Case Report

A Case of Relapsing Polychondritis with Positive C-ANCA

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Abstract

Relapsing polychondritis (RP) is an auto-immune disease with varied presentation. We are reporting a case of RP presented with auricular chondritis, sensory neural hearing loss, joint pain, skin rash with ulcer over knee. This patient also had positive anti-neutrophil cytoplasmic antibody (C-ANCA for PR3) which makes it an attention-grabbing case.

Key-words: Relapsing polychondritis, C-ANCA

(Journal of The Indian Academy of Geriatrics, 2017; 13:162-164)

INTRODUCTION

Relapsing polycondritis (RP) is an auto-immune disease of unknown etiology. Auricular chondritis with sparing of ear lobe is characteristic feature of this disease.[1,2] It is also associated with other connective tissue diseases i.e. Rheumatoid arthritis, wegeners granulomatosis, oral/genital ulcers, SLE, microscopic polyangitis.[3,4] Anti-neutrophil cytoplasmic antibody (ANCA) positivity is not uncommon in this disease.

CASE SUMMARY:

A 55 year old Hindu male, married visited rheumatology outdoor complaining of recurrent nasal blockage along with pain and swelling in both wrist, hands, knees and ankles since two years. He also noticed change in voice since 18 months and had decreased hearing with tinnitus for 15 days. He experienced pain, redness and swelling in both the auricles intermittently for 6 months which tends to resolve in 3-4 days. He also noticed non-itchy, erythematous, variable size rash over the face and body which lasted for 10-12 days occurred two times in last 6 months. There was single ulcer over left knee for 15 days.

There was no history of prolonged fever, hemoptysis, haematuria, oral/genital ulcers, allergy, sneezing. He was smoker and occasional alcoholic. His blood pressure was normal (120/80 mm Hg) and all peripheral pulses were palpable with a rate of 90/minute. He was afebrile (oral temperature 98.6 F) with saturation of oxygen 94% by pulse oximetry at room air.

On examination erythematous, maculopapular, scaly rash was present over face and below the neck with slightly oedematous eyelids. Skin sensations were intact. Both auricles were red, tender and swollen with sparing of ear lobules (Figure 1). Alae nasi were erythmatous, tender and nasal septum was deviated. Tenderness and swelling were found on smaller and large joints. Single erythematous non-tender, 2cm ×2cm size ulcer with clean margins was observed over left knee. On systemic examination bilateral rhonchi were present. Pallor, cyanosis, icterus and clubbing were not present.

On investigation mild anemia (hemoglobin 11 gm/dl) was found with normal leukocyte (6000 cells/ mm3) and platelet count (3.2 lakh cells/mm3). Erythrocyte sedimentation rate was 70 in Ist hour. C-reactive protein and Rheumatoid factor were positive. Fasting blood sugar was 90 mg/dl. Liver and renal function tests were normal. Urine examination was also normal. Skiagram chest,

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skiagram both hands and electrocardiogram were also normal. Possibility of tuberculosis was ruled out by negative sputum test for Acid fast bacilli and negative Montoux test. HBs antigen and anti HCV were negative. Sensory neuropathy involving both lower limbs nerves (left > right) was revealed on Nerve conduction studies. Bilateral sensory neural hearing loss (affecting right ear >left ear) was found on audiometry analysis. On immune-profile analysis, anti-nuclear antibody and ACPA were negative. C-ANCA for PR3 was positive (by indirect Immunofluorescence Antibody Assay (IFA) method).

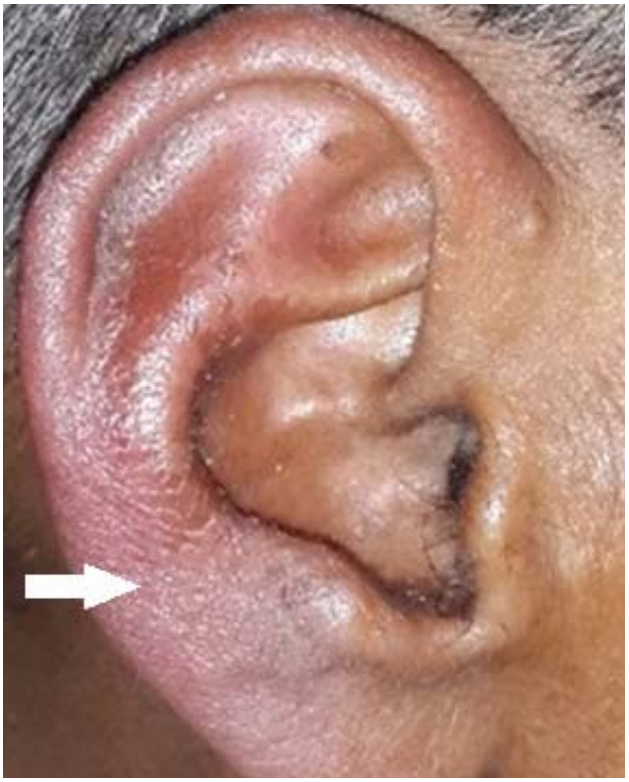

Figure 1. Relapsing polychondritis. Red, tender and swollen auricle (white arrow) with sparing of ear lobules

Biopsy of nasal septum revealed inflammatory infiltrate consisting of lymphocytes, neutrophils with cartilage destruction.

This patient had recurrent chondritis of both auricles, nonerosive inflammatory arthritis, history of change in voice suggestive of chondritis of the laryngeal and/or tracheal cartilages, cochlear damage as manifested by sensory neural hearing loss (affecting right ear >left ear) on audiometry analysis and biopsy of nasal septum showing chondritis of nasal cartilage. On the basis of these features diagnosis of relapsing polychondritis was established and treatment started with oral steroids (1 mg/kg), methotrexate 20 mg/week and folic acid 5 mg/week. He started improving.

DISCUSSION

Relapsing polychondritis (RP) is an autoimmune disorder of ill defined etiology characterized by recurrent inflammation of cartilageous structures i.e. cartilages of ear, nose, larynx and trachea-bronchial cartilage.[1] Auricular cartilage is the most affected cartilage in RP characterized by involvement of the pinna of the ear with sparing of the earlobe as earlobe is devoid of cartilage.[2] About 30% of cases of RP are associated with other auto-immune diseases including rheumatoid arthritis, microscopic polyangitis, wegeners granulomatosis, Behcet syndrome and SLE.[3] Anti Nutrophilic cytoplasmic antibody (ANCA) may be found in about one forth of RP cases.[4]

Many non-specific skin manifestations (50%) are seen in RP. These lesions includes aphthous ulcers, erythema nodosum, purpura, sterile pustules, papules, livedo reticularis, limb ulcer and distal necrosis.[4]

Sensory neural hearing loss occurs due to involvement of internal auditory artery or its cochlear branch. The diagnosis of RP is by criteria as suggested by McAdam et al (1976)[5] and later modified by Damiani and Levine (1979).[6]

Like other auto-immune disease, the presentation of RP can be highly ambiguous. In RP, ANCA may be found in 25% of cases.[4] These ANCA positive patients should be monitored closely for development of vasculitis in future.[7,8]

Treatment is oral steroid and immunosuppressant (i.e. methotrexate, azathioprine, cyclophosphamide, or cyclosporine) in steroid unresponsive/ dependent cases.

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