

**RELAPSING POLYCHONDritis WITH ERYTHEMA NODOSUM LIKE LESIONS - A
CASE REPORT****Dr. Rowan Monteiro***

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INTRODUCTION

A 21 year old male presented with a history of episodic, polyarticular and migratory joint pains over a period of 1 year. Four months later, he developed swelling of both his ears simultaneously, which was initially painful, but subsequently became painless (See Fig 1 and 2). The swelling involved the entire ear, sparing the ear lobe. In addition two months after the ear swelling, he developed firm skin colored Erythema nodosum like nodules over the shins, which were symmetrical in distribution (See Fig 3).

On examination, there was no evidence of hoarseness, dyspnoea, photophobia, hearing loss, ataxia and bony deformities. Investigations revealed a mild derangement of liver enzymes, while Rheumatoid factor and CRP were positive. Biopsy of the left ear pinna and leg nodules were done. Histopathology findings were similar from both sites showing a dense lymphoplasmacytic infiltrate, rich in histiocytes and plasma cells and associated with myxoid changes. Additionally, the biopsy from the ear pinna showed a complete absence of cartilage (See Fig 4).

McAdams criteria (Table 1)^[1] and Damiani and Levine criteria (Table 2)^[2] were used to aid the diagnosis as the patient had bilateral auricular chondritis and non-erosive inflammatory polyarthritis and was diagnosed as a case of relapsing polychondritis. No evidence of nasal or respiratory chondritis, ocular inflammation and audiovestibular damage were present after detailed evaluation.

The patient was started on a combination of corticosteroids and methotrexate and his joint symptoms and erythema nodosum like soft tissue swellings over the lower limbs decreased, but his ear swelling remained status quo. He was continued on the same treatment for a period of 3 months during which his symptoms were well controlled. He stopped taking the medication for two months, which lead to a flareup of his joint pains. He was restarted on the previous regimen but was subsequently lost to followup.

Relapsing polychondritis is a rare multisystem autoimmune disease with both autoantibodies and cellular immune reactions to collagen type II, present in hyaline and sometimes elastic cartilage. Usually

manifests between the third to fifth decade with a male to female ratio of 1:3.^[3] It gained its name of relapsing polychondritis due to its chronic relapsing and remitting nature (which was not the case in our patient) as well as progressive destruction of cartilage, which is seen in the end stage of the disease. It is regarded as an orphan disease and because of the rarity of the disease clinical series with only a finite number of patients with anecdotal therapy being the key to patient management. Systemic features are common and life threatening complications such as laryngotracheal strictures and obstruction being reported in 55% cases^[4] The mean delay between symptom onset until diagnosis is about 2.9 years,^[3] mostly owing to the slow progress, chronicity and multisystem involvement of the disease, which inturn makes identifying and diagnosing a case of relapsing polychondritis more challenging and difficult, as was the case for Patel et al.^[5]

An atypical feature that was noted were the extraarticular nodules located symmetrically over both shins. Such nodules have been reported in literature and have been described as erythema nodosum like lesions with histopathology features varying from septal panniculitis to thrombosis and vasculitis of skin vessels, however in our patient it showed a similar image to the ear, probably indicating the presence of a similar pathological process occurring in both the ear and the legs. The frequency of dermatological manifestations in patients who have "pure RP with no associated diseases" is around 35% and includes oral aphthosis, livedo reticularis, purpura, urticated papules, limb ulcerations, superficial phlebitis and limb ulcerations.^[6]

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