Otological Manifestations of Relapsing Polychondritis

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Citation

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Abstract

A rare disorder of unknown etiology “relapsing polychondritis” affecting head neck region was presented with literature review and management. Relapsing polychondritis is a rare disorder associated with high morbidity and mortality. The management depends up on early diagnosis and oral corticosteroids with or without other immunosuppressant.

INTRODUCTION

Relapsing Polychondritis is a rare systemic inflammatory disorder of unknown etiology first described by Jacksch-Warten horst in 1923 and characterized by an inflammatory reaction occurring in the cartilages of several different organs, the commonest being the auricular cartilage.

All types of cartilages, fibro cartilage, elastic, hyaline cartilage and cartilage in tracheo bronchial tree and other proteoglycan-rich structures such as eye, heart, blood vessels and inner ear may be involved. Constitutional symptoms are common and vasculitis affecting skin or internal organs (heart, kidneys, nervous system) may occur. Patients may be presented with symptoms that often confuse the physicians and it may be major diagnostic dilemma[4].

Hence we describe the clinical presentation and management of a case that mislead us initially and eventually the diagnosis was made on basis of the modified criteria by Damiani and Levine. (Table I)

Figure 1

Table 1: Criteria for diagnosis of relapsing polychondritis as suggested by McAdam/Michet (,) with Damiani-Levine () modification

Bilateral auricular chondritis
Nonerosive seronegative inflammatory polyarthritis
Nasal Chondritis
Ocular inflammation-Conjuctivitis, Keratitis, sceleritis/episceleritis, Uveitis
Respiratory chondritis-Laryngeal and /or tracheal cartilages
Cochlear and /or vestibular dysfunction-neurosensory hearing loss, tinnitus and /or vertigo
To establish the diagnosis, all patients were required to have one of the following:
At least three of the above clinical criteria
One or more of the above clinical criteria with positive histological confirmation
Chondritis at two or more separate anatomical locations with response to steroids and /or dapsone

CASE REPORT

A 45-year-old Malay man presented with a painful swelling of his left pinna, which has progressively worsened over one week. He had concomitant malaise. There was no past history of Meniere's disease or rheumatic fever.

On examination his left pinna was erythematous, swollen and tender on palpation. There was no evidence of mastoid tenderness. The facial nerve was intact. External ear canal and the tympanic membranes were normal on both sides. The hearing was normal in both ears. Hence diagnosis of left ear perichondritis was made and he was treated with Intravenous cefuroxime 750 mg three times daily and oral paracetamol. After three days he was discharged with oral antibiotics with some improvement.

After one week he presented to our department with history of vertigo and vomiting and decrease left hearing for one-

day duration. Clinical examination revealed nystagmus to the right. The left pinna was erythematous and tender on palpation. Both eyes were congested and diagnosis of bilateral scleritis was made by the Ophthalmologist. Tuning fork test confirmed senosorineural hearing loss on left. Hence the provisional diagnosis of sudden sensory neural hearing loss secondary to acute labyrinthitis was made and treated with Intravenous ciprofloxacin 200 mg twice daily, Indomethacin and beta histine 16 mg three times daily. Initial blood investigations revealed a normal full, but ESR was 79mm/ hour. Rheumatoid factor was negative. Anti nuclear antibody was negative. The renal profile and thyroid function test were normal. Initial pure tone audiogram performed on admission day showed left sided severe to profound sensori neural hearing loss

One-week post admission the right pinna was erythematous and tender on palpation. The ESR was raised to 130mm/ hour and audiogram showed bilateral sensori neural hearing loss more on left. His clinical features and blood ESR reading suggest the final diagnosis of relapsing polychondritis as the patient fulfilled the diagnostic criteria of Mcadam-Michet-Damiani-Levine[1, 2, 3]. Finally the patient was treated with oral prednisolone 60 mg per day and the dose was tapered over 2 weeks to 30 mg per day. The patient symptomatically improved and pure tone audiogram showed some response on left but very little response on right. He is still on our regular follow up at our clinic.

