

Relapsing Polychondritis Associated with Rheumatoid Arthritis

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Introduction

Relapsing polychondritis is a rare multisystem autoimmune vasculitic disorder¹⁻³. Its manifestations characteristically episodic in nature⁴ comprise inflammation of cartilaginous structures including the eyes and ears⁵. Approximately thirty percent of the cases are associated with other autoimmune disorders⁶. We present a case associated with Rheumatoid Arthritis (R.A.).

Case Report

A 50 years old male teacher presented with a 3 years history of painful symmetrical arthropathy starting from hands and later involving wrists, elbows, knees and ankles. He received non-steroidal anti-inflammatory drugs which gave him relief initially but 20 months later he had a relapse. We saw him with a 3 week history of high grade fever, swelling, redness and pain in both ears and eyes and arthralgias in hands, feet, ankles and knees. The swelling of ears started simultaneously and involved the auricle sparing the lobule causing them to increase about two times their size. The ears were red, very painful and caused closure of the meatus affecting hearing. His eyes began to get red with excessive watering, got painful and his vision became blurred. Examination revealed a middle aged, heavily built male. He had a temperature of 100°F. His ears excluding the lobules were swollen red, warm and tender. There was periorbital edema and intense lacrimation with intensely congested sclerae in both eyes. The cornea were hazy and the pupils were small (0.2 mm) in size. The locomotor system revealed swollen warm and tender proximal interphalangeal and metacarpophalangeal joints bilaterally without any deformities. Examination of other system was normal. His hemoglobin was 11.9 Gms/dl, total; leucocyte count 7400 per cu mm and ESR 115 mm in the first hour. R.A. factor was positive in a titre of 1:160, although titres of up to 1:240 had been recorded previously. The anti-DNA and complement levels were normal. X-ray chest, ECG and Echocardiogram were normal. X-ray hands revealed periarticular osteoporosis. He was diagnosed as having relapsing polychondritis associated with rheumatoid arthritis and oral prednisolone in a dose of 1 mg/kg body weight was started with topical steroid eye drops. He responded well and steroids are currently being tapered.

Discussion

Less than 250 cases of this disease have been reported in the World literature⁷. Relapsing polychondritis occurs in the forty to sixty years age group with peak frequency in the fourth decade³. It may rarely affect children⁸ and the elderly². There is no familial tendency but an association with HLA-DR4 has been reported⁹. Diagnostic criteria have been proposed for this disease but are not universally accepted. The disease may cause valvular abnormalities, otitis media, glomerulonephritis, stroke, skin lesions, pulmonary infiltrates and small airways disease. None of these abnormalities had been detected in our patient. He does not have involvement of the nasal cartilage which occurs commonly in this disease. Relapsing polychondritis can cause collapse of the upper airways requiring intubation or tracheostomy¹⁰. The R.A. factor is occasionally present in low titre. In our case the titre was high due to co-existent rheumatoid arthritis. Relapsing Polychondritis may develop in association

with SLE Sjogren's syndrome, Wegener's granulomatosis and other vasculitides. McAdam et al report that approximately 30% cases of relapsing Polychondritis have a preceding or coexistent rheumatic disorder⁶. Steroids are the cornerstone of therapy and most patients require 40 to 60mg/day initially. In most cases steroids can be tapered and stopped but some may require long term treatment of up to 25 mg/day. A mortality of 22-30% has been described for this disease.

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