RED EARS, RED EYES AND HEARING LOSS: A DIAGNOSTIC DILEMMA

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Abstract

Relapsing polychondritis (RP) is a disease that confounds clinicians worldwide due to the difficulty in confirming its diagnosis and determining the best treatment plan. We report a case of a 34-year-old lady who presented with bilateral inflamed auricular swelling and red eyes, which turned out to be relapsing polychondritis. The patient was successfully treated with long-term oral steroids and is on regular follow-up. This report highlights how the subtle signs of her condition were picked up, leading to her diagnosis of RP and the treatment regime that successfully reversed her symptoms. The challenge of picking up subtle signs of the disease is best met by working as a team in a multidisciplinary setting. A literature study shows that RP is a rare disease that can present with many symptoms.

Keywords: Relapsing Polychondritis, Immune-mediated, Conjunctivitis, Auricular Perichondritis

Introduction

RP is an immune-mediated disease that inflicts the cartilaginous structures of the body. Traditionally, the patients present with an episodic or progressive condition ensuing inflammation of any of the cartilaginous structures in the body. Nonetheless, the ear, nose, airway, and joints remain the most common cartilaginous structures. RP has also been reported to affect the heart, eyes, inner ears, and skin, which are proteoglycans-rich structures (1-3). Diagnosing RP remains a conundrum for most clinicians worldwide following its vague clinical features, episodic nature, and multiorgan involvement, which frequently leads to a delay in diagnosis. It is worth noting that the duration from the first presentation to the diagnosis of RP has been reported to be between 1.9 to an astounding ten years, with an average of five clinical visits before its diagnosis (4, 5).

Clinical features play a cardinal role in diagnosis. Concurrently, a myriad of criteria has been developed over time to aid diagnosis. Amongst the notable criteria that exist to date include McAdam et al. (1976) (6), Damiani and Levine, (1979) (7) and Michet et al. (1986) (8).

McAdam et al.'s (1976) criteria remain the most favored, whereby at least three out of six of the following clinical findings must be present to diagnose RP. These include 1) auricular chondritis bilaterally, 2) non-erosive seronegative polyarthritis, 3) nasal chondritis, 4) eye inflammation, 5) respiratory tract chondritis, and 6) cochlear and/or vestibular disturbance. Damiani and Levine expanded these criteria, including meeting one of McAdam et al.'s criteria in addition to histopathogical examination of the sample and response to corticosteroids. On the other hand, Michet et al.'s (1986) criteria requires the presence of proven chondritis in at least two of three of the auricular, nasal or laryngotracheal cartilages, or chondritis in one of these cartilages in addition to two other signs, including ocular inflammation, vestibular dysfunction, seronegative inflammatory arthritis, and hearing loss.

Case study

A 34-year-old lady with a premorbid condition of Evans syndrome presented with a one-month history of bilateral inflamed and swollen pinna. According to the patient, she noticed bilateral pinna swelling, which progressively worsened and subsequently became inflamed with tenderness. The patient also noticed bilateral eye redness with no visual disturbance. No recent trauma or fall history was elicited, and the patient has no known allergies. Upon further enquiry about her symptoms, the patient mentioned that she noticed reduced hearing over the past few months and was unsure of the laterality. There was no accompanying tinnitus, vertigo, facial asymmetry, otalgia or otorrhea. She sought treatment from a general practitioner and was given a course of antibiotics which did not alleviate her symptoms. Additionally, there were no other recurrent

nasal or throat symptoms. Her menstrual history was normal, her sleep and appetite were not disturbed, and her bowel and bladder habits were normal. She also had no prior complaints of joint pains or swellings.

Upon examination, the patient appears comfortable under the room air. Pinna inspection revealed a bilaterally swollen and erythematous pinna (Figure 1a & 1b). Upon palpation, both pinnae were firm but tender. Otoscopic examination revealed intact tympanic membranes bilaterally and bilateral normal external auditory canal. Examination of her eyes revealed bilaterally erythematous sclera. Her nasal, throat and neck examinations were unremarkable.

