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CASE REPORT

Limited auricular relapsing polychondritis in a child treated successfully with infliximab

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SUMMARY

Relapsing polychondritis (RP) is a rare progressive and destructive multisystem disorder characterised by recurrent inflammation of cartilaginous structures. It is a rare disease in paediatrics compared with adults. In children, the diagnosis is either delayed or overlooked due to low incidence. Auricular chondritis has been described in more than half of paediatric cases with RP. However, isolated auricular chondritis has not been reported as the only presentation of pediatric-onset RP. We described a lad who presented with isolated auricular chondritis, which is refractory to conventional treatment, including glucocorticoids and methotrexate as steroid-sparing agent. Remission of his disease's relapses was sustained with infliximab. Limited auricular involvement as a presenting feature of RP in the absence of systemic association is very rare in children. We describe a case of successful use of infliximab on limited auricular chondritis disease.

BACKGROUND

Relapsing polychondritis (RP) is a rare progressive and destructive multisystem disorder characterised by recurrent inflammation of cartilaginous structures. The disease commonly affects the ear, nose, larynx, trachea, bronchi, peripheral joints, eye, heart and skin. It is a rare disease in paediatrics compared with adults. In children, the diagnosis is either delayed or overlooked due to low incidence.[1] Less than 10% of reported cases occur in children and adolescents.[2] French series of 10 pediatric-onset RP cases revealed that the most frequent manifestations are auricular chondritis followed by arthritis. The same author did a systematic analysis of 37 reported paediatric cases and found auricular chondritis in 58% and arthritis in 71%.[3] None of these cases has limited the disease to the auricles. Isolated auricular chondritis is a very rare presentation of pediatric-onset RP.

We report a paediatric case of RP with limited auricular involvement in the absence of systemic associations treated with infliximab.

CASE PRESENTATION

We describe an 18-year-old Egyptian boy who presented to the paediatric rheumatology clinic at the age of 13 with diffuse painful swelling and redness of the right auricle. Similar symptoms were reported a year earlier over the left ear and he received multiple courses of antibiotics that resulted in no improvement (figure 1A). Before attendance at the clinic, he has been started on systemic steroid by a dermatologist with a presumptive diagnosis of perichondritis.

There was no history of fever, no skin rash, no proceeding infection, no trauma and no allergy to food or drugs. There were no respiratory or musculoskeletal symptoms and review of systems was unremarkable.

INVESTIGATIONS

Over the course of 1 year prior to his presentation to rheumatology clinic, he was seen at different medical centres. Investigations were done for him, which included an inflammatory marker, chest CT with contrast, hearing test, eye examination, antinuclear antibodies (ANA), antineutrophil cytoplasmic antibodies, C3 and C4, collagen type II antibodies and urine analysis; they were all negative.

TREATMENT

By the time of his visit, he was on prednisone 45 mg/day which resulted only in a mild improvement of his symptoms; therefore, subcutaneous (S/C) methotrexate 20 mg/week with folic acid supplement was added. Subsequently, prednisone was tapered over 4 months to 7.5 mg/day. Further steroid taper was not possible due to a flare of swelling and a redness of the right auricle. Following two episodes

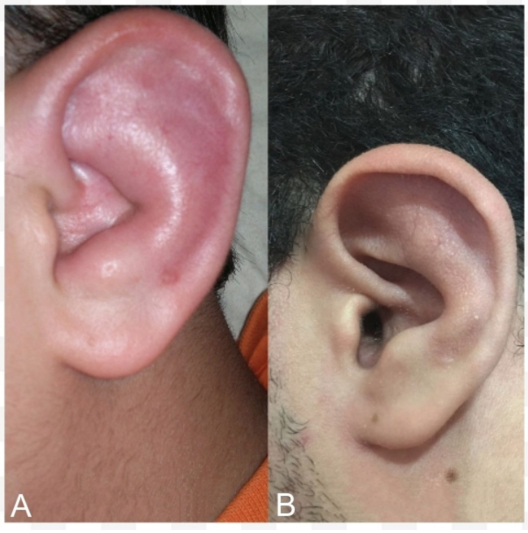

Figure 1 Left ear of the patient. (A) At the age of 13 years, it was swollen, warm, tender and erythematous before infliximab. (B) At the age of 18 years, normal after treatment.

