Case Report

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# Relapsing Polychondritis Associated with Miscellaneous Ocular Symptoms and Increased IgA: a Case Report

Yu Zhang[1], Li Zhang[2], Zhenzhen Wu[1], Jianping Tang[1]*, Xuan Wang[1]*

[1]Department of Rheumatology and Immunology, Tongji Hospital, Tongji University School of Medicine, Shanghai, People's Republic of China; [2]Department of Ophthalmology, Tongji Hospital, Tongji University School of Medicine, Shanghai, People's Republic of China

## ABSTRACT

A male patient had suffered miscellaneous ocular symptoms for 20 years with auricular dysmorphosis and was diagnosed with Relapsing Polychondritis (RP) in the ear, nose, joints, and costal cartilage. The patient lost his vision owing to recurrent ocular symptoms for decades. He presented an increased IgA and was diagnosed with monoclonal gammopathy of undetermined significance (MGUS) and treated by prednisone and cyclophosphamide. His ocular symptoms relieved and serum IgA decreased after six months. In conclusion, RP is a systemic disease with a wide range of clinical symptoms and may lead to serious complications.

*Corresponding authors:
Jianping Tang,
Xuan Wang,
Department of Rheumatology and Immunology, Tongji Hospital, Tongji University School of Medicine, Shanghai, 200065, People's Republic of China
**Tel:** 13918745569
**Email:** tangjp6512@126.com (Jianping Tang)
xuan2018@tongji.edu.cn (Xuan Wang)

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## INTRODUCTION

Relapsing polychondritis is a rare disease characterized by inflammation and degeneration of the cartilage such as eyes, nose, ears, airways, and joints, which can be fatal in some cases with the destruction of the respiratory tract or hematological malignancies (1). For the varied symptoms and absence of specific diagnostic markers, RP is easy to be misdiagnosed at the early stage. A study has illustrated that about 47% of RP patients had misdiagnosis experience, with a time from symptom gambit to a final diagnosis of as long as 14.4 months (2). It is a difficult task for clinicians to differentiate RP from related systemic symptoms, thus patients with RP always experience a complicated diagnostic process. Patients with RP generally suffer a roundabout diagnostic

process. Differentiating RP from related complex situations is a huge challenge for clinicians. . Here, we report a case of a patient with RP associated with increased IgA, suffering from diverse ocular symptoms.

## CASE PRESENTATION

A 67-year-old Chinese man was referred to Immunology in 2018, with 20-year recurrent ophthalmitis and 15-year auricula dysmorphosis history. In 1999, scleritis occurred to his left eye and then right eye, he was treated with irregular antibiotic eyewash and oral prednisone with a low dose for recurrent ophthalmitis more than five times once a year in the past 20 years. In 2005, (at age 42) there appeared, again, recurrent pain, tenderness, swelling of the left auricle along with tinnitus, and the increased binaural auricle cartilage gradually blocked the ear canal leading to the deformation of auricle and hearing loss. In 2009, arthralgia in his right elbow joint led to the destruction of articular cartilage and then hyperosteogeny. In December 2019, he visited a rheumatologist for recurrent ophthalmitis and auricula dysmorphosis. The physical examination showed saddle nose (Figure 1a), deformation

