Int. Adv. Otol. 2010; 6:(2) 295-297

CASE REPORT

Clinical Challenges and Management of Relapsing Polychondritis

Gul Ozbilen Acar, Muhammet Tekin, Devrim Bellek, Osman Ilkay Ozdamar, Berrin Gucluer

Goztepe Training and Research Hospital, Ear Nose Throath Department (GO, MT, DB, OO)
Goztepe Training and Research Hospital, Pathology Department, (BG)

Relapsing (recurrent) polychondritis (RP) is a rare inflammatory disease with clinical features of autoimmune, episodic and progressively outgoing character which can involve all cartilaginous structures. The most common involved cartilaginous structures are auricular and nasal septal cartilages in order. One of the most common finding is an arthritis which is non-erosive and not resulted with deformation. Involvement of other cartilaginous areas such as trachea, larynx, epiglottis, costal joints is widely encountered. Also, this disease can be seen in organs of eyes, aorta, heart, skin that are rich in proteoglycans. Diagnosis is generally done on clinical findings for RP.
In this paper; clinical course, histopathological features, diagnostic methods, and treatment modalities of this rarely seen disease has been discussed accompaniment with the current literature and in the sight of a case which has been diagnosed as having RP.

Submitted : 05 August 2009 *Accepted : 03 January 2010*

Introduction

Relapsing Polychondritis (RP) is a rarely seen, progressive autoimmune disease with unknown aetiology. Although this disease primarily affects cartilaginous tissues (auricula, nasal septum, laryngotracheobronchial system), inflammatory injury can also progress to non- cartilaginous body structures (kidneys, blood vessels) [1]. Diagnostic criterias of the disease were defined firstly by McAdam et al. in 1976 [2]. Although it has been explained as an autoimmune disease, and the pathogenesis has not been known exactly. Reasons of that are rarity of the disease, not constitution of appropriate animal models, and lack of specific laboratory findings.

Our case that is presented in this paper is a typical form of the disease with the classical findings of recurrent attacks of perichondritis and remission periods. These clinical findings made up the diagnosis of RP because of the fact that diagnosis is primarily done on clinical findings for RP. Also, dramatic healing with the usage of corticosteroids and anti chemotheraupathic agent (methotrexate) was observed in our patient.

Case Report

Fifty-three years old female patient applied to our clinic with complaints of severe ache, erythema and swelling of both auriculas, decline in hearing bilaterally, and both side of ear flow for a few weeks in March 2007 (Figure 1). In detailed anamnesis; the complaints of widespread joint pain, and wetting, erythema and burning of eyes were also present. The complaints have been repeated for 20 years with remission periods. The patient was given various medical treatments at remission periods, but she did not explain exactly which drugs were given to her previously.

In physical examination, both auricles were diffusely erythematous and edematous except lobules. Bilateral external ear meatus were obliterated and tympanic membranes were not observed. Nasal septal mucosa was observed as atrophic and dry. Bone conduction was measured bilaterally as 23 dB, air conduction at right ear was 35 dB, and at left ear 30 dB respectively on audiometric examination. Temporal bone magnetic resonance imaging (MRI) was normal. At WBC; Hct: % 31.8, Hgb: 11.1 g/dl, LDH and CK (4 folds) were elevated. ESR was 35 mm/h, and CRP (> 0.800 mg/dl) was elevated. ANA (+) and C3 was elevated (180 mg/dl). Other ANA related panel results were negative. Rheumatologic findings were found to be compatible with seronegative arthritis. In ophthalmologic examination; bilateral conjunctivitis, episcleritis and cataract had been detected. In Schirmer test, bilateral eye tears were reduced (<5mm). Flexible

Corresponding address:
Gul Ozbilen Acar
Ata 3/4 Daire: 249 Kat: 20 Sedef Cad. 38. Ada 34750 Atasehir/Kadıköy-İstanbul/TURKEY
Phone: +90 532 2669023; Fax: +90 216 4557777; E-mail: gulozbilenacar@gmail.com

bronchoscopic examination was performed by a Chest Disease specialist. Although trachea and right lung structures were normal, inflammated areas were observed at left bronchial system that had been confirmed as bronchitis by biopsy. Degenerative changes on aortic valve and limited cardiomegaly had been detected by echocardiographic examination at Cardiology consultation. Nevertheless left ventricular function of the patient was normal. Prominent matrix degeneration and formation of fibrous connective tissue were detected in hyaline cartilage tissue at auricular cartilage biopsy evaluation of the patient (Figure 2).

Diagnosis of RP was confirmed with bilateral characteristic auricular involvement, seronegative arthritis, and involvement of eye and lung. Treatment; prednisone dose of 64 mg/day was started and tapered in two weeks to 16 mg/day as a maintenance dose. Additionally methotrexate dose of 7.5 mg/week was started and elevated to 22.5mg/week as a maintenance dose. It was decided to continue prednisone and methotrexate maintenance dose for four months. Topical anti-inflammatory treatment was applied to both auricles and eyes. Ear complaints of the patient were improved approximately in one month (Figure 3). In audiometry, bilateral bone conduction thresholds were 23 dB HL. Air conduction thresholds were 32 dB HL and 28 dB HL on the right and left side respectively. Pronounced improvement was gained on eye and rheumatologic findings. The patient has still followed up by our clinic for approximately 2 years and her complaints were repeated 2 times. In both of the attacks, the patient presented with similar clinical and laboratory findings, and the same medical therapy was applied to her. The both recurrences were succesfully cured with our treatment. The patient was free of disease in our follow-up for the last 6 months.

