Relapsing Polychondritis: A Case Report

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Relapsing polychondritis is a chronic, recurrent, episodic, inflammatory disease of unknown cause affecting predominantly the cartilaginous tissues of the body. Episodes last few days to weeks. All types of cartilages may be involved. The most common presentation is the involvement of pinnae cartilages. The term was coined by Pearson and was first described by Jacksch-Wartenhorst in 1923, who called it polychondropathies. Annual incidence is about 3.5/million with the peak age at onset ranging from 40 to 50 years. Autoimmune etiology is suspected. Diagnosis is based on McAdam criterion. Corticosteroid remains the mainstay of treatment along with the anti-inflammatory drugs. Here we present a case of 18-year-old male who presented with the involvement of the laryngeal cartilages, which is a rare presentation and later there was an involvement of other cartilages too. Our patient had four of McAdams criteria and also responded to steroids.

Keywords: Chondritis, Relapsing polychondritis, Stenosis, Stridor

INTRODUCTION

Relapsing polychondritis is a rare multisystemic disease with an unknown etiology, characterized by the recurrent inflammation of cartilage tissues, and may lead to their destruction. It affects men and women equally and is more prevalent in Caucasians. It is manifested mainly in the fifth and sixth decades of life. Its cause is unknown, certain genetic predisposition and an association to human leukocyte antigen-DR4 has been observed.¹

The diagnosis is established according to MacAdams criterion. No specific histopathologic finding is considered pathognomonic for relapsing polychondritis. When the clinical picture is clear, biopsy is unnecessary.²

Mild auricular involvement can be treated with non-steroidal anti-inflammatory agents and/or low dose steroids. In severe disease, prednisone in the dosage of 1 mg/kg is administered. Immunosuppression with methotrexate, azathioprine, cyclosporine, and cyclophosphamide may be needed in patients who do not respond to steroids. Surgical intervention may be needed in case of complications involving the respiratory tract like tracheal stenosis and tracheomalacia and stents may be required for tracheobronchial collapse.³

5-year survival of up to 74% and 10-year survival of 55% have been reported in a study of 112 patients from Mayo clinic, major causes of death being infections, and systemic vasculitis.³

CASE REPORT

A 18-year-old male came to our department with complaints of cough sore throat and noisy breathing since 2 months. He was febrile and his speech was normal. Laryngeal tenderness was present, fiberoptic laryngoscopy revealed both the vocal cords in paramedian position with 3-4 mm glottic chink (FLP) (Figure 1). Complete abduction was absent on deep inspiration, but adduction was present.

Blood investigations showed hemoglobin to be 10.5 g%; erythrocyte sedimentation rate was 55 mm/h, the white blood cell count was 14,000 cells/cumm, and platelet count was 4.6 lakhs/cumm. Rheumatoid factor and antinuclear antibody were negative, but a raise in anti-collagen-II antibody was seen.

Pulmonary function test suggested upper airway obstruction. Bronchoscopy revealed a definite subglottic narrowing with normal mucosa. Bronchial washings were negative for acid-fast bacilli. Tracheostomy was done in view of narrow glottic
chink and patient having difficulty in breathing. At the same time biopsy from the narrowed subglottic area was taken, which was reported as chronic inflammation with fibrosis.

During regular follow-ups, he came with complaints of the hardness of hearing, pain, and swelling of both pinna and swelling of the nasal dorsum.

On examination, the cartilaginous part of both pinnae were reddened and swollen with sparing of the lobules (Figure 2a and b).

Dorsum of the nose was swollen with septal abscess for which incision and drainage were done which later healed with supra-tip depression (Figure 2c). Furthermore, there was synchiae between both the vocal cords on the fiber optic laryngopharyngoscopy done as a part of the routine follow-up, due to some surgery done over vocal cords elsewhere.

It was then that the diagnosis of relapsing polychondritis was opined according to McAdam et al. diagnostic criteria. Patient was referred to a rheumatologist for further management. He was prescribed prednisolone (1 mg/kg/day) and methotrexate (15 mg/week). Later the dose of prednisolone was tapered to 5 mg/day.

Patient underwent laser cordotomy elsewhere and came back for review of vocal cord status. FLP was done, which showed worsening of the situation with glottic chink of <2 mm with synchiae between both vocal cords (Figure 2d). Patient is still on tracheostomy with low-dose steroids (prednisolone 2.5 mg on alternate days) and methotrexate (10 mg/week).

**DISCUSSION**

Relapsing polychondritis was coined by Pearson and was first described by Jacksch-Wartenhorst in 1923, who called it polychondropathies. Annual incidence is about 3.5/million with Peak age at onset ranging from 40 to 50 years.

Autoimmune etiology is suspected as antibodies to collagen Type II in the acute phase can be detected. The course is characterized by episodes of inflammation and fibrosis that destroy all types of cartilage especially those with high concentrations of glycosaminoglycans.

According to McAdam et al., diagnostic signs (1976) presence of three or more of the following are necessary for diagnosis of relapsing polychondritis:

1. Recurrent chondritis of both auricles
2. Non-erosive inflammatory polyarthritis
3. Chondritis of nasal cartilages
4. Inflammation of ocular structures, including conjunctivitis, keratitis, scleritis/episcleritis, and/or uveitis
5. Chondritis of the respiratory tract involving laryngeal and/or tracheal cartilages-HOV, tenderness, airway obstruction
6. Cochlear and/or vestibular damage manifested by sensorineural hearing loss, tinnitus, and/or vertigo
7. Cartilage biopsy confirmation of a compatible histological picture.

Damiani and Levine (1979) modified it to following criteria:

- Three or more of McAdam et al. – No histological confirmation needed
- One or more of McAdam et al. – Histological confirmation needed
- Involvement of two or more separate anatomic locations with the response to steroid and/dapsone.

Our case had four of McAdams criteria and also responded to steroids.
The most common presentation is involvement of pinnae (89%), which is also a presenting symptom in 1/3 rd of patients. Hearing loss is seen in 36.3%, dysphonia in 36.3%, nasal chondritis in 26.6%, and approximately 50% of patients have larynx and trachea involvement.

Corticosteroid remains the mainstay of treatment. Overall survival rates were 74% at 5 years and 55% at 10 years. About 15% of the deaths were as a direct result of cardiovascular or respiratory tract involvement.

CONCLUSION

Relapsing polychondritis is a destructive disorder of cartilages, with the most common presentation being the involvement of pinnae cartilages and in our case presented with cricoarytenoid ankylosis with noisy breathing. The mainstay of treatment is anti-inflammatory agents.

REFERENCES