International Journal of Health Sciences and Research
Vol.13; Issue: 12; December 2023
Website: www.ijhsr.org
Case Report *ISSN: 2249-9571*

Relapsing Polychondritis Presenting with Cauliflower Ears and Saddle Nose Deformity in a Patient of Syphilis Incognito: Two Rare Diseases in One Patient

Kaul Neenu[1], Kumari Neeti[2]

[1]Assistant Professor, Department of Dermatology, Shri Guru Ram Rai Institute of Medical and Health Sciences, Dehradun,Uttrakhand India.
[2]Professor, Department of Dermatology, Shri Guru Ram Rai Institute of Medical and Health Sciences, Dehradun,Uttrakhand India.

Corresponding Author: Dr Neenu Kaul

DOI: https://doi.org/10.52403/ijhsr.20231231

ABSTRACT

Relapsing Polychondritis (RP) is a rare immune-mediated systemic disease. The etiology of RP is unknown but both infective and non-infective causes have been suggested to play a role in causation. It manifests as recurrent episodes of pain, redness and swelling of the cartilaginous parts of body commonly involving the auricular and nasal cartilage.

Keywords: Relapsing Polychondritis, chondritis, saddle nose, syphilis, prednisolone.

INTRODUCTION

Relapsing Polychondritis (RP) is a rare clinical entity and affects up to 3.5 in one million people per year. It is characterized by progressive inflammation in cartilage tissue. It commonly affects the ear, nose, respiratory tract. Other tissues that may be affected are blood vessels, proteoglycan rich-eyes and heart. [1] We report a case of relapsing polychondritis with cauliflower ear and saddle nose deformity for its rarity. The etiopathogenesis and the management of RP is also discussed.

CLINICAL AND IMAGING FINDINGS

A 48-year-old female presented at our clinic with recurrent episodes of pain, redness and swelling in both ears since eighteen months. Recurrent episodes of inflammation resulted in cauliflower like deformity in right ear. Similarly saddle nose deformity was noticed twelve months ago following two episodes of redness and pain over the bridge of nose. There was history of joint pains with redness and swelling in bilateral ankle and wrist joints for ten months. The symptoms started insidiously and there were recurrent episodes occurring over a period of two to three weeks followed by spontaneous resolution. The patient also took consultation from ear, nose and throat clinic as well as rheumatology clinic and was prescribed pain killers and antibiotic treatment for the same which did not provide much relief to the patient. There was no history of genital or oral ulceration in the past. The patient did not give history of any skin lesion. There was no history of ear or nose piercing, assault and iatrogenic injuries. The patient as well as her partner was prescribed intramuscular benzathine penicillin after their VDRL AND TPHA came reactive. However even after six months of penicillin treatment, the RP

didn't improve and dramatic response was seen within one week of starting 30 mg of oral corticosteroids

Cutaneous examination:

Local examination of ear revealed mild erythema, induration and thickening of bilateral auricles; cauliflower like deformity of right ear with sparing of both ear lobes as visualised in the image (Fig 1,2). On palpation there was mild tenderness in the auricles. Saddle nose deformity of nose was present (Fig 3). Rest of cutaneous, mucosal and systemic examination was normal. Nerve examination was normal.

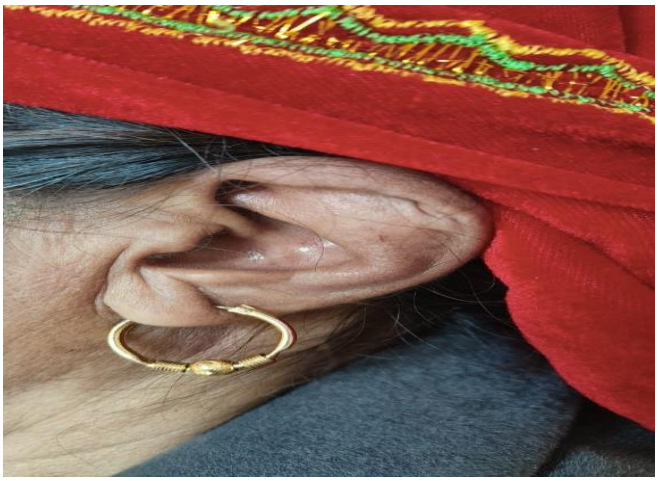

image 1

image 2

image 3

Laboratory investigation:

Baseline investigations like complete blood count, liver function tests, renal function tests, blood sugars were within normal range. ESR and CRP were mildly raised. Other clinical parameters like urinalysis, thyroid tests, ACE levels were within normal range.