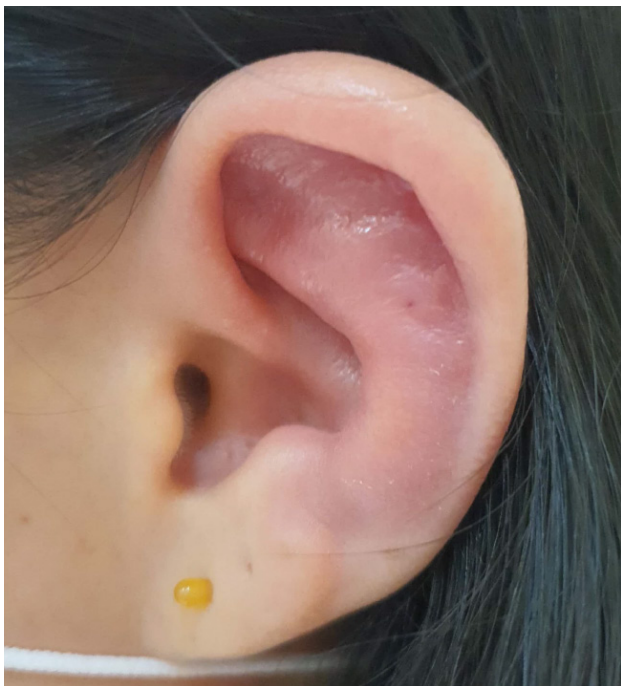

Figure 1a: Left swollen and erythematous pinna

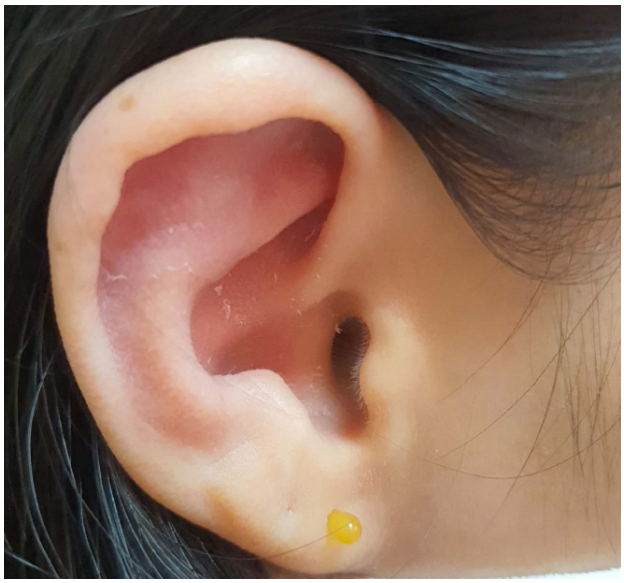

Figure 1b: Right swollen and erythematous pinna

We sent baseline blood investigations which revealed no significant abnormalities. The white blood cell count was 7.8×10^9/L, and both hemoglobin and platelet levels were within normal ranges. In addition, her kidney and liver function tests were within normal parameters. However, her C-reactive protein test was slightly raised at 16.5 mg/L, and the antineutrophil cytoplasmic autoantibodies (ANCAs) screen was positive. All other autoimmune workouts were not significant.

Pure tone audiometry was also done and revealed right moderate sensorineural hearing loss with normal hearing on the left. She was subsequently referred to the Rheumatology and Ophthalmology team. Eye assessment was normal. She was started on a short course of oral prednisolone 20 mg daily for six weeks. Upon follow-up, her pinna and eye redness improved. However, the repeated audiogram showed similar findings.

Results and discussion

Relapsing polychondritis (RP), first mentioned in 1923 by Jaksch-Wartenhorst was initially termed as polychondropathia (9). The term RP was coined in 1960 by Pearson et al. after observing a cohort of 12 patients experiencing on and off recurrent symptoms (10). McAdam et al. (6), Damiani and Levine (7), and lastly, Michet et al. (8) came up with different variations of the diagnostic criteria (Table 1).

