Cryptogenic cirrhosis in relapsing polychondritis

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Summary

A case of relapsing polychondritis with histologically proven cirrhosis is reported. This association has not been described before.

KEY WORDS: relapsing polychondritis, cirrhosis.

Introduction

Though the first documented case was described by Jaksch-Wartenhorst (1923), the name 'relapsing polychondritis' was suggested by Pearson, Kline and Newcomer as late as 1960. The extreme rarity of the condition and its possible confusion with other common conditions in Bangladesh, for example, leprosy, gout and pyogenic conditions of the pinna prompted us to report this case. In addition, the patient had cryptogenic cirrhosis, a previously unrecorded association.

Case report

The patient, a 37-year-old male cultivator, from Barisal about 200 km south of Dacca, reported to us with a history of recurrent attacks of pain and swelling of the auricle and nose associated with low grade pyrexia for the previous 2 years. Occasionally, the swelling of the auricle caused obliteration of the external auditory meatus. Subsequently, he noticed depression of the bridge of the nose. During the previous one year the pain became most distressing. There was also pain and swelling of costochondral and sternocostal joints. There was loss of weight, weakness and loss of hair during the previous 6 months. There was no history of jaundice, transfusion, or intake of any hepatotoxic drugs or alcohol.

On examination, the bridge of the nose was depressed and both auricles were hyperpigmented, slightly swollen and deformed. There was tenderness over the nose, auricle and costochondral joints. The liver was palpable 3 cm below the right costal margin

and the spleen was palpable 2 cm below the left costal margin. There was evidence of keratitis. Laboratory investigations showed haemoglobin 55%, white cell count $12 \times 10^9/l$ (polymorphs 74%), erythrocyte sedimentation rate 130 mm/hr, serum albumin 28 g/l, globulin 43 g/l, serum bilirubin and transaminases normal. Rheumatoid Factor, L.E. cells, hepatitis B surface antigen tests, and nasal scrapings for acid-fast bacilli were negative. Antismooth muscle and antimitochondrial antibody tests were not available. Liver biopsy showed evidence of macronodular cirrhosis. Right auricular cartilage biopsy showed chronic inflammatory cells, disintegration of cartilage and growth of fibrovascular tissue. The findings were considered consistent with the diagnosis of relapsing polychondritis.

The patient was put on oral prednisolone, 30 mg daily in divided doses. This was gradually reduced to 15 mg daily. The patient remained symptom free for about 4 weeks after which he developed diabetes mellitus requiring insulin.

Discussion

Relapsing polychondritis is an inflammatory disease of tissues that have a high glucosaminoglycan (mucopolysaccharide) content. Cartilage, bones, arterial tissues, sclera, cornea and inner ear components being rich in glucosaminoglycans, are prone to this disease.

McAdam *et al.* (1976) suggested 6 clinical diagnostic criteria. The presence of 3 or more of these together with histological evidence of chondritis confirms the diagnosis. The criteria are: (i) recurrent chondritis of auricles; (ii) non-erosive inflammatory polyarthritides; (iii) chondritis of nasal cartilages; (iv) inflammation of ocular structures including conjunctivitis, keratitis, scleritis or episcleritis and uveitis; (v) chondritis of respiratory tract affecting laryngeal or tracheal cartilages or both; and (vi) damage to

cochlea, vestibule or both, manifest by neurosensory hearing loss and tinnitus or vertigo or both. The case report here fulfils the criteria for a diagnosis.

Approximately 30% of patients may be associated with rheumatic or autoimmune diseases. The rheumatic diseases are of seronegative type while the autoimmune diseases include thyroid disease, ulcerative colitis, glomerulonephritis, dysgammaglobinaemias and non-caseating granulomas (McAdam *et al.*, 1976). Minor liver function abnormalities have been described in relapsing polychondritis (Herman, 1981), but not, to our knowledge, actual liver disease.

The absence of other aetiological factors such as viral hepatitis does not, of course, exclude their possible role in the development of the cirrhosis, but we feel that the presence of cirrhosis in our case may be yet another example of autoimmune disorder associated with relapsing polychondritis.

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