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CASE REPORT

A Case of Relapsing Polychondritis with Multisystemic Involvement

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Abstract: Relapsing polychondritis is a rare immunologic disorder that can involve all cartilage and proteoglycan-rich tissues. Clinical symptoms of relapsing polychondritis are often associated with recurrent inflammatory manifestations and functional impairment of such tissues. The disease has an insidious onset, and the first symptoms and clinical manifestations vary, making it easy to misdiagnose and miss the diagnosis. Here, we report a case of relapsing polychondritis with multi-systemic disease in a single ear and a single eye. A 44-year-old Chinese man suddenly developed redness, swelling, and pain in the right auricle without any causative factors half a year ago, followed by other multi-systemic symptoms, and he went to an outside hospital several times. He was diagnosed with sinusitis and bronchitis after completing some investigations in a foreign hospital, and his condition did not improve after taking oral medication, so he came to our hospital and was diagnosed with relapsing polychondritis after completing some investigations.

Keywords: relapsing polychondritis, auricular chondritis, systemic autoimmune disease, diagnosis

Introduction

Relapsing polychondritis (RP) is a severe systemic immune-mediated disease characterized by relapsing inflammation and progressive structural destruction of local cartilage structures and proteoglycan-rich tissues, which can occur independently or in association with other diseases.[1] RP is considered to be a rare disease, with a large number of single case reports, but few patient series have been reported in the literature. The estimated incidence is 0.71/1,000,000/year.[2] Currently, the pathogenesis of RP is unclear and involves the humoral immune system and cell-mediated immune responses, with type II collagen, matrilin-1 and cartilage oligomeric matrix proteins (COMP) as potential target antigens.[3]

Here, we report a case of a middle-aged male patient. Six months ago, he suddenly developed redness, swelling and pain in the right auricle without any triggers, and then other multi-system symptoms appeared one after another during several visits to an outside hospital, where he was diagnosed with sinusitis and bronchitis, and was given medication to improve his condition. One week ago, his right external auricle was red, swollen and mildly painful again, and at the same time, his right eye was congested, painful and tearful, and he had pain in his ribs near the sternum bilaterally, coughing and coughing up white sputum, and he was diagnosed with relapsing polychondritis after completing the relevant examinations in the dermatology clinic of our hospital. We reviewed the relevant literature and found that diagnostic delays and even misdiagnosis are frequent due to the variety of patients' first symptoms and the inability to rely on specific tests alone to confirm the diagnosis. The misdiagnosis rate of our patients is as high as 47%, and the average delay in diagnosis is 14.4 months.[4] Our patient was also

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frequently misdiagnosed during his visits to outside hospitals, and we report this case to raise the awareness of clinicians in various departments of this disease, to reduce omission and misdiagnosis, and to provide timely and effective treatment to patients, which is the key to improving the prognosis of this disease.

Case Presentation

Medical History

A 44-year-old Chinese male patient suddenly developed right auricular erythema with mild pain in April 2023 without any causative factors and was treated with dexamethasone 5 mg IV at a local clinic, and the lesions disappeared after 4 days. However, shortly after stopping the medication, he again developed erythema and pain in the right auricle, accompanied by runny nose, sneezing, nasal congestion and bloody nasal discharge. He felt that he had phlegm flowing into the laryngeal trachea, which caused coughing, coughing up white sputum, occasional choking on water, and coughing accompanied by sweating. In August, the results of nasal CT and chest CT examinations at the Third People's Hospital of Haikou City suggested bilateral sinusitis, bilateral turbinate hypertrophy, and bronchial wall thickening of an indeterminate nature, and further clinical examination was recommended. The doctor treated him according to sinusitis and bronchitis, and his condition did not improve, and the patient was not clear about the specific medication. A week ago, his right external auricle became red and swollen again and he felt mild pain, and his right eye was congested, painful and watery. He felt mild pain in both ribs near the sternum with cough and white sputum, but no fever, headache, dizziness or hearing loss. Therefore, he came to our dermatology clinic. The patient had no history of chronic illness or family history of genetic, psychiatric, neoplastic or similar disorders.