Figure 2

Figure 1: Left pinna was erythematous, swollen and tender on palpation

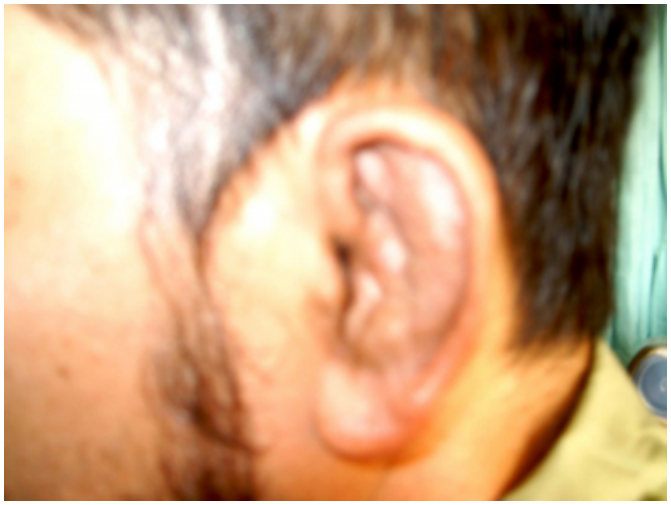
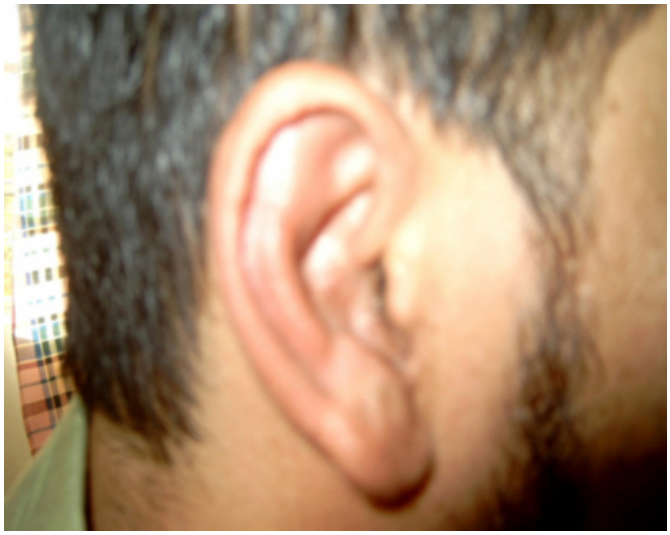


Figure 3

Figure 2: Right pinna was erythematous and tender on palpation.



DISCUSSION

The above case was diagnosed after a month as ‘Relapsing polychondritis' which is shorter than that of reported in the series of Trentham et al. Relapsing polychondritis can present with variety of clinical features and pose a diagnostic problem. Auricular Chondritis is the common presenting feature observed in 90 per cent of the cases. It is typically bilateral but it may be unilateral with sudden onset. Pinna will be erythematous in appearance without any purulent discharge. The other otological feature may include secretory otitis media due to involvement of the Eustachian tube cartilage. Sensorineural or mixed deafness, which may be bilateral or unilateral, which may be sudden or progressive over the weeks[6]. Associated vestibular symptoms include vertigo and vomiting. Our patient presented with above otological features except secretory otitis media. The other clinical features[5] of Relapsing polychondritis include: Arthropathy, Nasal Chondritis, and Chondritis of respiratory system, which may some times present with airway complications. Relapsing polychondritis may involve the cardiovascular system resulting in aortic incompetence, mitral regurgitation, pericarditis, cardiac ischemia, and anerysms of large arteries, vasculitis of central nervous system, Phlebitis and Raynaud's phenomenon.

Skin involvement causes cutaneous vasculitis and non-specific eruptions, which are absent in our patient. Commonest manifestation of eye includes episcleritis, which was present in our patient. Other eye presentations include keratitis, sceleritis, and uveitis. The commonest laboratory finding is raised ESR which follows the progress of disease.

Rarely neurological and renal systems will be involved. Medical treatment of relapsing polychondritis consists primarily of cortico steroids, immunosuppressive drugs and dapsone[5]. Prednisolone is the drug of choice. It reduces the inflammatory response by stabilizing lysozymes that come from the leucocytes with some evidence of recovery of hearing in patients with early sensorineural hearing involvement. Symptoms tend to relapses when the drug is discontinued or dose is markedly reduced.

CONCLUSION

Relapsing polychondritis is a rare condition and can be easily misdiagnosed. Lethal complications are often associated with airway or cardiovascular involvement. Treatments include oral cortico steroids and supportive therapy. A larger patient group is needed to provide more insight in clinical presentations and further management strategy to this condition amongst the Malaysian population.

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