CASE REPORT

Fever of unknown origin in a patient with red ears: relapsing polychondritis

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SUMMARY

Relapsing polychondritis (RP) is a rare autoimmune disease that affects cartilage and multiple organ system. We describe a case of RP presenting with fever, conjunctivitis and inflammatory signs of the right knee. Infectious and neoplastic diseases were excluded by imaging and laboratory examinations. After 17 days of admission the patient reported coughing and odynophagia, and, a physical examination showed red ears. Otorhinolaryngological examination demonstrated arytenoid chondritis. RP was diagnosed based on four McAdam's criteria. The patient was medicated with oral prednisone, and a positive clinical response was noted. Severe disease may require high doses of corticosteroids or immunosuppressive agents and the biological treatment may be beneficial. Beginning manifestations, such as fever, are often unspecific, leading to RP misdiagnosis. In this case, close attention to the patient's clinical history and a detailed physical examination were fundamental in concluding the correct diagnosis, and consequently beginning the appropriate medication.

BACKGROUND

Relapsing polychondritis (RP) is a rare systemic autoimmune disease of unknown aetiology, characterised by recurrent inflammation of cartilaginous and connective tissue. The most commonly affected cartilages are the ears, nose and laryngotracheobronchial tree. It can also involve multiple organ system, including the eyes, skin, musculoskeletal system, kidneys, cardiovascular system and central nervous system.

Patients may present with general and unspecific signs, such as fever, long before cartilaginous inflammation begins. Ocular manifestations and arthralgia are very common and may be the presenting symptoms.

We report a case of RP that proved to be a diagnostic challenge.

CASE PRESENTATION

A 79-year-old man was admitted to the emergency department of our hospital for fever during the preceding 3 days, accompanied by neck pain and conjunctivitis during the previous month. He had a known history of stroke, heart failure, arterial hypertension and diabetes. On physical examination, the patient was febrile and presented inflammatory signs on his right knee. He was admitted to the internal medicine ward to continue diagnostic work-up. The erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) were elevated

without leucocytosis or neutrophilia. Kidney function, urinalysis and acid uric serum concentration were normal. Infectious and neoplastic diseases were excluded by imaging and laboratory examinations (blood, urine and joint cultures; serological tests for syphilis, hepatitis B and C, HIV infection protein electrophoresis; chest radiography; cardiac ultrasound; joint ultrasound; brain, thoracic, abdominal and pelvic CT-scan; and cervical MRI).

At day 17, the patient continued febrile, reported pain in both auricles, coughing and odynophagia. He had inflammatory signs in both auricles, not including the lobules (figures 1 and 2).

Otorhinolaryngological examination demonstrated arytenoid chondritis and no involvement of nose cartilage.

Rheumatoid factor and antinuclear antibodies were negative; serum complement C3 and C4 levels were normal. Also antineutrophil cytoplasmic antibodies (ANCA) were negative excluding ANCA-associated vasculitis such as granulomatosis with polyangiitis.

RP was diagnosed based on four McAdam's criteria: symmetrical inflammation of the cartilage of the auricle, inflammation of the cartilage of respiratory tract (arytenoid), seronegative non-erosive

Figure 1 Inflammation of both auricles. Note that inflammatory signs do not extend to the lobules.

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Figure 2 Inflammation of both auricles. Note that inflammatory signs do not extend to the lobules.

arthritis (knee joint) and inflammation of the eye tissue (conjunctivitis).

OUTCOME AND FOLLOW-UP

The patient began oral prednisone at a dose of 50 mg daily and a favourable clinical response was observed. The patient no longer reported cough or odynophagia; body temperature returned to normal and the inflammatory signs of auricles and knee receded. ESR and CRP began to subside and the patient was discharged from the hospital, continuing follow-up assessment in ambulatory internal medicine consults.

DISCUSSION

RP is a rare autoimmune disease which affects cartilaginous tissues as well as proteoglycan-rich tissues including the media of the arteries, the conjunctiva and sclera of the eye.¹ The aetiology and pathophysiology are still unclear, yet it is believed to be a connective tissue disease. Antibodies to type II collagen in cartilage are often present and sometimes indicate RP activity.^{2,3} Other potential biomarkers are associated with RP, such as soluble triggering receptor expressed on myeloid cells-1.⁴

Ocular manifestations occur in 50–70% of patients and may be the presenting symptoms of disease in one-third of the patients.⁵

Cricothyroid chondritis may be presented by odynophagia and dysphonia and its progressive destruction can lead to airway stenosis.^{6,7}

Patients may present with general, unspecific signs such as fever and progressive malaise, and this may cause a significant delay between diagnosis and treatment.¹

The clinical presentation is the basis for establishing the diagnosis and requires three of the seven McAdam's criteria: symmetrical inflammation of the auricles and/or external auditory meatus cartilage; inflammation of the basal cartilage resulting in saddle-nose deformity due to collapse of the dorsum of the nose; inflammation of the cartilage of the respiratory tract (the larynx and trachea); seronegative, non-erosive arthritis; inflammation of the eye tissue (the conjunctiva, cornea, sclera,

choroid); inner ear and labyrinth lesions; histological confirmation (biopsy of the cartilaginous tissue).¹

In this clinical case, finding the origin of fever was a challenge because of unspecific signs and symptoms at the beginning, leading to many examinations. When the patient developed inflammatory signs in both ears, the diagnosis became clear. Looking back in the history finding, the patient had conjunctivitis and arthralgia. He also reported cough and swallow, revealing arytenoid chondritis. On closer examination of the clinical history and physical signs, along with an increased degree of clinical suspicion, a diagnosis was eventually reached.

The treatment consists of low-dose corticosteroids when chondritis and peripheral arthritis is observed. If significant organ involvement is proven, high-dose corticosteroids are often necessary. Severe disease may require the treatment with immunosuppressive agents.⁸ When these options prove ineffective, the biological treatment with infliximab and tocilizumab may be beneficial.^{9–11}

Learning points

- ▶ Relapsing polychondritis (RP) is a rare disease which involves cartilage and multiple organ systems, and the diagnosis is established with three of the seven McAdam's criteria.
- ▶ Differential diagnosis includes vasculitis, other autoimmune diseases, infectious and neoplastic diseases.
- ▶ Treatment consists of corticosteroids, and severe disease may require treatment with immunosuppressive agents.
- ▶ RP is often misdiagnosed due to presenting general and unspecific manifestations, such as fever.
- ▶ All details of the clinical history and physical examination revealed to be fundamental in determining the correct diagnosis and beginning appropriate therapy.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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