Physical Examination

The patient was in good general condition, his superficial lymph nodes were not palpably enlarged, and there were no abnormalities in the heart, lungs or abdomen. Dermatological examination: his right auricle was diffusely oedematous erythematous and swollen, with localised elevated skin temperature and positive tenderness on pressure, and there was no erythema of the earlobe (Figure 1). His left ear was normal.

Auxiliary Examination

In October 2023, the patient's blood tests revealed leukocytes 15.63 $*10^9$/L, neutrophils 12.32$*10^9$/L, neutrophil ratio: 78.8%, CRP: 31mg/L, sedimentation rate: 60mm/h, immunoprofile: IgG: 18.87g/L, C3: 0.42g/L, Rheumatoid Factor, anti-nuclear antibody, anti-ENA antibody, anti-ds-DNA were all negative.

He underwent ophthalmological examination in October 2023, his visual acuity was 4.9+3 in the right eye, 4.5 in the left eye, 4.7 after correction, pressure 19 mmHg in the right eye, 19 mmHg in the left eye, mixed congestion in the right eye (+++), follicular hyperplasia, sclera with no ulcerated surfaces, corneal hyaline, staining was negative, the anterior chamber was clear, the lens was not cloudy, the pupil was 3*3 mm, and he was sensitive to light. There was no turbidity in the vitreous of his right eye, and the fundus of the eye had a mildly unclear optic papilla border that was reddish in colour, a flat retina with no oedema or exudation, and a normal macula (Figure 2A–C). The fundus examination of his left eye was unremarkable (Figure 2D).

In October 2023, he underwent a chest CT and observed thickening of the anterior and lateral walls of his bronchus and no thickening of the posterior wall of an indeterminate nature, and further clinical investigations were recommended. He had hypodense foci in the left inner lobe of the liver of uncertain nature, few calcifications in the wall of the left coronary artery, and no abnormalities in both lungs (Figure 3A). He underwent a CT scan of the nose and observed bilateral sinusitis with bilateral turbinate hypertrophy (Figure 3B).

In October 2023, the doctor took a biopsy specimen of the patient's right auricle skin and cartilage tissue. Pathological examination showed mild thickening of the epidermis, collagen fibre hyperplasia in the dermis, massive inflammatory cell infiltration including lymphocytes, neutrophils, plasma cells and a small amount of eosinophilic granulocyte infiltration was seen in the lower part of the dermis, inflammatory cell invasion of the cartilage, degeneration of the cartilage cell margins, and reduction of the chondrocytes (Figure 4A–C).