Antinuclear antibodies (ANA), Antiphospholipid antibodies, Anti Neutrophil Cytoplasmic Antibodies (ANCA), Rheumatoid factor, Hepatitis B, C and HIV tests were non-reactive.

DISCUSSION

RP is a rare clinical entity with insidious onset and characterized by progressive inflammation in cartilaginous and proteoglycan rich tissues. Disease has an acute inflammatory phase which is followed by spontaneous remission of variable duration. With repeated attacks there may be severe cartilage damage which is replaced by fibrous tissue leading to permanent deformities like floppy ear or cauliflower like ear, saddle nose and life-threatening sequelae particularly if cartilage of tracheobronchial tree is involved. Chondritis and polyarthritis represent the most common clinical presentation of the disease. [2]

The exact pathogenesis of RP is unknown; however, it is suggested that both humoral and cell-mediated immune systems are involved. Cartilage specific autoimmunity and therefore circulating autoantibodies against collagens II, IX, XI may play an important role in the pathogenesis of RP. Association with other autoimmune diseases is found in 30% of all RP patients, rheumatoid arthritis (RA) being the most common.[3,4,5]

Factors like infections, trauma, autoimmune or inflammatory diseases such as ulcerative colitis, Behcet's disease and myelodysplastic syndromes may cause the release of degraded cartilage antigens and hence can initiate the disease process. [6]

There are no specific laboratory tests, histopathological features or imaging which are diagnostic of RP. However in 1979, Damiani sets new diagnostic criteria, presence of one of the events: bilateral auricular chondritis, nasal chondritis, audio vestibular affection, seronegative arthritis, ocular inflammation and airway chondritis with relevant histopathology or two clinical symptoms with favorable response to prednisolone. [7]

There are no specific guidelines for the treatment of RP. Non severe cases can be managed with non-steroidal anti-inflammatory (NSAIDs) drugs, dapsone and colchicine. NSAIDs resistance or severe cases can be treated with systemic drugs like oral corticosteroids, cyclophosphamide, cyclosporine, methotrexate and tumor necrosis factor (TNF) blockers[8,9,10,11,12,13]

The middle-aged female patient in our case presented with recurrent attacks of chondritis of ear cartilage, saddle nose deformity and arthritis. There were no other systemic manifestations. VDRL in dilution showed significant titre (1:16), confirmatory test for syphilis (TPHA) was also positive. ESR and CRP were mildly raised. The patient as well as her partner was prescribed intramuscular benzathine penicillin. However even after six months of penicillin treatment, the RP didn't improve. The dramatic response was seen within one week of starting 30 mg of oral corticosteroids and was continued for three weeks and then patient was lost to follow up. Hence, we report a rare case of RP in a middle-aged Indian woman fulfilling the diagnostic criteria by Damiani with no evidence of internal organ involvement and showing good response to moderate dose of oral corticosteroids.

From our case of RP, we can propose that infection with Treponema pallidum could be one of the factors to stimulate cell mediated and or humoral immune system thus triggering an inflammatory cascade in RP. Further studies about the beginning, perpetuation and activation of the immune system in infections like syphilis is needed.

CONCLUSION

Relapsing polychondritis is an uncommon, multisystem disease with relapsing and remitting course which is difficult to diagnose. Association with infections, inflammatory and autoimmune diseases has been suggested. Prompt diagnosis and treatment is required to prevent cartilage destruction and other comorbidities associated with the disease.

