# Point-by-point responses to the comments from the editorial and reviewers.

First of all, we would like to thank the reviewers for their great comments to improve the quality of the manuscript.

#### Reviewer 1:

**#1.** "The authors have presented a rare case of RP with comprehensive clinical and radiological characteristics. The manuscript is of high quality except for minor mistake. "Fluid correction" should be fluid collection. "

### Response

Thank you for your valuable comments and thoughtful review. We have revised the word as per your pointing out (Page 3, line 11, page 9, line 5 and page 9, line 10 of the clean revised version):

From
"correction"
To
"collection"

#2. "However, this case has limited clinical importance."

#### Response

Thank you for your comment. Actually, this finding doesn't result in the clinical course of patients with relapsing polychondritis; however, the current finding might be the answer of why inflammation occurs in mastoid air cells that lacks of chondroid tissue. I added a sentence and amended table 1 to emphasize clinical importance (Page 8, lines 14-15 and Page 18, line 11 of the clean revised version):

"The edema and contrast enhancement of ETs disappeared in the follow-up MRI at 8 weeks."

Table 1 Summary of the clinical course of the patient

Time point	Remarks
July 24, 2021	Redness of the bilateral auricles
July 27, 2021	Antibiotics administration at a clinician
August 10, 2021	Referred to our hospital
August 18, 2021	Non contrast enhanced chest to abdominal CT without
	significant findings
August 20, 2021	Biopsy for auricular cartilage
August 21, 2021	Contrast enhanced head and neck MRI with presenting figures
	Prednisolone administration was started
September 1, 2021	Symptoms and laboratory abnormality improved remarkably
October 28, 2021	Disappeared edema and contrast enhancement of eustachian
	tubes on MRI

# **Reviewer 2:**

"none"

Response

Thank you for your review.

#### **Reviewer 3:**

#1-1. What's your new findings of this case?

# Response

Thank you for your valuable comments and thoughtful review. The most important point of current report is that the Eustachian tube (ET) is an involved region in patients with relapsing polychondritis (RPC), which haven't noticed previously. We have discussed it in discussion session (Page 7, lines 7-8 of the clean revised version):

"To the best of our knowledge, this is the first report of a patient with RPC manifesting enhancing and edematous ET on MRI."

#1-2. "What rules have you learned about diagnosing the disease?"

#### Response

Auricles are most familiar site of involvement in patients with RPC. We could learn MRI for patients with RPC reveals inflammatory auricles and ET at the same time. The follow up MRI have not been obtained to check up how the findings of auricles and ET are changed; however, the inflammatory findings might be getting better as the clinical symptoms have been disappeared soon after therapeutic intervention.

#2. Why did the patient's MRI show such characteristics? What is the mechanism behind it?

# Response

ET comprise chondroid tissue in it. Relapsing polychondritis (RPC) is an inflammatory disease targeting systemic chondroid tissue so that ET should be an inflammatory site in patients with RPC. Namely, the mechanism of edematous and contrast enhancing of ET is an inflammation, which is a manifestation of RPC. We have discussed it in discussion session (Page 9, lines 9-11 of the clean revised version):

"Fluid collection in middle ear in patients with RPC could be due to ET dysfunction derived from inflammation of comprising cartilage as this case;

however, this have not been proven as a pathological examination is impossible at this site."

#3. References are too old, please add some references of recent years.

# Response

We added sentences in discussion session and added the references below (Page 8, lines 18-21 and page 10, lines 2-34 of the clean revised version):

"To the best of our knowledge, this is the first report of a patient with RP manifesting as enhanced and edematous ET on MRI by reviewing previous mass reports and imaging review[3-5,9,10]. The only head and neck lesion other than auricles and nasal cartilage was orbital involvement reported by Moore et al[11]"

- "1 Lekpa FK, Chevalier X. Refractory relapsing polychondritis: challenges and solutions. *Open Access Rheumatol* 2018; **10**: 1-11 [PMID: 29391837 DOI: 10.2147/oarrr.S142892]
- 2 Kent PD, Michet CJ, Jr., Luthra HS. Relapsing polychondritis. *Curr Opin Rheumatol* 2004; **16**: 56-61 [PMID: 14673390 DOI: 10.1097/00002281-200401000-00011]
- 3 Chen N, Zheng Y. Characteristics and Clinical Outcomes of 295 Patients With Relapsing Polychondritis. *J Rheumatol* 2021: [PMID: 34334365 DOI: 10.3899/jrheum.210062]
- Dion J, Costedoat-Chalumeau N, Sene D, Cohen-Bittan J, Leroux G, Dion C, Frances C, Piette JC. Relapsing Polychondritis Can Be Characterized by Three Different Clinical Phenotypes: Analysis of a Recent Series of 142 Patients. *Arthritis Rheumatol* 2016; **68**: 2992-3001 [PMID: 27331771 DOI: 10.1002/art.39790]
- 5 Shimizu J, Yamano Y, Kawahata K, Suzuki N. Relapsing polychondritis patients were divided into three subgroups: patients with respiratory involvement (R subgroup), patients with auricular involvement (A subgroup), and overlapping patients with both involvements (O subgroup), and each group had distinctive clinical characteristics. *Medicine* (*Baltimore*) 2018; **97**: e12837 [PMID: 30334986 DOI: 10.1097/MD.0000000000012837]
- Rampelberg O, Gerard JM, Namias B, Gerard M. ENT manifestations

of relapsing polychondritis. *Acta Otorhinolaryngol Belg* 1997; **51**: 73-77 [PMID: 9241371]

- Bachor E, Blevins NH, Karmody C, Kühnel T. Otologic manifestations of relapsing polychondritis. Review of literature and report of nine cases. *Auris Nasus Larynx* 2006; **33**: 135-141 [PMID: 16427754 DOI: 10.1016/j.anl.2005.11.020]
- 8 McAdam LP, O'Hanlan MA, Bluestone R, Pearson CM. Relapsing polychondritis: prospective study of 23 patients and a review of the literature. *Medicine (Baltimore)* 1976; **55**: 193-215 [PMID: 775252]
- 9 Lin DF, Yang WQ, Zhang PP, Lv Q, Jin O, Gu JR. Clinical and prognostic characteristics of 158 cases of relapsing polychondritis in China and review of the literature. *Rheumatol Int* 2016; **36**: 1003-1009 [PMID: 26951051 DOI: 10.1007/s00296-016-3449-8]
- Thaiss WM, Nikolaou K, Spengler W, Spira D, Xenitidis T, Henes J, Horger M. Imaging diagnosis in relapsing polychondritis and correlation with clinical and serological data. *Skeletal Radiol* 2016; **45**: 339-346 [PMID: 26490679 DOI: 10.1007/s00256-015-2270-x]
- Moore GH, Rootman DB, Roybal CN, Goldberg RA. Orbital Relapsing Polychondritis: A Unique Presentation, Complication, and Treatment. *Ophthalmic Plast Reconstr Surg* 2016; **32**: e34-36 [PMID: 25072220 DOI: 10.1097/IOP.0000000000000222]

# Round 2

#### **Reviewer 1:**

"After the author's modification, this article has basically reached the acceptable level."

# Response

Thank you.

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