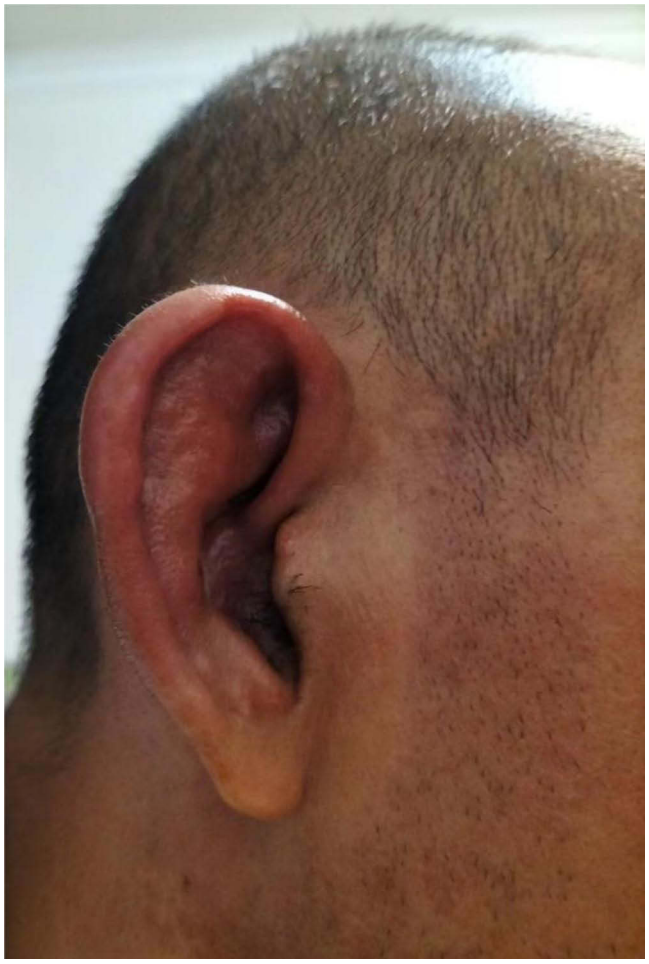

Figure 1 Diffuse oedematous erythema and swelling of the right auricle with no involvement of the earlobe.

Diagnosis and Treatment

According to the history, clinical manifestations and examination results, the disease was clearly diagnosed as "multiple chondritis", and the patient was given oral prednisone tablets of 30mg once a day, and topical tobramycin dexamethasone eye drops for the right eye four times a day. The patient felt that his right eye was congested, his cough and sputum decreased, and his sternal pain decreased, but his right ear erythema and pain did not decrease significantly, so he was given oral prednisone tablets at 45 mg once a day because his condition was not relieved. After one week of treatment, the patient had no erythema and pain in the auricle (Figure 5), and no congestion in the right eye, so he was instructed to discontinue tobramycin dexamethasone eye drops, and oral prednisone continued to be reduced to 35 mg once a day for maintenance treatment. When his prednisone was reduced to 30mg once a day, he was added to oral treatment with tretinoin tablets 20mg three times a day. He is now maintained on prednisone at 20mg and tretinoin tablets 20mg twice a day orally. Currently, his condition is stable and he is on maintenance treatment at a reduced dosage.

Discussion

RP is a severe systemic immune-mediated disease characterised by recurrent inflammation of the affected tissues leading to progressive anatomical deformation and dysfunction of tissue structures. Widespread inflammation of cartilage in the ear, nose, larynx, trachea – bronchial tree and joints predominates. Proteoglycan-rich structures such as the eyes, heart, blood vessels, inner ear and skin may also be involved. The disease may occur independently or in association with other diseases.[1] RP is considered a rare disease with a large number of single case reports, but few patient series have been

