RELAPSING POLYCHONDRITIS
(Case Report)

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Summary

A case of relapsing polychondritis is reported which showed depression of nose and flattening of both pinnae. Erythrocyte sedimentation rate was high but rheumatoid factor was negative.

Relapsing polychondritis is a disease of unknown aetiology affecting the articular and non-articular cartilages.

It was described originally in 1923 by Jaksch-Wartenhorst[1]. It is a disease of middle age with slightly increased incidence in females. Dolan et al[2] stated that the course of this disease may be rapidly fulminant or a low grade smouldering one. Ear involvement is the commonest presentation. If nasal involvement is present, the bridge simply sinks without features of inflammation and this may occur startingly in a period of 2—3 days.

The associated arthropathies described are : simple arthralgia without joint destruction, dislocations[3] "arthritis mutilans" or a joint disease indistinguishable from rheumatoid arthritis[4]. Involvement of rib cartilages and larynx was described by Rowell et al[10].

Chest involvement is of grave importance. Recurrent respiratory tract infections resistant to antibiotics[5], pneumothorax, flail chest due to costochondral cartilage dissolution[3] and miliary tuberculosis have all been described.

Eye involvement in the form of episcleritis and iridocyclitis was described by Dolan et al[2]. Rucker et al[7], reported that severe keratitis, unilateral exophthalmos and lateral rectus muscle weakness may be associated with it. Occasionally, eyes and ears may show unilateral lesions[2].

Hainer et al[6] described the various visceral manifestations associated with this disease as myocarditis and aortic valvular insufficiency with aneurysmal dilatation of aorta. Recurrent thrombophlebitis was found in some cases by Rowell et al[10].

Skin lesions, inconsistently reported with this disease are alopecia[8], retarded nail growth, erythematous nodules[9] and increased incidence of psoriasis.

Elevated erythrocyte sedimentation rate has been consistently reported in these cases. Other biochemical features like increased urinary acid mucopolysaccharides, presence of rheumatoid factor, hypoalbuminaemia, increase or

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decrease in alpha-2 globulins, leukocytosis and elevated antistreptolysin "O" titre were found to be inconsistently present.

Verity et al[15] reported that death could occur due to tracheal and bronchial obstruction, necrotising broncheolitis, miliary tuberculosis or renal involvement.

The pathogenesis of this disease is still not clear. Feinerman et al[13] reported that proteolysis of the chondroitin sulphate-protein complex with release of acid mucopolysaccharides, may be the primary responsible factor in chondrolysis. Rogers et al[14] assumed that the chondrolysis takes place on the basis of an immune mechanism since indirect immunofluorescent testing revealed the presence of anti-cartilage antibodies in the serum of a patient with relapsing polychondritis. Rowell et al[10] reported the presence of gastric and thyroid antibodies, a positive rheumatoid arthritis latex fixation test and association with autoimmune thyroid disease, especially Hashimoto's thyroiditis.

Dolan et al[2] found that treatment with prednisolone initially with 30-60 mg/day followed by a maintenance dose of 5-10 mg was quite effective in patients with predominant respiratory symptoms. Long term use of salicylates was also found to be effective. Recently Martin et al[11] and Barranco et al[12] tried treatment with dapsone (100-200 mg/day), on the assumption that dapsone may be effective in certain immune related diseases because of its inhibitory action on the lysosomal enzymes.

Case Report

55 years old female, attended the Skin and V. D. Outpatient Department of Rajendra Hospital, Patiala, with a two year history of painful swellings of all joints, especially of lower limbs associated with facial puffiness after intake of telepaque tablets (for cholecystography). These symptoms subsided after sometime. Three months after the onset of the complaints she developed painful swellings of ears and nose which have been showing remissions and relapses since then.

Cholecystectomy was performed two years earlier for recurrent cholecystitis. There was no other significiant past history.

General physical examination revealed no abnormality except swelling of proximal interphalangeal joints of hands. No respiratory, cardiovascular or associated skin disease was found.

Eyes showed an alternating concomitant convergent squint.

Changes were present in ears and nose.

