CASE REPORT

Relapsing polychondritis: case report and literature review

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KEY WORDS

arthritis, auriculitis, conjunctivitis, fever of undetermined etiology, relapsing polychondritis

ABSTRACT

Relapsing polychondritis (RP) is a rare connective tissue disease which affects cartilaginous tissues of the nose, earlobes, respiratory tract, and joints, as well as proteoglycan-rich tissues including the media of the arteries, the conjunctiva and sclera of the eye. The disease is most common in patients aged 40–60 years. It may for a long time cause unspecific signs (fever, malaise), which may cause a significant delay in establishing the diagnosis and initiating the appropriate treatment. The mean time from symptom onset to diagnosis is 2.9 years. Steroids are the mainstay of therapy, but methotrexate and dapsone may also be beneficial in some patients. A case of RP presenting with fever, anemia, fatigue, arthritis, conjunctivitis, and auricular deformation is described. There were signs and symptoms suggestive of an infectious disease but no infection had been found, and despite tentative administration of antibiotics, the patient's condition steadily deteriorated. Eventually, the diagnosis was established based on McAdam's criteria. After intiating steroid therapy, the patient's condition improved significantly. We discuss the diagnostic criteria and treatment of this rare disease.

INTRODUCTION Relapsing polychondritis (RP) is a rare connective tissue disease which affects the cartilaginous tissues of the nose, earlobes, upper and lower respiratory tract, and joints, as well as various proteoglycan-rich tissues. A peak incidence is observed in patients aged 40–60 years. Patients may for a long time present with general, unspecific signs such as fever and progressive malaise. We describe the case of RP that posed a considerable diagnostic challenge.

CASE REPORT A 55-year-old patient was admitted to our department because of fever, which lasted several weeks, progressive malaise, and generalized arthralgia. He also complained of a productive cough and hoarseness. He reported inflammation and edema of both auricles, which occurred several months earlier. He was addicted to alcohol and tobacco, and his living conditions were destitute.

The physical examination revealed the following abnormalities:

1 periods of everyday hectic fever >38–38.5°C alternating with periods of subfebrile state

2 deformation of the auricular cartilage – it was flabby, resembling the "cauliflower ear" of professional boxers, but without signs of inflammation. There was a slight saddle nose deformity.

3 massive crural edema with skin lesions typical of chronic venous insufficiency with no evidence of acute deep-vein thrombosis.

4 disturbed sense of joint position resulting in an unsteady gait

5 conjunctivitis of the left eye treated successfully with a topical corticoid and antibiotic preparation.

Additional tests performed on admission revealed the following abnormalities:

1 hipochromic and microcytic anemia (hemoglobin 8.3 g/dl, hematocrit 25%, mean corpuscular volume 77 fl (norm, 80–96 fl) with low serum iron concentration (<25 μg/dl, reference values, 35–250 μg/dl) and elevated ferritin levels (697 ng/ml, reference values, 3–244 ng/ml)

2 elevated inflammatory markers: C-reactive protein – 232 mg/l, α_1-globulin – 0.5 g/dl (0.2–0.35 g/dl), α_2-globulin – 1.1 g/dl (0.4–0.7 g/dl) and γ-globulin – 1.6 g/dl (1.0–1.4 g/dl), white blood

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Received: May 18, 2009.
Accepted: August 18, 2009.
Conflict of interest: none declared.
Pol Arch Med Wewn. 2009; 119 (10): 680-683

cell count 12,000/mm^3 with neutrophilia, elevated platelet count of 616,000/mm^3

3 hypoalbuminemia – 2.0 g/dl.

Based on the preliminary findings, the initial differential diagnosis included connective tissue, malignant, and infectious diseases.

Clinical management Endoscopic examination of the upper and lower alimentary tract showed an ulceration on the minor curvature of the stomach, which was histologically proven to be of peptic origin, with coexisting *Helicobacter pylori* infection. Abdominal ultrasonography, chest X-ray, otorhinolaryngological examination, and bronchoscopy were normal. Cardiac ultrasound examination excluded endocarditis.