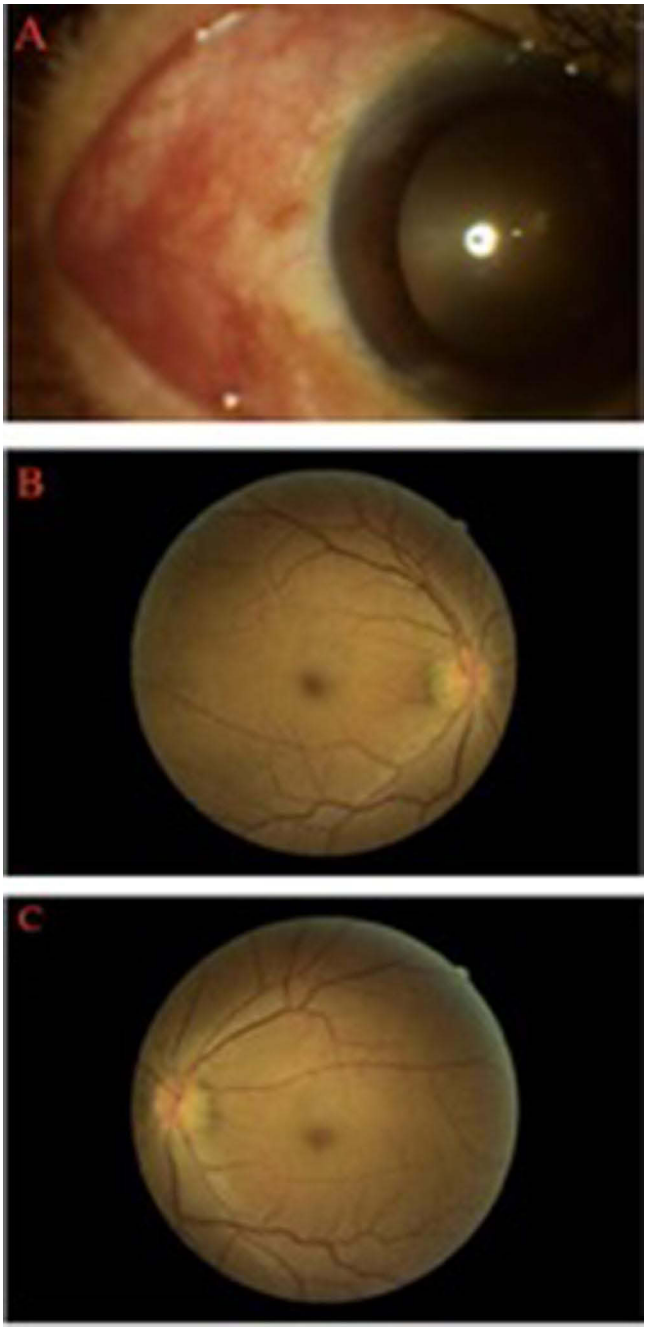

Figure 2 (**A**) Mixed congestion in the right eye. (**B**) Right fundus with mildly unclear optic papilla border. (**C**) No abnormality in the left fundus.

reported in the literature. The estimated incidence is 3.5/1 million/year,[5] but a recent UK population-based cohort study reported an incidence of 0.71/1 million/year.[2]

The disease was first described by Jaksch-Wartenhorst in 1923 and named "polychondritis".[1] In 1960, Pearson et al introduced the term "recurrent polychondritis" to emphasise the peculiar intermittent course of the disease observed in 12 patients.[2]

The exact pathogenesis of RP has not been clarified. Genetic studies have identified HLA-DR4 as the major risk allele for RP and a negative correlation between the severity of organ involvement and HLA-DR6.[6] Circulating autoantibodies against collagens II, IX and XI have been found in patients with RP. Collagen type II (CII) accounts for 95% of the total collagen content of cartilage and may be a major target of autoimmunity. In fact, circulating antibodies against CII were detected in one-

