International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Relapsing Polychondritis-Early Diagnosis and Treatment'

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Abstract: ***Relapsing polychondritis is a infrequent immune mediated systemic disease most commonly presenting as inflammation of the cartilage of the ears and nose. It can also affects the airway if involved laryngeal cartilage, then may result as a drastic complication if not timely treated. Patient with auricular and nasal involvement usually pesented as outdoor patient in the otorhinlaryngology department with painful, swollen and erythematous external ear (with sparing of ear lobules) and nose. Intialtreament with oral antibiotics and nonsteroidalanti inflammatory drugs does not result complete recovery. Patient responds to oral steriod. Oral steriod given for longer period followed by the use of steriod sparing drugs like methotrexate has been given in weekly doses along with folic acid in our case. Early diagnosis help in the prevention of developing permanent deformities like cauliflower ear, saddle nose.***

Keywords: Relapsing polychondritis, saddle nose deformity, autoimmune disorder, immunosuppressive drugs

1. Introduction

Relapsing polychondritis (RP) is a infrequent rheumatologic disorder in which repeated episodes of inflammation result in damage of cartilage of the ears and nose. The joints, eyes, audio vestibular system, cardiovascular system, and respiratory tract can also be involved.[1] It is characterized by recurrent inflammation of the cartilaginous structures and proteogly can-richorgans.[2] Relapsing polychondritis (RP) is a multisystem disorder of cartilaginous tissues.[3] RP Iis classified as a systemic vasculitis in the past.[4] it can cause the permanent destruction of cartilage if not timely treated. it rarity usually results in delayed diagnosis.[6]

Jaksch-Wartenhorst had apparently described it as polychondropathia and was later as chondromalacia, chronic atrophic polychondritis. It was Pearson et al in 1960 who named it as relapsing polychondritis. it commonly occurs between the ages of 20 and 60 years, with a peak in the 40s. male to female ratio is equal but Trentham et al reported afemale-male ratio of 3:1.1

First clinical diagnostic criteria for RP was proposed by, McAdam et al, then later modified by Damiani and Levineand Michet et al. The estimated incidence is 3.5 per 1, 000, 000 per year in UK. There were Diagnostic criteria of relapsing polychondritis, according to different authors.
Mc Adam et al.1976

At least three clinical features among auricular chondritis, nonerosive inflammatory polyarthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis, audiovestibular damage; histologic confirmation not required Damiani and Levine 1979

At least one of the six clinical features suggested by Mc Adam et al. plus histological confirmation or two of the six clinical features suggested by Mc Adam et al. plus positive response to administration of corticosteroids or dapsone
Michet et al.1986

Confirmed inflammation in two of three cartilages among auricular, nasal or laryngotracheal or proven inflammation in one of the above cartilages plus two other minor criteria among hearing loss, ocular inflammation, vestibulary disfunction, seronegative arthritis.5

It is a diagnosis of exclusion and early diagnosis can be made by the most common presenting feature of auricular chondritis.1

There are many treatment option available for this like nonsteroidal anti-inflammatory drugs, corticosteroids, dapsone, azathioprine, cyclosporine, cyclophosphamide, and methotrexate. Some cases do not respond to conventional therapy and may cause chronicity, irreversibility, and finally death. Biological therapy can be used in these cases that result in good disease control and improved quality of life. oral steroid followed by one of immunosupressant is good for limited disease.7

2. A Case Report

A 57 year old women came in the department of otohinolaryngology at Deendayal Upadhya zonal hospital Shimla as outdoor patient on march 28, 2022. she presented with the complaint of pain and redness in nose and both pinna (right>>left) for a week. She had generalized weakness, loss of appetite, decreased sleep because of pain. She took treatment for joint pain from some local quack in the past. She had no history of ear discharge and no history of ear trauma with no history of insect bites, fever, cough, and runny nose. She had no history of diabetes mellitus, hypertension, asthma.

On physical examination, the generalist status results were within normal limits. The erythema over the tip of nose and the nasal vestibule was present. It was tender. There were

swollen and erythematous external ears with sparing of ear lobules (right>>left). The examination of the right and left ear canal and ear drum was found within normal limits. Blood and internal medicine tests were within normal limits.

On 28 March, we prescribed her a course of antibiotics, antiinflammatory along with adequate hydration for a week. She showed sign of improvement within a week. The erythema over the nose and left pinna subsided. Only mild erythema over the right pinna was present. We continued same treatment for 3 more days.

After 15 days she again presented with redness and pain but only in the right external ear with sparing of ear lobule. We prescribed the tapering doses of steriod for two weeks along with antiinflammatory drugs twice a day for three days and then SoS. After two weeks she was improved. We stopped the treatment. She again had recurrence in the right ear. We made a diagnosis of Relapsing Polychondritis. We asked her continue steriod for one more month in tapering doses after getting the blood test. After one month we had put the patient on oral methotrexate 20mg in a weekly dose along with folic acid 5mg in two divided doses for three month. Follow up was done after every month, patient was improved now. Repeat liver function test and complete heamogram was within normal.

3. Discussion

It is a infrequent inflammatory disorder results in repeated inflammation of cartilages. It can affects mainly cartilages of ear and nose but can also involve the tracheobronchial tree.6It is multisystem disease that can also affect the cardiovascular system, central nervous system, renal system, involvement of ocular system, joints.3 If there is involvement of laryngeal and tracheal cartilage rings that results in delayed diagnosis because of rarity of disease. If it is not treated then there will be the damage of cartilage and finally collapse of airway.[6]

Patient usually presents with red ear in 20-41%. There can be one or both ear can be involved. There is involvement of pinna with sparing of ear lobules. The nasal cartilage involvement incidence varies from one region to another. There is main morbidity and mortality if airway involved in the relapsing polychondritis. Its prevalence is 50%.[2]

There are many treatment option available for this like nonsteroidal anti-inflammatory drugs, corticosteroids, dapsone, azathioprine, cyclosporine, cyclophosphamide, and methotrexate.[7]

We have reported a case of relapsing polychondritis with limited disease. We have examined the patient in detail. As patient presented in department of otorhinolaryngology in our hospital with bilateral ear involvement with sparing of ear lobules and nose, with complaint of multiple joint pains in the past three months. Initially we prescribed the course of oral antibiotic and antiinflammatory for a week but not improved. Later we have given oral steroid for a month in tapering doses for a month followed by oral methotrexate. Patient was improved and continued on oral methotrexate for 6 month.

We have found early diagnosis helped in lesser relapse of disease. It can be treated by oral steroid and steroid sparing drugs for maintenance.

4. Conclusion

Relapsing polychondritis is an autoimmune disorder having variable presentation. It can affect the cartilage of body anywhere most common involvement auricle and nasal cartilages. Early diagnosis of disease helps in avoiding the associated complication of disease without permanent sequele like cauliflower ear and saddle nose deformity in the otorhinolaryngology department. Patient can be treated with the tapering doses of steroid followed by steriod sparing drug methotrexate in our case. Average duration of treatment varies 3 months to one year.

Acknowledgment

We would like to thank the patient who had agreed to have her case reported.

Declaration of patient consent

We certify that w have obtained all appropriate patient consent form. In the form the patient has consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil

Conflicts of interest

There are no conflicts of interest.

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