Relapsing Polychondritis: 
A Rare Disease of Multisystem Involvement

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ABSTRACT
Relapsing Polychondritis (RP) is a rare connective tissue disease of unclear pathogenesis and may present with multisystem involvement. In this report we describe a case of Relapsing Polychondritis, a rare autoimmune disease of varied presentation, course, and response to therapy.

Key words: Relapsing polychondritis, autoimmune disease

INTRODUCTION
Relapsing polychondritis (RP) is a rare connective tissue disease of unknown aetiology. This disease primarily involves cartilaginous structures throughout the body: ears, joints, nose, eyes, laryngotracheobronchial tree, and costal cartilages are mainly affected. Apart from inflammation of these structures, it is associated with systemic vasculitis and various autoimmune diseases. Diagnosis is based upon the clinical manifestations of multiple organ involvement supported by histological findings in doubtful cases. Treatment involves use of anti-inflammatory agents (nonsteroidal and steroids); immunosuppressive agents are useful in severe cases. Here, we describe a case report of a young man with Relapsing polychondritis who had severe respiratory and other-organ involvement.

CASE REPORT
A 20-year old boy from Sarlahi was admitted to the medical ward of Patan Hospital with ear, eye, and throat involvement of one-year duration. Initially, he had painful swelling of the right auricle which, after a few days, became blackish. Three months later, the left ear was similarly affected. The same process recurred in both ears again in six months, this time leaving his ears soft and flabby.

Shortly after the first episode of ear involvement, he had pain and redness of the eyes along with blurring of vision. These symptoms resolved with a topical steroid drop from a local pharmacy. However, the eye problem recurred and his eyes were still red and painful at the time of admission.

Six months before admission, this patient had an episode of dry cough, sore throat, hoarse voice, and difficulty swallowing; these symptoms remitted partially with the use of systemic steroid given by the local medical shop. He claims, however, that those symptoms were persistent even at the time of his being admitted in our hospital. In addition, he also reported that he was having recurrent painful swelling of joints and stiffness in all four limbs for the last few months.

A few days prior to admission he noticed pain and swelling over the nasal bridge; this part later collapsed giving an appearance of a saddle nose.

Review of the cardiovascular, gastrointestinal, and nervous systems revealed no significant problems. Similarly, he had no remarkable medical illnesses in the past, including no allergy to drugs.

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intravenous steroids in the ICU. Extubation was accomplished after 3 days. However, he continued to experience recurrent episodes of brief, acute respiratory distress that were accompanied by mild stridor. Direct laryngoscopy after extubation confirmed previous findings of collapse of periglottic structure; there was near occlusion of the glottis during inspiration, which opened up during expiration. Bronchoscopy could not be carried out because of the narrowed glottis and the potential risk of acute respiratory distress.

Over the next two days, the patient continued to experience intermittent respiratory distress. Because the anatomic defect seemed not to be responding to steroids, the patient was taken to the OR for tracheostomy. This was attempted under local anesthesia, during which the patient was restless, the procedure was difficult, and the patient arrested. He could not be resuscitated and died.

DISCUSSION

Relapsing polychondritis (RP) is a rare connective tissue disease characterized by an episodic and progressive inflammation and subsequent destruction of the cartilage of various parts of the body. It can affect people of all ages and both sexes, and is associated with many rheumatic and nonrheumatic diseases.

The exact cause of RP is uncertain but various genetic, immunological, and environmental factors may be implicated in its pathogenesis. The disease starts with acute inflammation of the cartilaginous tissues of the external ear, nose, tracheobronchial tree and sclera. Repeated inflammation in the same areas results in destruction of the cartilage with subsequent fibrosis. Ultimately the cartilage loses structural integrity and assumes a distorted shape: the ear may flop or become cauliflower-like and the nose saddle-shaped.

As in our patient, RP may go unrecognized for several months particularly in patients who present with intermittent symptoms. The diagnosis was delayed for about one year in our patient. The pattern of cartilage involvement and the frequency of episodes also vary widely among patients.
Recurrent inflammation of the auricular cartilage is the most common manifestation; it may present as an acutely inflamed pinna, distorted external ear, hearing loss, or vestibular impairment. Asymmetrical, non-erosive arthritis of both small and large joints occurs in three-fourths of patients which usually resolves spontaneously without any residual joint deformity. Upper respiratory tract cartilage is affected in 50% of cases and may present as saddle nose, dry cough, hoarseness, stridor, and breathlessness. Sometimes patients develop life-threatening airway obstruction necessitating intubation, and later tracheostomy. Our patient also needed an emergency intubation during the hospitalization. Ocular inflammation results in episcleritis, scleritis, uveitis and even blindness. Some patients develop neurologic features like stroke, seizures, and various cranial nerve palsies due to vasculitis. The heart (aortic regurgitation, aortic aneurysm, pericarditis, heart blocks), kidneys (glomerulonephritis), and skin (erythema nodosum) are also affected in some cases.

Laboratory abnormalities in RP are nonspecific and simply reflect chronic inflammatory process. Diagnosis is thus made on the basis of typical clinical manifestations. McAdam and colleagues proposed a clinical criteria consisting of six features (See Box). Later, Damiani and Levine modified these slightly and suggested that the diagnosis could be made with:

- 3 or more of these criteria
- at least one clinical criterion plus histological confirmation
- chondritis in two or more separate anatomical locations with a response to treatment.

However, a histological diagnosis is not necessary in most patients with clinically evident disease.

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<tr>
<th>Box : McAdam et al. criteria for diagnosis of RP</th>
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<tr>
<td>1. Recurrent chondritis of both auricles</td>
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<td>2. Non-erosive polyarthritis</td>
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<td>3. Chondritis of the nasal cartilage</td>
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<td>4. Ocular inflammation including conjunctivitis, keratitis, scleritis/ episcleritis, uveitis</td>
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<td>5. Involvement of laryngeal and/or tracheal cartilage</td>
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<td>6. Cochlear and/or vestibular involvement</td>
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Our patient had almost all of these criteria except features denoting inner ear involvement. He had floppy ears, due to repeated episodes of auricular chondritis, asymmetric polyarthritis, tender nasal cartilages with depressed bridge, scleritis, and – above all – life-threatening laryngotracheal involvement.

Delays in diagnosis, multiorgan involvement and the unpredictability of recurrences all pose significant challenges in the management of RP. Treatment involves nonsteroidal anti-inflammatory drugs, steroids, and dapsone for active chondritis, and immunosuppressants like methotrexate and cyclophosphamide for severe forms of disease. Supportive measures include tracheostomy, tracheal stents, hearing aids, etc. The prognosis is somewhat worse in patients with more widespread disease. We highlight here our patient’s precipitous and unexpected demise as a warning to others managing this challenging disease.

REFERENCES

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