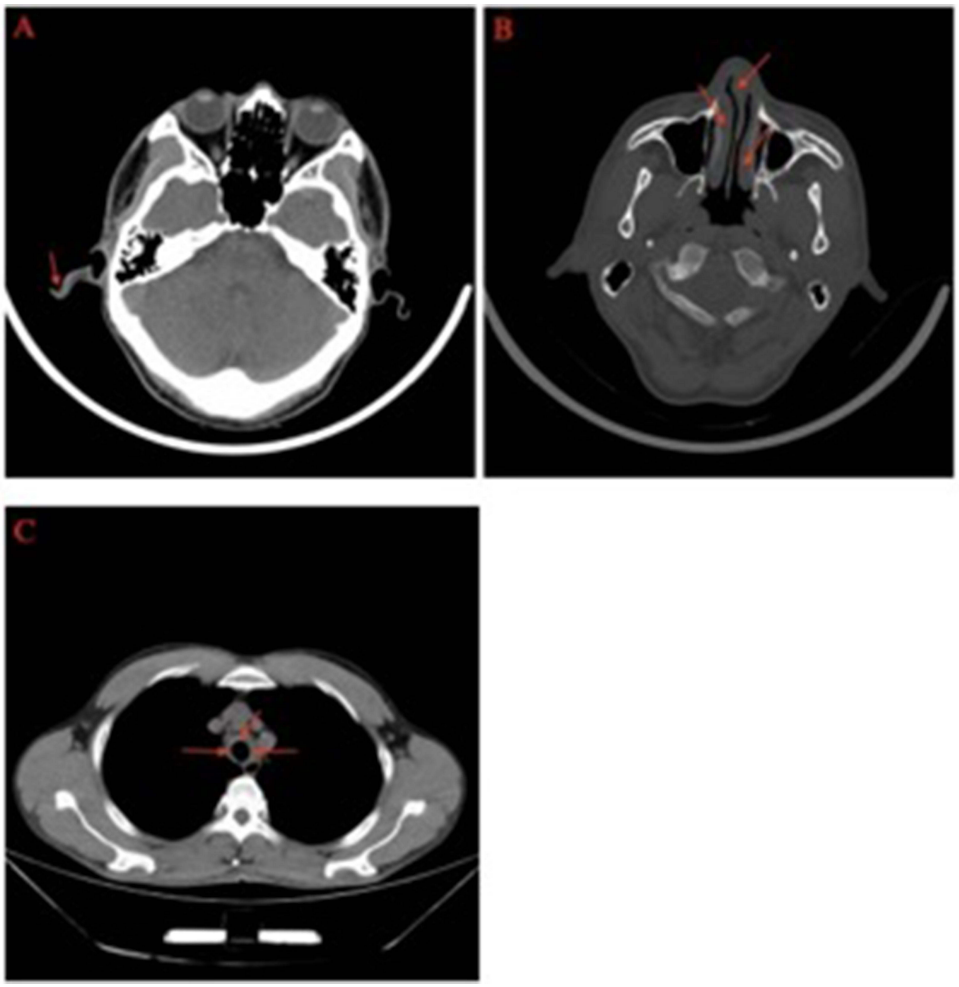

Figure 3 (**A**) Hypertrophy of the right auricular cartilage. (**B**) Hypertrophy of the nasal turbinate and nasal septum cartilage. (**C**) Thickened bronchial ring cartilage.

third of patients with active recurrent chondritis, with a positive correlation between serum titres and disease severity,[7] suggesting that cartilage-specific autoimmunity may play a key role in the pathogenesis of RP.[7–9] Other known target autoantigens are matrilin-1 and cartilage oligomeric matrix protein (COMP). COMP is expressed predominantly in tracheal, nasal, auricular and sternal cartilage, but not in normal adult articular cartilage.[10] The involvement of COMP in the trachea, nasal cavity, and auricle was confirmed in our patient, and the pain at the sternocostal joints in our patient was highly suspicious of the presence of anti-COMP antibodies in his body, but there is no clinical method to detect anti-COMP antibodies, so we can only guess for the time being. PET/CT (positron emission tomography/computed tomography) may also be useful. For example, FDG (fluorodeoxyglucose) accumulation has been detected in subclinical auricular manifestations in various cases, or in the bronchial system.[11] Identification of FDG enrichment may help to detect subclinical cartilage involvement. This may also be accompanied by intensification of immunosuppressive therapy.[12]

In 1976, McAdam et al first proposed diagnostic criteria for RP based on the clinical presentation of 159 patients.[13] Three of the following 6 criteria: bilateral auricular chondritis; non-erosive, seronegative inflammatory polyarthritis; nasal chondritis; ocular inflammation (eg, conjunctivitis, keratitis, scleritis/scleral epitheliitis, uveitis); respiratory chondritis (eg, laryngeal and/or tracheal chondritis); and damage to the cochlea and/or vestibule (eg, tinnitus, vertigo, neurological hearing loss). These criteria were later modified by Damiani and Levine[14] as follows: at least 3 McAdams criteria; or at least 1 McAdams criterion plus a positive histological finding; or chondritis at two or more sites, responsive to glucocorticoids and/or dapsone. Michet et al[15] modified the criteria to read as follows: inflammation of two of the following three sites: ear, nose, laryngeal tube or Inflammation at one of these sites with two other symptoms

