

Refractory relapsing polychondritis, responsive to IL-6 blockade

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Abstract

Relapsing polychondritis is a rare immune-mediated condition, causing inflammation of cartilage and other structures, and associated with a high mortality. Its heterogenic nature and rarity raises a number of diagnostic and treatment challenges.

Keywords

Relapsing polychondritis; scleritis; IL-6 blockade; tocilizumab.

Case report

A 51-year-old Caucasian woman presented to the rheumatology clinic with refractory scleritis and inflammatory polyarthritis. She developed painful erythematous ears (Fig. 1) and pain across the bridge of the nose, and was diagnosed with relapsing polychondritis. Her chest radiograph was normal.

Over 3 years, treatment with mycophenolate mofetil, methotrexate, sulfasalazine, azathioprine, dapsone and colchicine was trialed but found to be ineffective.

There was a response to oral prednisolone, but daily doses of 40–60 mg were required for disease control and these doses resulted in severe weight gain, glucocorticoid-induced diabetes and poor diabetic control.

Two emergency admissions with severe costochondritis resulted in treatment with pulsed intravenous methylprednisolone, with only short-term benefit. Oral cyclophosphamide was commenced (3 mg/kg), but this was not tolerated due to gastrointestinal side effects.

There was no history of stridor, and chest radiographs and pulmonary function tests were normal. Clinical and echocardiographic examinations did not reveal any cardiac involvement. A computed tomography-positron emission tomography scan did not show any large vessel vasculitis.

Treatment with interleukin (IL)-6 blockade was commenced rather than anti-tumor necrosis factor (TNF) therapy because we could not secure funding for the latter. Tocilizumab was given (8 mg/kg) at monthly intervals, with a response after each infusion (Table 1).

The patient reported ongoing symptomatic benefit and treatment with IL-6 blockade was continued.



Fig. 1. Right ear chondritis during a flare.

Table 1. Clinical markers of response to treatment, at monthly intervals

	Baseline	Month 1	Month 2	Month 3	Month 4	Month 5
Average prednisolone dose (mg)	50	15	35	20	20	50
C-reactive protein (mg/l)	29	5	<5	<5	<5	<5
Erythrocyte sedimentation rate (mm/h)	25	2	2	5	4	5
VAS chest wall pain	80	30	0	0	20	50
VAS ear pain	85	45	15	30	20	20
Swollen joint count	14	4	12	8	4	1
Tender joint count	47	8	53	24	8	1
Functional score (HAQ)	3.125	2.25	3.125	2.25	2.13	2.57

VAS, visual analogue scale; HAQ, Health Assessment Questionnaire.

Discussion

This patient initially presented with refractory scleritis and inflammatory arthritis. One third of cases of relapsing polychondritis coexist with connective tissue disease or a myelodysplastic syndrome^[1].

As this patient fulfilled four of McAdam's criteria^[2,3] for diagnosis on clinical grounds, further histological confirmation was not obtained.

Due to the rarity of relapsing polychondritis, there are no clinical trials to determine the efficacy of treatments. Treatment decisions are based on case reports and small series and consensus. Non-steroidal anti-inflammatory drugs (NSAIDs), dapsone, colchicine and prednisolone have been used with good effect. Visceral involvement requires aggressive escalation of therapy, and methotrexate, azathioprine, cyclosporine and cyclophosphamide have been used for refractory cases. Response to biological agents is limited to case reports, the majority of these being TNF blockade with infliximab and adalimumab.

Relapsing polychondritis is thought to be driven by a cell-mediated immune response. Although normal serum TNF- α and IL-6 levels have been demonstrated in active relapsing polychondritis^[2], there is a growing body of case reports showing response to TNF blockade, and more recently, tocilizumab has been used for relapsing polychondritis refractory to conventional immunosuppressants and TNF blockade. It has been reported to allow reductions in steroid dosage, with nose, ear and tracheal inflammation^[3,4] and aortitis^[5].

Teaching points

- Relapsing polychondritis is a rare condition that presents with painful inflammation of the cartilage of the ear and nose, but can also affect the eyes, joints, respiratory tract, cardiac valves and aortic root. It has a reported mortality of 20–30%.
- One third of cases coexist with connective tissue disease or a myelodysplastic syndrome^[1].
- Corticosteroids remain the mainstay of treatment, but conventional immunosuppressants and biological agents are used for refractory cases.

References

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