Table 1: Diagnostic criteria for relapsing polychondritis

Diagnostic criteria	Details of criteria
McAdam et al. (6)	At least three criteria out of the following six: 1) bilateral auricular chondritis, 2) non-erosive seronegative polyarthritis, 3) nasal chondritis, 4) ocular inflammation, 5) respiratory tract chondritis, and 6) cochlear and/or vestibular dysfunction
Damiani and Levine (7)	One of McAdam et al.'s criteria plus histological confirmation and/or efficacy to corticosteroids.
Michet et al. (8)	There is proven chondritis in at least two of three of the auricular, nasal or, laryngotracheal cartilages, or chondritis in one of these cartilages plus two other signs including ocular inflammation, vestibular dysfunction, seronegative inflammatory arthritis, and hearing loss.

In our case, we utilized Michet et al.'s criteria (8) for the diagnosis of our patient. She fulfilled the presence of chondritis in her auricular cartilage plus two other signs, which were ocular inflammation and hearing loss. This is

in line with literature that states that the most common feature of RP is aural chondritis, which is observed in up to 90% of patients (11). She was also found to have bilateral sensorineural hearing loss, which is present in 46% of patients with RP (12). Traditionally, hearing loss secondary to autoimmune or connective tissue disorders are bilateral, although unilateral presentation has also been reported.

Our patient also had episcleritis which is the most common ocular symptom in RP. Ocular symptoms are present in 50–60% of cases of RP (11).

Due to its vast involvement of organs and systems, previous studies have suggested a certain regime of investigations to be carried out. Inflammatory markers such as CRP are suggested even though they might not be raised in a mild flare-up episode (13). In our patient, her CRP was slightly raised.

A typical baseline assessment may include otorhinolaryngology and ophthalmology examinations, cardiovascular screening with an electrocardiogram, renal function testing, and testing for antineutrophil cytoplasmic autoantibodies (ANCAs) (14). All these investigations were carried out for our patient, which aided us with our diagnosis.

In our patient, due to the manifestation of RP, mostly in her ears, we had a few differential diagnoses to consider. We ruled out infection via a diagnostic aspiration which yielded no pus or collection. Her white blood cell count was also not raised. Another differential diagnosis was chondrodermatitis helices nodularis, an inflammatory and degenerative skin lesion of unknown etiology (15). In our patient, this was ruled out because her ear lesion was not a localized, circumscribed lesion and did not show any other signs of chondrodermatitis helices nodularis.

To date, no universally accepted guidelines exist for treating RP. Treatment is based on the practitioner's clinical experience and empirically given based on the severity of each flare-up. Mild forms are treated with NSAIDs, colchicine, dapsone, and low-dose corticosteroids (16), although a standardized regime does not exist to date. In the same vein, the efficacy and outcome of each of the pharmacological agents show varying results. Continuous use of prednisolone has been reported to decrease the severity, frequency and duration of relapses. In the acute phase, prednisolone is administered 20-60 mg daily and tapered subsequently to 5-25 mg daily for maintenance. Life-threatening or organ-threatening complications such as ocular and laryngotracheal involvement and systemic vasculitis require high-dose systemic corticosteroids or immunosuppressants (16). Immunosuppressant agents such as methotrexate, azathioprine and cyclosporine are commonly reserved for patients intolerant to steroids or those unresponsive to steroids (16). Our patient benefitted from low-dose corticosteroids for two weeks, which helped clear her initial presenting symptoms. It is noteworthy that follow-ups for patients with RP are typically long-term owing to the possibility of relapse and to monitor disease.

Conclusion

Relapsing polychondritis (RP) is a rare entity and can easily be missed if the criteria that justify its diagnosis are not detected. Therefore, all suspected cases of RP must be approached by a multidisciplinary team so that all the criteria required for a diagnosis can be confirmed either via examination or investigation. In our case, although the patient did not complain of hearing loss, we managed to confirm it via a hearing assessment which helped to fulfil the criteria for diagnosis. The treatment of RP also needs to be catered to each presentation due to its vast and varied presentations. The patient reported in our case responded well to corticosteroid therapy. We also concluded that the differential diagnoses of auricular swelling need to include RP, even though auricle perichondritis is a more frequent presentation. As with this case, when we confirmed the diagnosis of RP and catered our treatment towards it, the patient's symptoms resolved.

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Ethical clearance

No ethical clearance was required as this is a case report.

Competing interests

The authors declare that they have no competing interests.

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