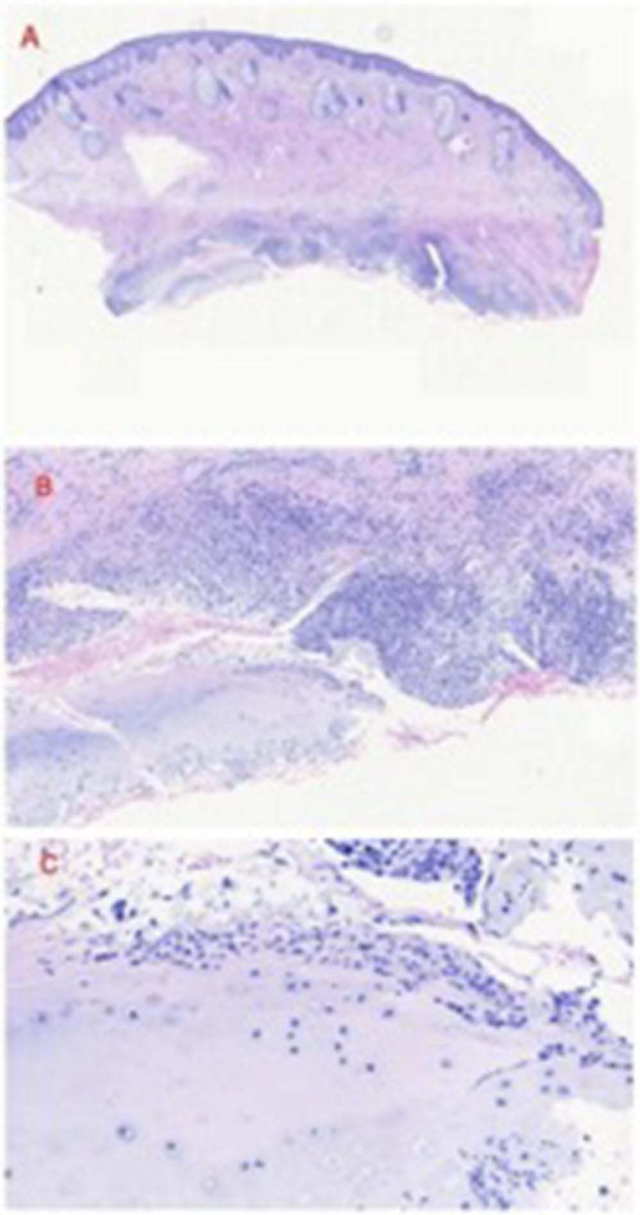

Figure 4 (**A**) Mild thickening of the epidermis, collagen fibre hyperplasia in the dermis, and a large infiltration of inflammatory cells was seen in the lower part of the dermis and cartilaginous tissue (HE, X20). (**B**) A large number of lymphocytes, histiocytes, neutrophils infiltration and vasodilatation and congestion were seen in the lower dermis and around the cartilage tissue (HE, X100). (**C**) Degeneration of cartilage tissue margins with invasion of inflammatory cells and reduction of basophils (chondrocytes) (HE, X400).

Inflammation of the eyes, hearing loss, vestibular dysfunction, or seronegative inflammatory arthritis. This patient has otitis media, nasal chondritis, tracheitis, scleritis, arthritis (pain in the thoracic and rib joints), effective glucocorticoid hormone therapy, and histopathological examination of the skin of the external auricle showing cartilage damage. The patient fulfilled the diagnostic criteria for all three RPs.

Because most cases of polychondritis develop in both ears, this disease clinically develops in one ear. In China, Huang Yanping[11] reported a case of multiple chondritis in which the right ear developed half a year before the left ear developed. Therefore, bilateral auricular cartilage inflammation in polychondritis does not necessarily develop at the same time, and one auricle may be affected first, and the other auricle may be affected several months later. In China, Liang Siyu et al[12] reported a case of multiple chondritis misdiagnosed as auricular chondritis. The patient first had a red, swollen and painful left auricle for 3 months, and was treated with local debridement combined with anti-infective therapy. On the second postoperative day,