Blood cultures obtained during fever were sterile. Urinalysis was normal. Serological tests for syphilis, hepatitis B and C, and HIV infection yielded negative results. Thyroid-stimulating hormone and prostate-specific antigen were within normal ranges.

The patient's condition deteriorated, his red blood count and serum protein concentration gradually decreased. Therefore, although no septic focus had been found and suspected infection had not been confirmed, a broad-spectrum antimicrobial therapy was started (ceftriaxone and amikacin). However, the antibiotics did not improve the patient's condition, body temperature, or inflammatory parameters.

In the second week of hospital treatment, arthralgia exacerbated. The metacarpophalangeal and interphalangeal joints of the right hand, the metatarsophalangeal and interphalangeal joints of both feet, the knee and ankle joints were swollen, tender and warm. Pain and effusion severely limited joint mobility. The latex test for rheumatoid factor was negative. A uric acid serum concentration was normal.

RP was diagnosed based on three McAdam's criteria, including a history of auricle chondritis, saddle deformity of the nasal cartilage, and seronegative, nondestructive arthritis. The patient was then referred to a rheumatologist. He received prednisone at an initial dose of 50 mg daily. A significant improvement in the patient's condition was observed, the joint pain subsided, the body temperature returned to normal, and the red blood cell count and serum protein concentration began to rise. He was able to start rehabilitation and was discharged from hospital.

DISCUSSION

RP is a connective tissue disease characterized by recurrent flares of inflammation of the cartilaginous tissue (chondritis) and of other tissues rich in proteoglycans, with progressive lesions of the affected organs.

Peak morbidity occurs in patients aged 40–60 years, although cases of children have also been described.[1] Inflammation may affect the auricular and nasal cartilage, the cartilage of the joints, airways, the media of the arteries, heart valve, sclerotic coat, and ocular conjunctiva. The mean time from the onset of symptoms to diagnosis is 2.9 years[2], although cases of up to 8 years of diagnostic uncertainty have been reported[3].

Diagnostic criteria The clinical presentation is the basis for establishing a diagnosis. The classic work of McAdams in 1976 required that 3 of the 7 criteria be fulfilled:

1 symmetrical inflammation of the cartilage of the auricle and/or the external auditory meatus

2 inflammation of the nasal cartilage resulting in saddle-nose deformity due to collapse of the dorsum of the nose

3 inflammation of the cartilage of the respiratory tract (the larynx and trachea)

4 seronegative, nonerosive arthritis

5 inflammation of the eye tissue (the conjunctiva, cornea, sclera, choroid)

6 inner ear and labyrinth lesions

7 histological confirmation (biopsy of the cartilaginous tissue).

RP may also be diagnosed on the basis of one criterion and typical histological changes, or two criteria and response to immunosuppresion. The symptoms and abnormal imaging findings in PR have been discussed in detail in the articles by Franczak-Drygalska and Kucharz[4] and by Fliciński et al.[5]

Histological findings The typical changes in the affected cartilage include considerable amounts of vacuoles in chondrocytes, absence of basophils usually present in the stained tissue, presence of fibrous tissue instead of the regular cartilage, as well as perivascular polymorphonuclear and mononuclear cell infiltrates. However, a cartilage biopsy is not necessarily required to establish the diagnosis. Grabysa et al.[6] described a patient, in whom a diagnosis of RP was also established on the basis of an auricular biopsy.