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of flare up, adalimumab 40 mg S/C injections every 2 weeks were then introduced in May 2012. There was an improvement of the swelling for 4 months but the dose was interrupted due to family travel and he had a severe flare that necessitated pulse steroid and an increase in methotrexate to 25 mg S/C injection weekly. Despite resuming adalimumab, he continued to have a frequent flare up. An auricular biopsy was then done, which showed features of degeneration of chondroid tissue in the form of loss of chondroid basophilia, vacuolisation and degeneration of chondrocytes. Chondroid/connective tissue interface shows only a few lymphocytes. However, due to active clinical symptoms, he was switched to 6 mg/kg/dose of infliximab infusion, given at 0, 2, 4 then every 6 weeks. His symptoms resolved (figure 1B) and methotrexate was shifted to an oral dose of 20 mg po weekly and subsequently tapered down to 7.5 mg weekly. Prednisone was further tapered to 2.5 mg every other day but attempts to take him off were challenged by two minor flares that had been managed with an increase in prednisone for a short course.

OUTCOME AND FOLLOW-UP

Fortunately, no deformity over his ear cartilages was observed. The patient continues to have no evidence of eye, upper airway, major vessel, musculoskeletal and other cartilages or muscuocutaneous involvement. Surveillance laboratory, including complete cell counts, liver enzymes, renal function and ANA, remains unremarkable. At the age of 18 years, the patient moved back to Egypt to join a medical school.

DISCUSSION

Pediatric-onset RP is rare and occurs in 5%–10% of the reported cases.[2 3]

Belot *et al* described 10 patients and conducted a systematic review of another 37 reported cases of RP in children. The most frequent presenting features of those cases were auricular chondritis and arthritis.[3] Knipp *et al* reviewed 13 paediatric cases of RP and the incidence of auricular chondritis was 61%.[4] Isolated auricular chondritis might be the only presenting clinical sign for RP, characterised by inflammation of the cartilaginous portion of the pinna, with pain, redness, swelling or tenderness, leading to a nodular or verrucous appearance and becoming soft and flabby after repeated attacks, or sometimes after a single prolonged episode. RP typically spares the auricular lobe, which has no cartilage. Even though the presentation of our patient is not usual and does not fulfil any of the three known set criteria for RP,[5–7] it was easy to exclude other differential diagnoses such as infections and allergy due to bilateral involvement and sparing ear lobe as well as the remitting-relapsing nature of the disease.

The diagnosis of RP in children is delayed for 5 years. The time since the left ear symptoms started in our patient to his presentation to rheumatology clinic with right ear inflammation was 2 years. Cartilage biopsy is rarely conducive, and most histopathological findings are not specific.[3 8] In our case, it was not contributive in diagnosis, neither did it change the plan for management.

All types of cartilage may be involved, such as the hyaline cartilage of peripheral joints and the fibrocartilage of extra-articular sites, as well as proteoglycan-rich tissues including the media of the arteries, the conjunctiva and sclera of the eye.[9] Musculoskeletal examination, ophthalmological examination, hearing assessment, echocardiogram and CT angiogram were non-revealing in this child.

There is a lack of randomised therapeutic trials; therefore, the treatment of RP remains mainly empirical.[10] Corticosteroids are the main form of treatment and, in patients with a sustained or refractory disease, immunosuppressive agents such as cyclophosphamide, azathioprine, cyclosporine, methotrexate and mycophenolate mofetil have been used as steroid-sparing agents with varying results. The need for other drugs to prevent the unwanted side effect of long-term steroid is paramount. Unfortunately, there is no rigorous clinical research to support the use of new therapeutic modalities including biological agents.

Infliximab was used to induce and maintain remission in a 14-year-old girl with severe saddle nasal deformity.[11] Transient response to infliximab was reported in another 14-year-old girl with saddle nose who responded well to anakinra.[12] de Oliveira *et al* described a young girl with persistent and destructive arthritis who had partial response infliximab and etanercept.[13] Other indications that infliximab has been used in treating children with RP include severe episcleritis, pyoderma gangrenosum and tracheal chondritis.[12 14 15]

Complications of RP include vanishing of the auricular cartilage resulting in drooping of the pinna, which becomes floppy and has a cauliflower appearance.[10]

Hearing impairment due to a conductive hearing loss secondary to external meatus obstruction or damage to the cochlea and vestibular system may occur, leading to a sensorineural hearing loss.[16]

In conclusion, pediatric-onset RP is a very rare disorder. Isolated auricular chondritis is even rarer, which makes the diagnosis challenging and could contribute to its delay. Biological treatment might be justified in limited cases to prevent morbidity. In our patient, a good therapeutic response was obtained with infliximab and it prevented auricular cartilage damage and resulted in no hearing deficit.

Learning points

- Pediatric-onset relapsing polychondritis (RP) is a very rare disorder.
- The treatment of RP remains mainly empirical due to rarity and lack of clinical trials.
- Different biological agents have been reported in the treatment of RP with variable response.

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REFERENCES

1 Herrera I, Mannoni A, Altman RD. Relapsing polychondritis: commentary. *Reumatismo* 2002;54:301–6.
2 Trepel RJ, Lipnick RN, D'Angelo L. Relapsing polychondritis in an adolescent. *J Adolesc Health Care* 1989;10:557–60.
3 Belot A, Duquesne A, Job-Deslandre C, *et al*. Pediatric-onset relapsing polychondritis: case series and systematic review. *J Pediatr* 2010;156:484–9.
4 Knipp S, Bier H, Horneff G, *et al*. Relapsing polychondritis in childhood-case report and short review. *Rheumatol Int* 2000;19:231–4.

5 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.
6 Damiani JM, Levine HL. Relapsing polychondritis-report of ten cases. *Laryngoscope* 1979;89:929???946–46.
7 Michet CJ, McKenna CH, Luthra HS, *et al*. Relapsing polychondritis. Survival and predictive role of early disease manifestations. *Ann Intern Med* 1986;104:74–8.
8 Valenzuela R, Cooperrider PA, Gogate P, *et al*. Relapsing polychondritis. Immunomicroscopic findings in cartilage of ear biopsy specimens. *Hum Pathol* 1980;11:19–22.
9 Cantarini L, Vitale A, Brizi MG, *et al*. Diagnosis and classification of relapsing polychondritis. *J Autoimmun* 2014;48-49:53–9.
10 Mathian A, Miyara M, Cohen-Aubart F, *et al*. Relapsing polychondritis: A 2016 update on clinical features, diagnostic tools, treatment and biological drug use. *Best Pract Res Clin Rheumatol* 2016;30:316–33.
11 Bell D, Wright D, Witt PD. Durability of nasal reconstruction in an adolescent with relapsing polychondritis treated with infliximab. *Plast Reconstr Surg* 2007;120:1087–8.
12 Buonuomo PS, Bracaglia C, Campana A, *et al*. Relapsing polychondritis: new therapeutic strategies with biological agents. *Rheumatol Int* 2010;30:691–3.
13 de Oliveira SK, Fonseca AR, Domingues RC, *et al*. A unique articular manifestation in a child with relapsing polychondritis. *J Rheumatol* 2009;36:659–60.
14 Ghosn S, Malek J, Shbaklo Z, *et al*. Takayasu disease presenting as malignant pyoderma gangrenosum in a child with relapsing polychondritis. *J Am Acad Dermatol* 2008;59(5 Suppl):S84–S87.
15 Terrier B, Aouba A, Bienvenu B, *et al*. Complete remission in refractory relapsing polychondritis with intravenous immunoglobulins. *Clin Exp Rheumatol* 2008:26:136–8.
16 Trentham DE, Le CH, Ch L. Relapsing polychondritis. *Ann Intern Med* 1998;129:114–22.