that may mimic RP but does not cause chondritis (7). Moreover, hilar lymphadenopathies, interstitial lung disease, and tissue granulomas are very rare in RP. In our patient, the clinical findings described above were conclusive with RP.

Although the etiopathogenesis of RP remains unknown, there are many arguments to support the role of the immune system. First, as seen above, RP is frequently associated with autoimmune diseases, and responds to corticosteroids. Second, RP is linked to the HLA class II DR4 isotype, which is frequently associated with other autoimmune diseases (8). Third, autoantibodies to type II, IX and XI collagen and to matrilin-I, a cartilage protein, can be found in the sera of patients with RP (3; 9; 10). Moreover immunofluorescence studies of involved cartilages have shown deposits of Immunoglobulins G, M, A and C3 as well as the presence of CD4<sup>+</sup> T lymphocytes and plasma cells (11). Mice experimentally immunised with type 2 collagen or matrilin-1 develop auricular chondritis and symptoms resembling RP in humans (12).

Sarcoidosis is a systemic granulomatous disease that preferentially involves the lungs and lymph nodes. The etiopathogenesis also implicates the immune system. Sarcoidosis is the consequence of an exaggerated immune reaction mediated by CD4<sup>+</sup> T lymphocytes in response to unknown antigens (13). T-cells present in alveoli and pulmonary interstitium secrete chemotactic factors to monocytes and macrophages, in particular monocyte chemotactic protein-1, which is also secreted by CD4<sup>+</sup> T lymphocytes in patients with RP (5, 14).

Sarcoidosis and RP seem to share some etiopathogenic mechanisms: CD4<sup>+</sup> T lymphocytes are implicated by an immune reaction through recruitment of the monocytes that contribute to inflammatory lesions. This could explain why these 2 diseases can coexist in the same patient.

In conclusion, sarcoidosis should be added to the list of diseases potentially coexisting with RP. Given the multitude of associated diseases, it has been proposed that RP be considered as a syndrome which can be primary or secondary.

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**Table 1 Major diagnostic criteria for relapsing polychondritis (1; 2)**

<p><b>McAdam criteria (three of six conditions recommended to be present)</b></p> <p>Michet et al. criteria (one of two conditions recommended to be present)</p> <ol style="list-style-type: none"> <li>1. Recurrent chondritis of both auricles</li> <li>2. Inflammatory polyarthritis</li> <li>3. Chondritis of nasal cartilage</li> <li>4. Inflammation of ocular structures including conjunctivitis, keratitis, scleritis/episcleritis and uveitis</li> <li>5. Chondritis of the respiratory tract involving laryngeal and tracheal cartilages</li> <li>6. Cochlear or vestibular damage manifested by neurosensory hearing loss, tinnitus and</li> </ol>
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vertigo
<p><b>Michet et al. criteria (one of two conditions recommended to be present)</b></p> <ol style="list-style-type: none"> <li>1. Proven inflammation in two of three of the auricular, nasal or laryngotracheal cartilages</li> <li>2. Proven inflammation in one of three of the auricular, nasal or laryngotracheal cartilages plus 2 other signs including ocular inflammation, vestibular dysfunction, seronegative arthritis and hearing loss</li> </ol>

**Table 2 Diseases associated with relapsing polychondritis**

Diseases	Références
Vasculitis	(15)
Wegener's granulomatosis	(15; 16)
Polyarteritis nodosa	(15; 16)
MAGIC syndrome (mouth and genital ulcers with inflamed cartilage syndrome)	(15; 16)
Behçet's disease	(15; 16)
Hypothyroidism	(17)
Systemic lupus erythematosus	(15)
Rheumatoid arthritis	(15)
Sjögren's syndrome	(15)
Mixed connective tissue disease	(15)
Pernicious anemia	(17)
Diabetes mellitus	(17)
Myasthenia gravis	(17)
Ulcerative colitis	(17)
Common variable immunodeficiency	(18)
Myelodysplastic syndrome	(19; 20)
Hematologic malignancies	(21-23)
Solid tumor malignancies	(21-23)

**Figure 1. Auricular chondritis of the patient**