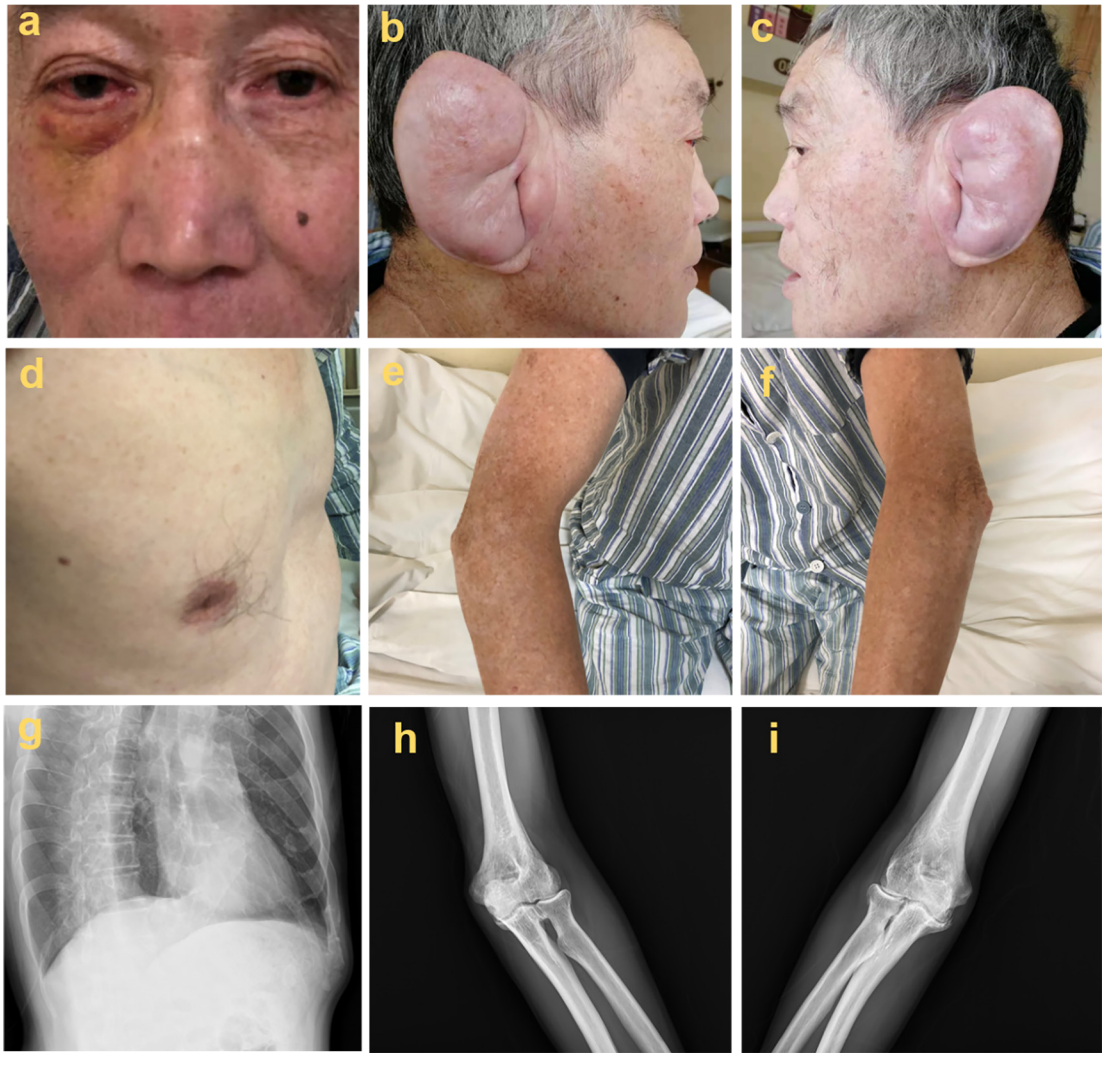


**Figure 1.** (a) shows the saddle nose, (b&c) shows the redness, swelling and deformation of right and left ear respectively, (d) shows the depression of the lower margin of sternums, (e&f) shows the deformation and limited extension of both elbow joints, (g) shows the collapsed sternum angel of the present patient with CT scanning, (h&i) shows the X-ray of both elbow joints of the present patient, both of the elbow joints show the destruction of cartilages and narrow joint space.

**Table 1. Shows the parameters of the Optic Disc of the present patient. The present patient was diagnosed with the optic nerve atrophy in the left eye for the mean thickness of RNFL is 52μm, which was lower than the average thickness (54.16-65.11μm) according to the age. RNFL represents the retinal nerve fiber layer.**

| | OD | OS |
|---|---|---|
| Mean thickness of RNFL | * | 52μm |
| Whether the RNFL is symmetrical or not | NO | |
| Acreage of the disk border | * | 0.63mm$^2$ |
| Acreage of optic disk area | * | 1.68mm$^2$ |
| Average ratio of cup-to-disc | * | 0.79 |
| Vertical cup-to-disk ratio | * | 0.77 |
| Volume of cup | * | 0.457mm$^2$ |

of the auricle (Figure 1b.c), depression of the lower margin of the sternum (Figure 1d), the destruction of articular cartilage, and limited extension of both elbow joints (Figure 1e. f). Eventually, a diagnosis of RP was made. Then he received a comprehensive inspection and was diagnosed with panuveitis in both eyes, secondary glaucoma in the right eye, lens defect in the right eye, senile cataract in the left eye, and optic nerve atrophy in the left eye (Table 1). He received symptomatic treatment to control the response of uveitis.