Declaration by Authors

Ethical Approval: not applicable
Acknowledgement: None
Source of Funding: None
Conflict of Interest: The authors declare no conflict of interest.

REFERENCES

1. Peebo B.B., Peebo M., Frennesson C. Relapsing polychondritis: A rare disease with varying symptoms. Acta Ophthalmol. Scand. 2004; 82:472–475. doi: 10.1111/j.1600-0420.2004.00276. x.
2. Borgia F,Giuffrida R,Guarneri F,Cannavo SP. Relapsing polychondritis: An updated Review.Biomedicines. 2018 Aug2;6(3): 84. doi: 10.3390 /biomedicines 6030084. PMID: 30072598; PMCID: PMC6164217.
3. Foidart J.M., Abe S., Martin G.R., Zizic T.M., Barnett E.V., Lawley T.J., Katz S.J. Antibodies to type II collagen in relapsing polychondritis. N. Engl. J. Med. 1978; 299:1203–1207. doi: 10.1056/NEJM197811302992202.
4. Yang C.L., Brinckmann J., Rui H.F., Vehring K.H., Lehmann H., Kekow J., Wolff H.H., Gross W.L., Müller P.K. Autoantibodies to cartilage collagens in relapsing polychondritis. Arch. Dermatol. Res. 1993; 285:245–249. doi: 10.1007/BF00371591.
5. Alsalameh S., Mollenhauer J., Scheuplein F., Stöss H., Kalden J.R., Burkhardt H., Burmester G.R. Preferential cellular and humoral immune reactivities to native and denatured collagen types IX and XI in a patient with fatal relapsing polychondritis. J. Rheumatol. 1993; 20:1419–1424.
6. Arnaud L., Mathian A., Haroche J., Gorochov G., Amoura Z. Pathogenesis of relapsing polychondritis: A 2013 update. Autoimmun. Rev. 2014; 13:90–95. doi: 10.1016 / j. autrev. 2013.07.005.
7. Damiani J.M., Levine H.L. Relapsing polychondritis-Report of ten cases. Laryngoscope. 1979; 89:929–946. doi: 10.1288/00005537- 197906000-00009.
8. Schumacher S., Pieringer H. Relapsing polychondritis: A chameleon among orphan diseases. Wien. Med. Wochenschr. 2017; 167:227– doi: 10.1007/s10354-017-0559-1.
9. Cuestas D., Penaranda E., Mora S., Cortes C., Galvis I., Patino M., Velasquez O. Relapsing polychondritis, an underestimated dermatological urgency: Case report and literature review. Int. J. Dermatol. 2017; 56:1379–1386. doi: 10.1111/ijd.13755.
10. Mark K.A., Franks A.G., Jr. Colchicine and indomethacin for the treatment of relapsing polychondritis. J. Am. Acad. Dermatol. 2002;46: S22–S24. doi: 10.1067/mjd.2002.105477.
11. Sharma A., Gnanapandithan K., Sharma K., Sharma S. Relapsing polychondritis: A review. Clin. Rheumatol. 2013; 32:1575–1583. doi: 10.1007/s10067-013-2328-x.
12. Sosada B., Loza K., Bialo-Wojcicka E. Relapsing polychondritis. Case Rep. Dermatol. Med. 2014; 2014:791951. doi: 10.1155/2014/791951.
13. Mathian A., Miyara M., Cohen-Aubart F., Haroche J., Hie M., Pha M., Grenier P., Amoura Z. Relapsing polychondritis: A 2016 update on clinical features, diagnostic tools, treatment and biological drug use. Best Pract. Res. Clin. Rheumatol. 2016; 30:316–333.doi: 10.1016/j.berh.2016.08.001.

How to cite this article: Kaul Neenu, Kumari Neeti. Relapsing polychondritis presenting with cauliflower ears and saddle nose deformity in a patient of syphilis incognito: two rare diseases in one patient. *Int J Health Sci Res.* 2023; 13(12):259-262. DOI: *10.52403/ijhsr.20231231*

******