Ears

Both pinnae were flattened out. The various landmarks were ill defined and

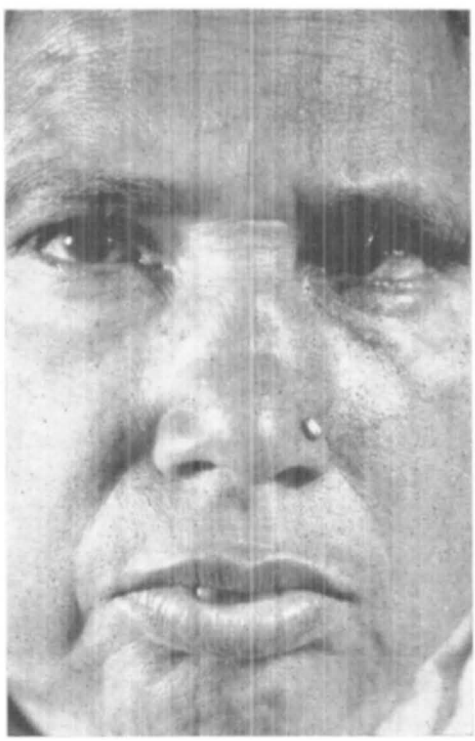

Fig. 1 Shows depression of cartilaginous portion of nose. Left eye shows convergent squint.

the external auditory meatuses were narrowed. On palpation there was thickening of cartilaginous portion. The lobules were flabby but freely mobile. Hearing was normal.

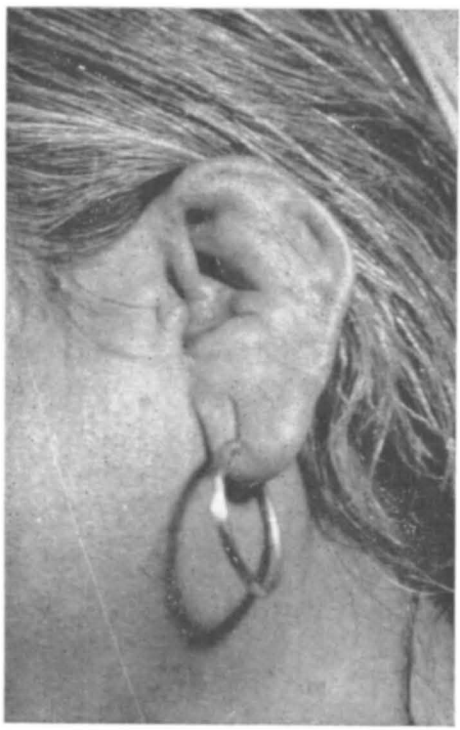

Fig. 2 Shows flattening of antihelix, antitragus, flattening of fossa of helix (scapha) and flabbiness of lobule of ear.

Nose

Depression at the junction of bone and nasal cartilage was seen. Skin over this region was mobile and nostrils were normal. Septum was irregularly depressed and flattened out.

Costochondral junctions were tender on deep palpation but there was no visible or palpable swelling.

Laboratory investigations : Hb. 13.0 5 per cent, TLC 6,400/cmm, DLC : P g3%, L 42%, E 3% and M 2%, ESR 65mm/1st hour Westergren, urine and stool examination — NAD, bleeding time 0.35", clotting time 3'-10", fasting blood sugar 60 mg%, blood urea 40mg%, L.E. cell phenomenon negative, rheumatoid factor negative. V.D.R.L. and Kahn tests were negative. Liver function tests showed a negative Vandenberg reaction, serum bilirubin 0.2 mg%, thymol turbidity 3 units, thymol flocculation nil and zinc turbidity 6 units.

Patient responded well to steroids and continues on a maintenance dose of 10 mg of prednisolone per day. There has been no relapse after starting therapy. Mild tenderness at the costochondral junctions persisted.

A case of relapsing polychondritis is reported which presented with acute generalised involvement of joints following intake of telepaque tablets. Patient was initially diagnosed to be suffering from rheumatoid arthritis but later developed polychondritis which resulted in flattening of ears and sinking of nose just below the nasal bridge. Laboratory investigations revealed no abnormality except for increased E. S. R. Rheumatoid factor was negative. The patient responded to steroids.

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