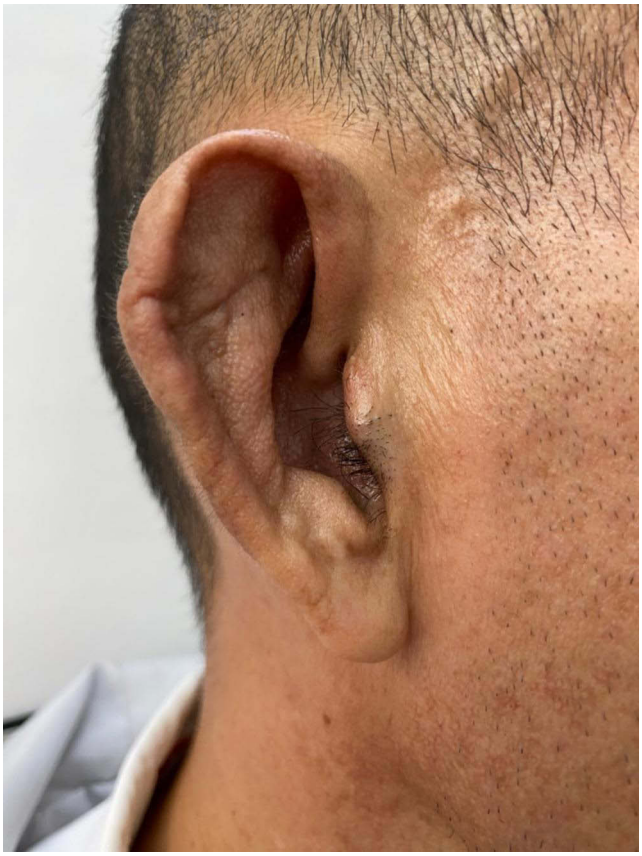

Figure 5 After two weeks of treatment, the patient's right ear swelling was basically free of redness, and the colour was significantly darker than before.

the right auricle became erythematous and painful. Postoperative pathology of the left auricle revealed chronic inflammation of the auricular cartilage. Finally, dexamethasone was added to the treatment, which was effective. After the patient was discharged from the hospital and stopped taking the medication, he again had redness, swelling and pain in both auricles, accompanied by generalised joint pain, and was diagnosed with multiple chondritis. Therefore, it is easy to misdiagnose multiple chondritis as auricular chondritis when one auricle is involved at the beginning. However, in this case, the patient's ear lesions were mainly infiltrative erythema, swelling, and pain, and for the dermatologist's first consideration could be inflammation of the skin and soft tissues, but the affected lesions were in the auricle, which has cartilaginous tissues in addition to the skin and soft tissues, and the patient also had respiratory involvement, so it was necessary to consider whether there was inflammation in the cartilaginous tissues as well. This patient was misdiagnosed several times in external hospitals during the first half year of his illness, because the first symptoms of RP are diverse, and it is not possible to rely only on specific tests to confirm the diagnosis, so diagnostic delays and even misdiagnosis occur frequently. The misdiagnosis rate of patients in China is as high as 47%, and the average delay in diagnosis is 14.4 months.[4] The prognosis of RP is related to whether the diagnosis and treatment of patients are timely and appropriate. Therefore, we report this case in order to raise the awareness of clinicians in various departments of this disease and to reduce missed diagnosis and misdiagnosis. Timely and effective treatment is the key to improve the prognosis of this disease.

There is a lack of evidence-based guidelines for the treatment of this disease, and treatment remains empirical, with glucocorticoids, immunosuppressants, and ampicillin as the main therapeutic agents. Steroid therapy is often recommended to prevent recurrence during long-term follow-up, but this does not alter the progression of the disease. In the case of arthritis, 15–20 mg/day are usually taken, and the dosage should be reduced slowly. In the presence of inflammation of the trachea, higher dosages of 0.5–1 mg/kg body weight are used initially. In severe courses of the disease, glucocorticoid shock therapies of 250 mg/day to 1 g/day over a short period of 1 to 3 days as well as therapies.[12]

Immunosuppressants are indicated in patients who are intolerant to corticosteroids or dependent on them, or in cases where there is a lack of response to corticosteroids or a need for corticosteroid replacement therapy.[10,16–18] Some reports

suggest that biologics may help to improve RP cases refractory to conventional therapies, but the number of cases is still too small to ensure the efficacy and safety of biologics in patients with RP, and further clinical trials are needed to verify the observations. Our patient responded well to glucocorticoid therapy and is currently on low-dose maintenance therapy.

Ethics Statement

The publications of images were included in the patient's consent for publication of the case. The Hospital Ethics Committees of the Fifth People's Hospital of Hainan Province approved to publish the case details.

Consent Statements

Written informed consent was provided by the patient to have the case details and any accompanying images published. Institutional approval was not required for this case study.

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Disclosure

The authors report no conflicts of interest in this work.

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