The laboratory tests revealed normal C reactive protein (CRP), increased erythrocyte sedimentation rate (ESR), slightly lower complements, and the PET-CT revealed a diffuse abnormal increase of fluorodeoxyglucose (FDG) metabolism in the bilateral auricle. A monoclonal IgA kappa band was noted total serum IgA 8.95 g/l (normal 0.82–4.53g/l), K/ λ = 3.43. Other immunoglobulins, antinuclear antibody (ANA), rheumatoid factor (RF), levels of C3 and C4 complement were normal. Bone marrow aspirate suggested that plasma cell ratio increased to 3% with morphological changes and FISH showed about 10% of cells losing the D13S319 gene, 12% of cells losing RB1 gene. The patient was confirmed with the diagnosis of monoclonal gammopathy of undetermined significance, was treated with prednisone 30mg/d, cyclophosphamide 600mg/m, and thalidomide 50mg/d. Panuveitis and chondritis were alleviated and IgA in serum decreased after six months of treatment.

## DISCUSSION AND CONCLUSION

The clinical features of RP include saddle nose deformity, external ear chondritis, sensorineural hearing loss, otitis media, subglottic strictures, and arthritis. McAdam described the diagnosis criteria of RP, in which patients must present three of the following six features: respiratory tract chondritis, non-erosive sero-negative inflammatory polyarthritis, nasal chondritis, ocular inflammation bilateral auricular chondritis, and audiovestibular damage (3).

65% of the patients with RP have been reported exhibiting ocular complications including not only the common, scleritis, conjunctivitis, uveitis, and keratitis, but also the rare eyelid edema, proptosis, extraocular muscle palsy, eyelid edema, retinopathy, and optic neuritis (4, 5). The patient in our case suffered not only from general ocular symptoms of RP such as conjunctivitis, scleritis, and keratitis but also a few reported severe vision losses, which were caused by optic atrophy. There have been few previous reports displaying such an extensive variety of ocular symptoms in a patient with RP. The present patient suffered from miscellaneous ocular symptoms for decades and was treated with corticosteroid eye drops and ointment.

More and more cases of RP have been reported relating to malignant tumors, particularly myelodysplastic syndrome (MDS) and not prevailing solid tumors such as lung, breast, colon, pancreas, bladder, or serious

hematological malignancies (lymphoma) (6). In this case, we found him with an increased IgA, losing the D13S319 gene and RB1 gene, which are common to be detected in sundry hematological malignancies including acute myeloblastic leukemia (AML), chronic myelogenous leukemia (CML), chronic lymphocytic leukemia (CLL), MDS and multiple myeloma (MM)(7-9). The correlation between RP and MDS has been extensively documented in the literature, showing up to 27% of patients with RP accompanying MDS, and on the contrary, the complication of MGUS is rare. MGUS is a condition where an abnormal protein called M-protein is found in the blood, which can develop life-threatening blood diseases such as MM and lymphoma (10). Considering the risk of underlying malignancy, the current patient's care should include regular follow-up for an extended period of time. It is necessary to distinguish the patients with MGUS and ameliorate hyperglobulinemia before any severe outcomes appear, such as malignancies.

Patients who were diagnosed with RP should be evaluated systemically. Clinicians especially ophthalmologists should consider RP when patients display multiple system symptoms associated with recurrent and refractory ocular inflammatory disorders. Hematologists should realize RP patients could appear with irregular MGUS and hematological malignancies, even those who might not have typical chondritis. Meanwhile, after diagnosing RP, rheumatologists should evaluate the risk of hematological malignancies and ocular symptoms to avoid misdiagnosis or other severe outcomes. This case shall unfold the mask of RP with irregulated manifestations and be helpful in prompt treatment of high-risk patients and decrease the morbidity and mortality related to the disease.

## LIST OF ABBREVIATIONS

RP: relapsing polychondritis; MGUS: monoclonal gammopathy of undetermined significance; CRP: C reactive protein; ESR: erythrocyte sedimentation rate; FDG: fluorodeoxyglucose; MDS: myelodysplastic syndrome; AML: acute myeloblastic leukemia; CML: chronic myelogenous leukemia; CLL: chronic lymphocytic leukemia; MM: multiple myeloma

## ETHICS APPROVAL AND CONSENT TO PARTICIPATE

The study was approved by the Human Ethics Committee of Tongji Hospital. Written informed consent was obtained from the patient.

## CONSENT FOR PUBLICATION

The patient gave written informed consent to the publication of their images, as well as their personal and clinical information.

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## AUTHORS' CONTRIBUTIONS

Xuan Wang collected the patient's information and contributed to supervision and final approval, Yu Zhang drafted this manuscript, analyzed and interpreted the patient's data, and edited the photos. Li Zhang and Jianping Tang made critical revisions to the manuscript. Zhenzhen Wu edited the photos and table. All authors read and approved the final manuscript.

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## AVAILABILITY OF DATA AND MATERIALS

All data generated or analyzed during this study are included in this published article.

**Conflict of Interest:** None declared.

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