CASE REPORT

Occurrence of relapsing polychondritis with a rising cANCA titre in a cANCA-positive systemic and cerebral vasculitis patient

Georg Mattiassich,¹ Markus Egger,² Georg Semlitsch,³ Franz Rainer⁴

¹Trauma Center Linz,
Linz, Austria

²Department of Internal
Medicine,
Diakonissenkrankenhaus
Schladming, Schladming,
Austria

³MED-CENTER Judenburg,
Judenburg, Austria

⁴Department of Internal
Medicine, Hospital
Barmherzige Brüder,
Graz-Eggenberg, Bergstrasse
27, 8020, Graz, Austria

Correspondence to

Dr Georg Mattiassich,
georg.mattiassich@gmx.at

SUMMARY

Relapsing polychondritis (RP) is a relatively rare disorder, with a high death rate that affects cartilaginous structures. RP can be either primary or secondary as part of autoimmune syndromes. We present a case of RP in a 49-year-old man suffering from cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) positive vasculitis, admitted to our hospital with red swollen left ear and painful sternoclavicular joint and larynx. The patient was in remission from the vasculitis but manifested a high cANCA titre indicating vasculitis activity. With his high cANCA titre vasculitis, full manifestation of RP concomitantly occurred. After a successful cortisone treatment for RP, the patient received cyclophosphamide treatment for his vasculitis that resulted in a decrease in cANCA titre and full remission of his RP symptoms.

BACKGROUND

Relapsing polychondritis (RP) is a relatively rare recurrent disorder affecting cartilaginous structures such as ear and nose elastic cartilages, peripheral joints hyaline cartilages or the tracheobronchial cartilage.^{1–2} Inflammation can also occur at other proteoglycan-rich structures such as the inner ear, eyes, blood vessels and heart.³ The cause of RP is unknown, probably the immune system plays a pathogenetic role in the RP aetiology.⁴ RP can be either primary or secondary as part of an autoimmune syndrome.^{5–6} Diagnosis is based on clinical criteria described by McAdam *et al*¹ as no specific clinical tests are currently available. In 1979, Damiani and Levine⁷ modified these criteria to include histological findings (box 1) for early diagnosis of the disease. An early diagnosis is of imperative importance owing to the fact that RP is a potentially life-threatening debilitating disorder, especially with respiratory manifestations.⁸ Although corticosteroids are the therapy of choice, still side effects limit a long-term cortisone therapy. Immune modulation with methotrexate, cyclophosphamide or mycophenolate mofetil^{9–10} among others seem to be beneficial in the treatment of RP as an adjuvant therapy to corticosteroids.

CASE PRESENTATION

We present a case of a 49-year-old man suffering from cANCA-positive cerebral vasculitis for 2 years, which initially started with non-destructive joint pain and effusion of the left ankle. The diagnosis was based upon a cANCA high titre in serum

and histologically proven glomerulonephritis with focal and segmental glomeruloscleroses. A remission of the disease was induced by six parenteral cyclophosphamide cycles. As the cANCA titre was normal and the patient showed no clinical signs of the vasculitis. This was followed by a mycophenolate mofetil therapy. Despite intensive therapy, a rising cANCA titre was detected 1 year later. The patient was complaining of painful sternoclavicular joint and dizziness. The sternoclavicular joint was not swollen. The patient gave a history of left ankle joint pain. In February 2007, the patient was admitted to our hospital with red and swollen left ear with suspicious erysipelas of the ear that was treated with antibiotics. The left ear lobe was not affected by the inflammation (figure 1). He also had pain in the right ear with no obvious signs of inflammation. An RP diagnosis was reached based on his symptoms of night dyspnoea, pain on pressure in the larynx, parasternal pain and chondritis of the left ear. Pulmonary function tests revealed possible extrathoracic tracheal stenosis. A biopsy of the left ear revealed unspecific inflammatory signs that could be attributed to RP (figure 2). Laryngoscopy and CT scan were normal. Bone scintigraphy revealed increased Technetium-PDP uptake in the sternocostal region and in the sternoclavicular joint. An MRI of ankle and CT of the sternoclavicular joint revealed no pathological findings (figure 3). According to the clinical criteria of Damiani and Levine,⁷ we diagnosed RP as a secondary manifestation of the cANCA-positive vasculitis.

The patient was treated with 50 mg/day cortisone for 1 week, which led to a decline of C reactive protein and erythrocyte sedimentation rate serum inflammatory markers. The inflammation of the left ear was completely resolved within few days of the therapy. Pulmonary function tests were normal. The patient showed no signs of dyspnoea. The patient started a cyclophosphamide therapy as his MRI revealed progressive cerebral vasculitis (figure 4) that responded to the therapy. He was discharged from the hospital as his condition improved.

DISCUSSION

RP is commonly encountered with different conditions such as vasculitis, connective tissue diseases and arthropathies.⁸ C-ANCA are characterised by perinuclear neutrophilic pattern and mainly recognise myeloperoxidase. RP is probably immune-mediated owing to the high incidence of autoimmune disorders

To cite: Mattiassich G, Egger M, Semlitsch G, *et al*. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-008717

Box 1

- ▶ Diagnostic criteria based on McAdam *et al* and Damiani and Levine
 - Initial criteria (at least three of the criteria must be met to establish the diagnosis)
 - Recurrent chondritis of both auricles
 - Non-erosive inflammatory arthritis
 - Chondritis of the nasal cartilage
 - Ocular inflammation
 - Chondritis of the larynx, trachea or both
 - Damage to the cochlea, vestibule or both, with hearing loss and tinnitus or vertigo or both
- ▶ Modified criteria
 - Three or more of the initial criteria, with no histological confirmation necessary
 - Chondritis in two or more anatomical locations with a response to corticosteroids, dapsone or both

in patients with RP and the occasional presence of collagen type II autoantibodies.¹¹ The combination of vasculitis and RP is encountered in as high as 56% in McAdam's series with considerably worse prognosis.¹

In contrast to infections or other inflammatory diseases of the ear, RP spares the inferior lobe, which lacks cartilage. Typically, the cartilaginous parts of the ear (the helix, antihelix, tragus and external auditory canal) are inflamed, tender, painful and swollen; often bilateral and hearing loss occurs in nearly 50% of the cases.

