Relapsing Polychondritis in an Elderly Male

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Abstract
A 65 years patient presented with left ear swelling, swelling of the nasal bridge with congestion of the eyes. Clinically there was evidence of left auricular chondritis, nasal chondritis and conjunctivitis with a history of multiple similar episodes in the past, features suggestive of relapsing polychondritis. The patient improved with oral prednisolone.

INTRODUCTION
Relapsing polychondritis (RPC) is a rare autoimmune disease of unknown aetiology characterized by recurrent inflammation and destruction of cartilaginous tissues. Its estimated annual incidence is about 3.5 per million in Rochester, Minnesota. Its peak age at onset ranges from 40 to 50 years even as cases have been reported in children and in the very elderly. Although the aetiology remains obscure, an autoimmune process seems to be responsible. Only four cases have been reported in the Indian literature and we report another case.

CASE REPORT
GH, a 65-year-old male was admitted to our hospital with a 20 day history of pain and swelling of left ear which involved the right ear as well over a period of seven days. This was followed by a painful swelling of the nasal bridge and two days later by intense irritation and redness of the eyes accompanied by excessive watering. About six weeks earlier the patient had experienced arthritis of both knees and right wrist which had subsided with NSAIDs. The patient denied a relation of these symptoms to cold and there was no history of skin rash, fever, epistaxis, purulent nasal or ear discharge, chest pain, haemoptysis, hoarseness of voice, haematuria, oliguria, visual or auditory disturbances or features suggestive of peripheral vascular insufficiency.

The patient had experienced three similar episodes of painful swelling of the ears, about two years, one year and 9-months earlier. In the last attack he had throat pain with sudden onset weakness of the ocular movements with congestion of the eyes, for which he had to be hospitalized. He had been found to have bilateral palsies of third and sixth cranial nerves with evidence of orbital cellulitis, the aetiology of which was thought to be infective and he had been managed with antibiotics and anti-inflammatory agents. During the hospital stay he had developed deep vein thrombosis of the left leg which responded to anticoagulation.

Clinical examination of the patient revealed active inflammatory swelling of both ears with sparing of the ear lobule (Fig. 1) with loss of the cartilaginous architecture of the left ear. There was bilateral conjunctivitis, and active inflammation of the nasal bridge. The movements of both knees were painful without any evidence of active arthritis. Rest of the general and systemic examination was normal.

Haemogram revealed an ESR of 55mm with mild anaemia (Hb 10.9g/dl). The serum levels of urea, creatinine, glucose, bilirubin, proteins, SGPT, SGOT, albumin, calcium, phosphates,
uric acid, sodium and potassium were normal. Urine examination was normal and the culture sterile. Radiographs of the chest, soft tissue of the neck and skull were normal. Latex agglutination test for rheumatoid factor was negative and antibodies to nuclear antigen, dsDNA and ENA were not detected. LE cell phenomenon and VDRL were negative. Bronchoscopy was normal.

In view of recurrent bilateral chondritis of the pinnae with sparing of the ear lobules, nasal cartilage chondritis, conjunctivitis and nonerosive polyarthritis, a diagnosis of relapsing polychondritis was made and the patient was put on prednisolone (0.5mg/kg). The patient improved dramatically and he was discharged and steroids tapered off over six weeks. He has been symptom-free during a follow-up of over six months.

**DISCUSSION**

Relapsing polychondritis (RPC) is a rare autoimmune disease of unknown aetiology with characteristic inflammation of the cartilaginous tissues. The peak age of onset ranges from 40-50 years, but cases have been reported in children and in the very elderly. No clear cut hereditary predisposition or HLA association has been reported. An autoimmune process seems to be responsible, the hypothesis strengthened by the demonstration of antibodies to native collagen and presence of circulating immune complexes in patients of RPC, both of which correlate with disease activity. The involved cartilage on histopathology demonstrates acidophilic transformation of the cartilage matrix, inflammatory infiltration by polymorphs followed later by lymphomononuclear cells and granulation tissue with fibrosis after repeated episodes of inflammation. RPC has been reported to be associated with SLE, Sjogren’s syndrome, rheumatoid arthritis, overlap syndromes, cryoglobulinaemias and spondyloarthropathies. However, our patient did not have clinical or serological evidence of any of such disorders.

Clinical characteristics of RPC are tabulated in Table 1. Other manifestations and diagnostic criteria are listed in Table 1.

**Table 1 : Diagnostic criteria for relapsing polychondritis**

1. Recurrent chondritis of both auricles
2. Non-erosive inflammatory polyarthritis
3. Chondritis of nasal cartilages
4. Inflammation of ocular structures, including conjunctivitis, keratitis, scleritis/episcleritis, and/or uveitis
5. Chondritis of the respiratory tract involving laryngeal and/or tracheal cartilages
6. Cochlear and/or vestibular damage manifested by neurosensory hearing loss, tinnitus, and/or vertigo
7. Cartilage biopsy confirmation of a compatible histological picture

The presence of 3 or more criteria is necessary for the diagnosis of relapsing polychondritis.

Neurological manifestations include cranial neuropathies (II, VI, VII, VIII), headaches, seizures, encephalopathy and hemiplegia.

Laboratory abnormalities are nonspecific ranging from leukocytosis, thrombocytosis, chronic anaemia, raised ESR and elevated gammaglobulin levels. Low titers of rheumatoid factor and ANA can be detected. High titers of anticollagen-II antibody have been reported. Radiographic abnormalities may include tracheal stenosis and cartilaginous calcifications.

Mild auricular involvement can be treated with nonsteroidal anti-inflammatory agents and/or low dose steroids. In severe disease prednisone in the dosage of 1mg/kg is administered. Immunosuppresion with methotrexate, azathioprine, cyclosporine and cyclophosphamide may be needed in patients who do not respond to steroids. Surgical intervention may be needed in case of complications involving the respiratory tract like tracheal stenosis and tracheomalacia and stents may be required for tracheobronchial collapse.

Five-year survival of up to 74% and 10-year survival of 55% has been reported in a study of 112 patients from Mayo clinic, major causes of death being infections and systemic vasculitis. About 15% of the deaths were as a direct result of cardiovascular or respiratory tract involvement. Bad prognostic indicators included coexistent vasculitis and early saddle nose deformity in the young and the presence of anaemia in older patients.

**REFERENCES**