Treatment Corticosteroid treatment is not always effective, although it proved beneficial in the present case. Immunosuppressive drugs represent a second-choice treatment. Targońska-Stępniak et al.[7] described a case of RP resistant to prednisone, which was successfully treated with methotrexate (the therapeutic weekly dose of 15–25 mg), while Goldenberg et al.[8] reported efficacious treatment with mycophenolate mofetil. When both corticosteroid and immunosuppressive drugs prove ineffective, biological treatment may be beneficial. Cazabon et al. and Kawai et al.[9,10] described successful treatment of PR patients with infiliximab (a chimeric anti-tumor necrosis factor-α antibody), etanercept (a soluble tumour necrosis factor-α receptor), and tocilizumab (an anti-interleukin-6 receptor antibody). The current study shows that even if one type of biological therapy fails, there are other biological drugs that may produce beneficial outcomes.

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Nawracające zapalenie wielochrzęstne – opis przypadku i przegląd piśmiennictwa

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SŁOWA KLUCZOWE

gorączka o nieustalonej etiologii, nawracające zapalenie wielochrzęstne, zapalenie małżowiny usznej, zapalenie spojówek, zapalenie stawów

STRESZCZENIE

Zapalenie wielochrzęstne jest rzadko spotykaną układową chorobą tkanki łącznej dotyczącą tkanki chrzęstnej nosa, małżowin usznych, dróg oddechowych i powierzchni stawowych, jak również tkanek bogatych w proteoglikany: błony środkowej tętnic, spojówek i twardówek. Szczyt zachorowalności przypada na 40–60 rż. W obrazie klinicznym choroby przez długi czas mogą dominować niespecyficzne objawy ogólne (gorączka, postępujące osłabienie), co może opóźnić postawienie diagnozy i wdrożenie odpowiedniego leczenia. Średni czas od pojawienia się dolegliwości do postawienia prawidłowego rozpoznania wynosi aż 2,9 roku. Podstawą leczenia jest sterydoterapia, zastosowanie znajduje również metotreksat i dapson. Poniżej omówiono przypadek zapalenia wielochrzęstnego, objawiającego się gorączką, niedokrwistością, złym stanem ogólnym, zapaleniem stawów, zapaleniem spojówek oraz zniekształceniem małżowin usznych. Objawy sugerowały chorobę infekcyjną, jednak nie wykryto ogniska zakażenia, a empiryczna terapia antybiotykami nie poprawiła stanu chorego. Ostatecznie rozpoznanie postawiono opierając się na kryteriach podanych przez McAdama. Zastosowanie sterydoterapii spowodowało znaczną poprawę stanu chorego. Omówione zostały także kryteria rozpoznania tej rzadko występującej układowej choroby tkanki łącznej oraz metody leczenia.

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Praca wpłynęła: 18.05.2009.
Przyjeta do druku: 18.08.2009.
Nie zgłoszono sprzeczności interesów.
Pol Arch Med Wewn. 2009; 119 (10): 680-683