Although there are no pathognomonic findings in cartilage histology, basophilic staining of the cartilage matrix, perichondral round-cell infiltration or cartilage destruction with fibrous replacement can be of diagnostic value. In our case, histological findings of the ear cartilage according to Damiani and Levine criteria seem to be useful in RP diagnosis, clinical involvement of the larynx and improvement under the cortisone therapy as well as the combination with vasculitis and non-destructive arthritis fulfilled the diagnostic criteria.⁷

Bone scintigraphy can be helpful in identifying atypical cases that can be histopathologically confirmed, especially in patients with chest pain or pain in the thyroid cartilage as it was the case in this patient.¹²

The respiratory tract is involved in approximately 50% of the patients. Clinical symptoms include cough, wheezing, stridor or dysphonia. Other features of respiratory tract involvement include tenderness over the thyroid and laryngeal cartilage and recurrent respiratory infections. Pulmonary function tests show

Figure 1 Red and swollen left ear.

Figure 2 H&E ×40: unspecific inflammatory signs.

plateaus on the flow-volume loops that suggest upper-airway obstruction. Involvement of the cartilaginous structures of the respiratory tract is one of the more serious complications of RP and accounts for 10–50% of RP-related mortality. Corticosteroids in the patient resulted in normal pulmonary function tests.

Figure 3 Bone-scintigraphy with increased Technetium-PDP uptake in the sternocostal region and in the sternoclavicular joint.

Figure 4 T2-weighted cerebral MRI revealing progressive cerebral vasculitis.

RP is considered by some as a separate comorbidity unrelated to vasculitis.¹³ However, in this patient the rising cANCA titre was directly related to the initial manifestation of RP, which probably indicates that RP arises as a secondary manifestation of vasculitis. This is of clinical importance in our case report as his RP symptoms were resolved as a result of his vasculitis-oriented therapy.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 McAdam LP, O'Hanlan MA, Bluestone R, *et al.* Relapsing polychondritis: prospective study of 23 patients and a review of the literature. *Medicine* 1976;55:193–215.
- 2 Lahmer T, Treiber M, von Werder A, *et al.* Relapsing polychondritis: an autoimmune disease with many faces. *Autoimmun Rev* 2010;9:540–6.
- 3 Tesche S, Wenzel S, Sagowski C. Relapsing polychondritis—a case report and review of the literature. *Laryngo- Rhino- Otologie* 2005;84:352–6. Polychondritis recidivans et atrophicans—ein Fallbericht und Literaturübersicht.
- 4 Gergely P Jr, Poor G. Relapsing polychondritis. *Best Pract Res Clin Rheumatol* 2004;18:723–38.
- 5 Handrock K, Gross WL. Relapsing polychondritis as a secondary phenomenon of primary systemic vasculitis. *Ann Rheum Dis* 1993;52:895–7.
- 6 Ullrich S, Gottschlich S. Secondary polychondritis in Wegner's disease. *Z Rheumatol* 2006;65:69. Sekundäre Polychondritis bei Morbus Wegener.
- 7 Damiani JM, Levine HL. Relapsing polychondritis—report of ten cases. *Laryngoscope* 1979;89(6 Pt 1):929–46.
- 8 Trentham DE, Le CH. Relapsing polychondritis. *Ann Intern Med* 1998;129:114–22.
- 9 Goldenberg G, Sanguenza OP, Jorizzo JL. Successful treatment of relapsing polychondritis with mycophenolate mofetil. *J Dermatol Treat* 2006;17:158–9.
- 10 Edrees A. Relapsing polychondritis: a description of a case and review article. *Rheumatol Int* 2011;31:707–13.
- 11 Foidart JM, Abe S, Martin GR, *et al.* Antibodies to type II collagen in relapsing polychondritis. *N Engl J Med* 1978;299:1203–7.
- 12 Shi XH, Zhang FC, Chen LB, *et al.* The value of 99mTc methylene diphosphonate bone scintigraphy in diagnosing relapsing polychondritis. *Chin Med J* 2006;119:1129–32.
- 13 Papo T, Piette JC, Le Thi Huong D, *et al.* Antineutrophil cytoplasmic antibodies in polychondritis. *Ann Rheum Dis* 1993;52:384–5.

Learning points

- ▶ Relapsing polychondritis (RP) is a relatively rare disorder associated with inflammation in cartilaginous structures and other tissues.
- ▶ RP can be either a primary disease or a secondary phenomenon of autoimmune syndromes such as vasculitis, connective tissue diseases or arthropathies.
- ▶ Typically the cartilaginous parts of the ear are inflamed, tender, painful and swollen, often bilateral and hearing loss occurs in nearly 50% of the cases.
- ▶ Diagnosis is based on clinical criteria as no specific clinical tests are currently available.
- ▶ An early diagnosis is of prime importance owing to the fact that RP is a potentially life-threatening disease.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow