

Relapsing Polychondritis: Case Report and Review of the Literature

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Abstract:

Relapsing polychondritis is a rare, severe, episodic and progressive inflammatory disorder of unknown aetiology and autoimmune pathogenesis, causing recurrent inflammation of involving cartilaginous structures, predominantly those of the ears, nose, and laryngotracheobronchial tree. Rarity of this disease, array of many possible presenting symptoms, lack of specific diagnostic investigation and episodic nature often causes significant delay in diagnosis. We are reporting a 55 year old woman presenting with recurrent episodes of pain, swelling, redness of nose, and both ear resulting in depressed nasal bridge and floppy pinna. Relapsing polychondritis was diagnosed and she was treated with systemic corticosteroid successfully.

Key words: Relapsing polychondritis

Introduction:

Relapsing polychondritis (RP) is a rare multisystem disease of unknown aetiology initially described by Jaksch-Wartenhorst in 1923. The patient described by Wartenhorst experienced an 18-month course of progressive degeneration of the peripheral joints, external ears, nasal septum, external auditory canals, inner ear, and epiglottis. He termed this condition polychondropathia.¹ In 1960, Pearson, Kline, and Newcomer reviewed 12 such cases. The term relapsing polychondritis was introduced in that review.² Relapsing polychondritis (RP) is characterized by recurrent inflammation occurring in cartilaginous structures, resulting in tissue destruction and their subsequent replacement by fibrous connective tissue. All types of cartilage may be involved, such as elastic cartilage of ears and nose, the hyaline cartilage of peripheral joints, the fibrocartilage of axial skeleton, and the cartilage of the tracheobronchial tree. RP can also affect other proteoglycan rich structures such as eye, heart, blood vessels and inner ear.^{3,4} In clinical reports and reviews, relapsing polychondritis is reported to be a rare disease. By 1997, only 600 cases had been reported

worldwide. The annual incidence in Rochester, Minnesota, was noted to be 3.5 cases per million population. International incidence of RP is unknown.⁴

The etiology of relapsing polychondritis is unknown; however, the pathogenesis is autoimmune. The evidence for an autoimmune etiology includes pathological findings of infiltrating T cells, the presence of antigen-antibody complexes in affected cartilage, cellular and humoral responses against collagen type II and other collagen antigens, and the observation that immunosuppressive regimens most often suppress the disease. Researchers have found that antibodies to type II collagen are present during acute relapsing polychondritis episodes and that the levels correlate with the severity of the episode.⁵ The hypothesis of an autoimmune etiology for relapsing polychondritis is also supported by the high prevalence of other autoimmune disorders found in patients with relapsing polychondritis. McAdam et al reported that 25%-35% of patients with relapsing polychondritis had a concurrent autoimmune disease.⁶ Autoimmune conditions associated with relapsing polychondritis includes rheumatoid arthritis, SLE, thyroid disease, cutaneous leucocytoclastic vasculitis, systemic vasculitis, Sjogren syndrome, inflammatory bowel disease etc.

Diagnostic criteria for relapsing polychondritis were proposed first by McAdam et al and have been modified several times. Biopsy is required only if clinical criteria are in question. According to McAdam et al criteria, 3 of 6 following clinical features necessary for diagnosis: a) bilateral auricular chondritis, b) nonerosive seronegative inflammatory polyarthritis, c) nasal chondritis, d) ocular inflammation, e) respiratory tract chondritis, f) audiovestibular damage.⁶ Damiani and Levine reviewed and modified the criteria of McAdam et al. One of three following conditions is necessary

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for diagnosis: a) three McAdam et al criteria, b) one McAdam et al criterion plus positive histology results, c) two McAdam et al criteria plus therapeutic response to corticosteroid or dapsone therapy.⁷ According to Michet et al criteria, 1 of 2 following conditions are necessary for diagnosis: a) proven inflammation in 2 of 3 of the auricular, nasal, or laryngotracheal cartilages; b) proven inflammation in 1 of 3 of the auricular, nasal, or laryngotracheal cartilages plus 2 other signs including ocular inflammation, vestibular dysfunction, seronegative inflammatory arthritis, and hearing loss.⁸

The array of possible presenting symptoms and the episodic nature of relapsing polychondritis may result in a significant delay in diagnosis. In a review of 66 patients, the elapsed time from patient presentation for medical care for a related symptom to diagnosis was reported to be 2.9 years.⁴

Case report:

Mrs. M, a 55 year old lady was admitted in Rajshahi Medical College Hospital with the complaints of recurrent episodes of pain, and swelling of nose for 9 months; pain, redness and swelling of both ear for 6 months; recurrent attacks of redness, and watering of both eyes for 6 months; depressed nasal bridge for 3 months and pain at multiple joints for 3 months. Her nasal pain was moderate in intensity, and was associated with redness & swelling of overlying skin. She didn't give any history of trauma, epistaxis, nasal congestion, or rhinorrhea. Each episode of nasal pain and swelling subsided spontaneously after about 10 days. These episodes occurred 6 times in the last 9 months and her nasal bridge gradually became depressed. She also complained of four episodes of bilateral pain and swelling of external ear, which first occurred 6 months back. It was associated with reddish discoloration of overlying skin of helix of both ear, but both ear lobes were spared. She didn't complain of hearing loss, vertigo, tinnitus, nausea, or vomiting. This episodes occurred at variable intervals leading to deformed, floppy ear; more marked in the left. She gave history of recurrent episodes of redness, watering & foreign body sensation of both eyes without any visual impairment. For last 3 months, she was having pain and mild swelling involving both ankles, right wrist, left elbow and right 1st MCP joints. It was associated with morning stiffness lasting for about an hour. She did not give any history of burning sensation during micturition or diarrhea in last few weeks before this episode. She had occasionally taken some NSAID's for her problems. She did not give any history of cough, haemoptysis, breathlessness, haematuria, skin rash, chest pain, or swelling of body. On general examination, she was mildly anaemic, there was depression of nasal bridge, erythematous, deformed external ear with bilateral floppy pinna, more marked

in the left. Her ankles, right wrist, and both elbow joints were swollen, and tender on palpation. Movements of these joints were restricted in all directions because of pain. There were also tenderness of 2nd, 3rd, and 4th costochondral joints bilaterally. Other joints and spine were normal. There was no tenderness over laryngeal cartilages. Her hearing was not impaired in any ear. All other system examination was normal. Investigations showed Hb: 9.3 gm/dl, TC: 9,000/mm³, N- 82%, L-14%, E-01%, M-03%, B-00% ESR : 130 mm in 1st hour, PBF was commented as normochromic normocytic anaemia with high ESR. Her CRP was 61 mg/L (normal < 6), RA test (Elisa) negative, anti-CCP negative, ANA Negative, C-ANCA negative, CXR P/A was normal, no erosion was noted in the X-Ray of right wrist joint. RBS, urine RME, serum creatinine, pure tone audiometry was also normal. Slit skin smear for AFB was negative. Biopsy from the ear cartilage was not done because it was not mandatory for the diagnosis and also because of the patient's refusal. Our patient fulfilled three McAdam et al criteria: bilateral auricular chondritis, nonerosive seronegative inflammatory polyarthritis, nasal chondritis. Her history of ocular involvement was also suggestive, but it was not included because there was no objective sign at the time of admission. She also fulfilled the Damiani & Levine and Michet et al criteria, thus confirming our diagnosis of relapsing polychondritis. As she had already fulfilled three criteria, histological proof was not deemed necessary. She was treated with 40 mg of prednisolone/ day initially, which was gradually tapered and maintained on 10 mg/day. She was doing well when last seen.



Fig-1: showing depressed nasal bridge



Fig.-2: showing floppy left ear

Discussion:

Relapsing polychondritis (RP) is a rare autoimmune disease of unknown aetiology with characteristic inflammation of the cartilaginous tissues. The peak age of onset ranges from 40-50 years, which is nearly concordant with our case. Disease onset of relapsing polychondritis is usually sudden and as the name implies there is recurrent inflammation of cartilages in the different parts of body. External ear involvement is the presenting symptom in almost all cases. RP typically attacks the cartilaginous portion of the pinna, sparing ear lobes, which lack cartilage. Auricular chondritis presents with pain, redness, violaceous discoloration, swelling and tenderness involving one or both ears. Episodes last a few days to weeks and patients recover from these episodes with or without treatment. Recurrent episodes of cartilage inflammation lead to tissue destruction and in case of ear, leads to floppy pinna. Conductive type hearing impairment may occur due to inflammatory closure of external auditory meatus, serous otitis media, eustachian tube obstruction. Sometimes, there is sensorineural hearing loss or vestibular dysfunction manifested as vertigo, dizziness, vomiting etc. The nasal chondritis is acute and painful and accompanied by a feeling of fullness over the nasal bridge. Mild epistaxis may be present. A saddle-nose deformity may develop in longstanding disease. The common ocular conditions seen in RP are episcleritis and scleritis, and uveitis. Relapsing polychondritis may involve any portion of the respiratory tree, including the distal bronchi. Tenderness to palpation may occur over the anterior trachea or thyroid cartilage. Chondritis weakens the tracheal cartilage rings, resulting in wheezing, dyspnea, cough, and hoarseness. The upper airways can eventually become stenosed and are replaced by collapsible fibrotic tissue. A

seronegative non-deforming arthritis is one of the McAdam diagnostic criteria. Most commonly, the arthritis is asymmetric, oligoarticular or polyarticular, non-deforming, and nonerosive. The ankles, elbow, wrists, proximal interphalangeal joints, metacarpophalangeal joints, and metatarsophalangeal joints are often involved, although any joint may be affected. The costochondral, sternoclavicular, and sternomanubrial joints may be involved.^{4,6,9} Laboratory abnormalities are nonspecific ranging from leukocytosis, thrombocytosis, chronic anaemia, raised ESR and elevated gammaglobulin levels. Low titers of rheumatoid factor and ANA can be detected.¹⁰ High titers of anti-collagen type II antibody have been reported.¹¹ Radiographic abnormalities may include tracheal stenosis and cartilaginous calcifications.¹²

No controlled trials of therapy for relapsing polychondritis (RP) have been published. The goal of treatment is to abate current symptoms and to preserve the integrity of cartilaginous structures. The mainstay of treatment is systemic corticosteroid therapy. Prednisone (20-60 mg/d) is administered in the acute phase and is tapered to 5-25 mg/d for maintenance. Severe flares may require 80-100 mg/d. Most patients require a low daily dose of prednisone for maintenance; however, intermittent administration of high doses during only flares of the condition is successful in rare cases. McAdam et al found that continuous prednisone decreased the severity, frequency, and duration of relapses.⁶ Immunosuppression with methotrexate, azathioprine, cyclosporine and cyclophosphamide may be needed in patients who do not respond to steroids. Surgical intervention may be needed in case of complications involving the respiratory tract like tracheal stenosis and tracheomalacia and stents may be required for tracheobronchial collapse.^{10,13} Case reports have described successful treatment with anti-tumor necrosis factor- α inhibitors infliximab, etanercept, and adalimumab. Anakinra, an IL-1 receptor antagonist; and rituximab, an anti-CD20 chimeric antibody, have also shown benefit.¹⁴ Five-year survival of upto 74% and 10-year survival of 55% has been reported in a study of 112 patients from Mayo clinic, 10 major causes of death being infections and systemic vasculitis.⁸

Conclusion:

Relapsing polychondritis, as the name implies, is the relapsing inflammation of the cartilaginous structures. It should always be considered when a patient presents with recurrent pain and swelling of ear, nose, laryngeal or tracheal cartilages; or when the patient presents with floppy ear or depressed nasal bridge.

Conflict of interest: None

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