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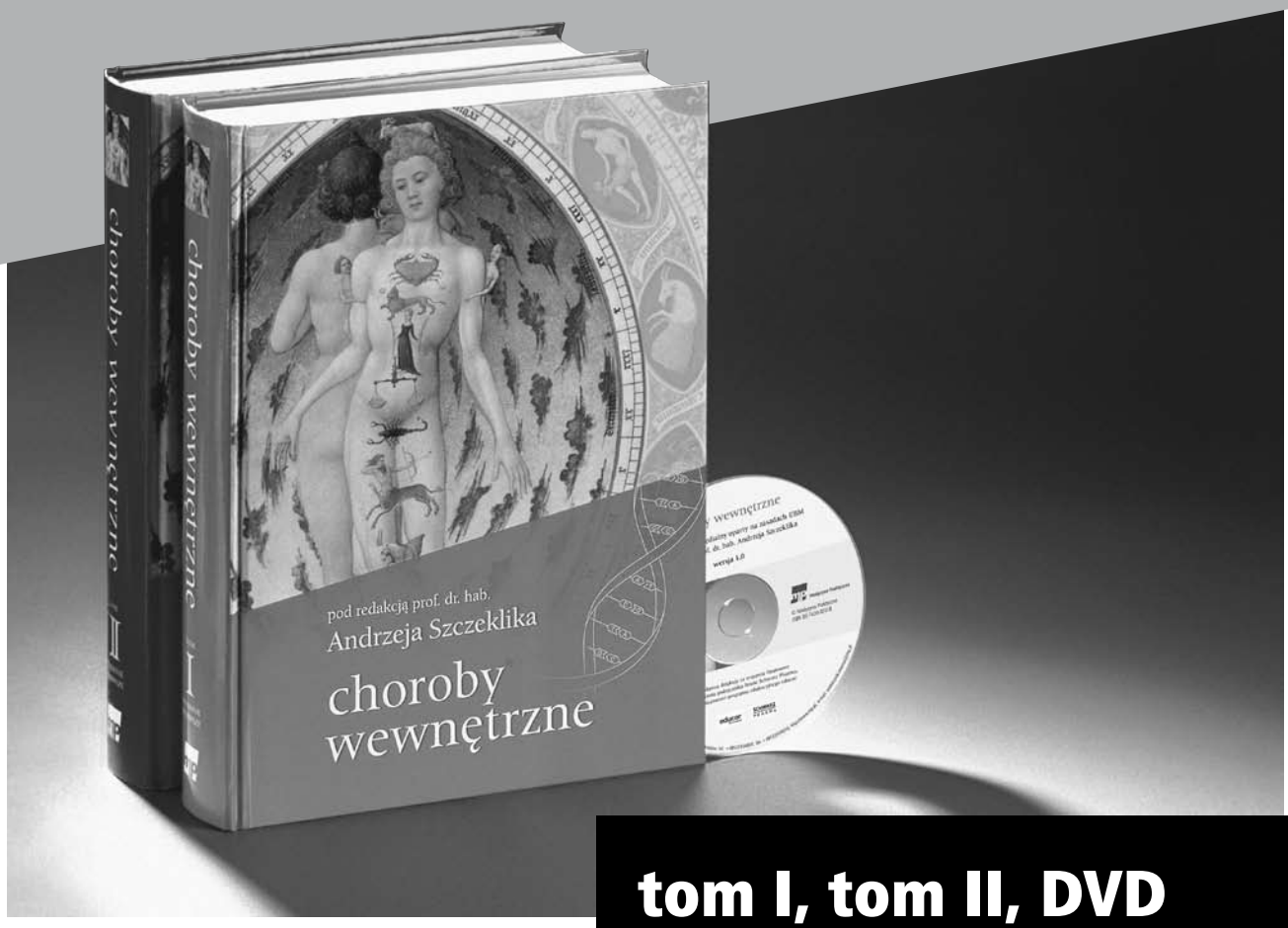

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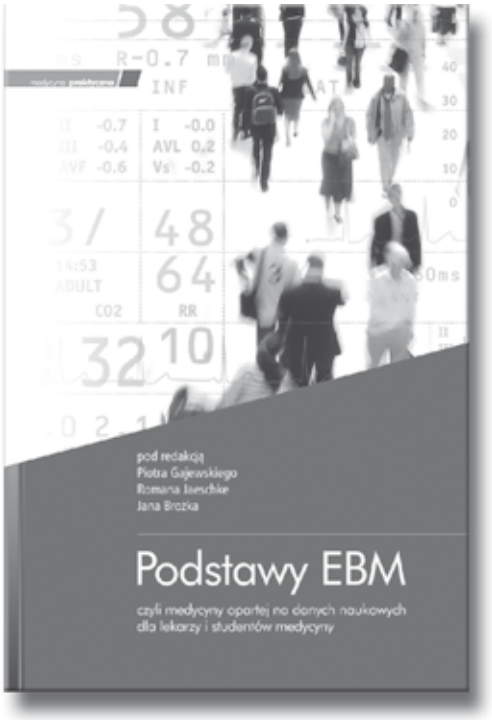

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