Discussion

Prevalence is pointed out as [3-5] per million at different studies in USA. Slightly female predominance exists, and fifth decade is the most affected period [3]. Nevertheless the etiology is not well-known, High incidence of HLA-DR4 antigens and low incidence of HLA-DR[6] antigens were detected in the patients [4]. The relapsing polychondritis (RP) is a rare, multisystemic and potentially fatal disease. The pathogenesis and optimal therapeutic approach is poorly understood. The disease is characterized by episodic inflammation of cartilaginous structures such as auricular, nasal and laryngotracheal. But the immune damage can spread to involve noncartilaginous tissues like the kidney, blood vessels, and so forth. The manifestations of the disease can take many different forms [5].

Differential diagnosis of RP is settled with the exclusion of other diseases and its diagnosis is primarily based on clinical investigation. Because of its rarity and attribution of symptoms to other coexistent disease, 1 or 2 years may elapse before the

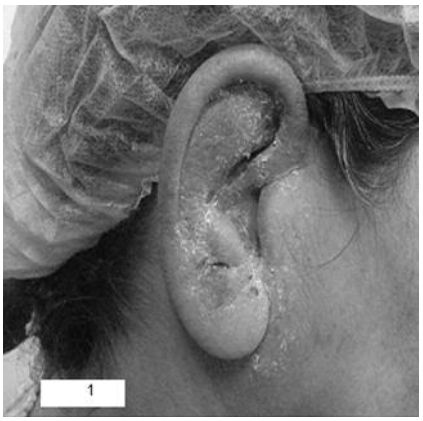

Figure 1. Pretreatment appearance of the rigth auricle of the patient.

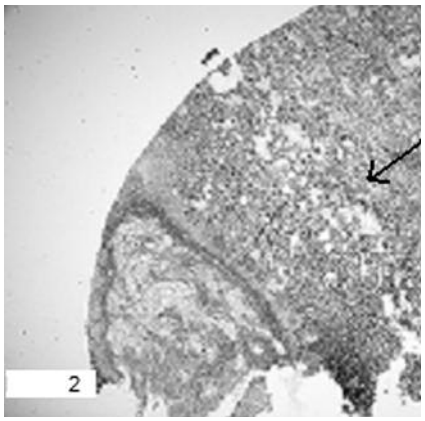

Figure 2. Histopathology of the right auricular cartilage biopsy. Extensive degenerative changes in cartilage tissue, missing of the matrix basophilia, markedly effected marginal chondrocytes, missing of the lacuna, degeneration of elastic fibres and irregularly increased collagen fibres are shown in specimen (HE X 100).

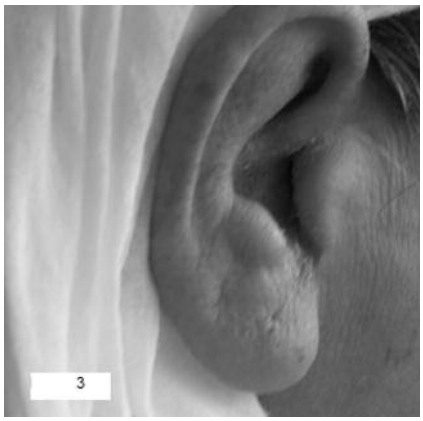

Figure 3. Posttreatment appearance of the rigth auricle.

diagnosis is made. Auricular chondritis is relatively unique to relapsing polychondritis and eventually occurs in nearly 90% of cases. However, necrotizing external otitis resulting from Pseudomonas aeruginosa should always be considered in the patient with a unilateral inflamed ear. Various diseases can cause inflammatory or granulomatous lesions, including Wegener granulomatosis, sarcoidosis, tuberculosis, amyloidosis, and rheumatoid arthritis [5].

Histologically, It is detected chondrolysis, chondritis, and perichondritis at patients with RP. Cartilage matrix finally converts to fibrous connective tissue [6]. In our case the most prominent histological changes were at the ear cartilage, and it was minimal at nasal cartilage. It was determined diffuse eosinophilia at matrix, intermittent lacunar loss at chondrocytes, near total effacement and increasing of irregular collagen at these sites in ear cartilage(Figure 2).

Auricular (% 39) and nasal septal cartilage(% 24) are the first and also the most common affected sites at RP [7]. The second most common seen affection is as nonerosive, seronegative, asymmetric, oligoarthritis or poliarthritis and it affects 70% of patients [8]. Pulmonary findings consist of tracheal and bronchial stenosis, thickening and calcification of airways, obstructive bronchiectasis and dynamic tracheal/ bronchial collapse [9]. It may be also audiovestibular disorders on patients. Sudden hearing loss is generally permanent and it was also reported as the first sign [10]. Ophthalmologic inflammation is episodically seen at uveal tractus, conjunctiva, sclera and/or cornea. Episcleritis and scleritis are the most seen ophthalmologic inflammation forms [11]. Cardiovascular, central nervous system, skin and renal involvement may be detected in the course of the disease.

Erythema and swelling of both auricles and external ear canals, and decline in hearing bilaterally were detected in clinic of the patient at approval to our unit. Other potentially disease related signs such as bilateral conjunctivitis, episcleritis, cataract, bronchitis were unraveled with the consultations of the related specialty branches.

The main component at the treatment of RP is systemic corticosteroids. When methotrexate is given with steroids, prominently decreases the need for steroid of the patient and also takes the symptoms under control[2]. In our case methotrexate therapy was applied with dose of 7.5 mg/week which was started and elevated to 22.5mg/week as a maintenance dose for four months. Additionally, corticosteroid was started 64 mg/day and the dose tapered to 16 mg/day as a maintenance dose. This treatment plan was effective in which the patient's complaints have been relieved in one month.

In conclusion; RP commonly affects otolaryngologic structures clinically. Therefore, ENT physicians must have knowledge about this disease. Interdisciplinary close relationships are unavoidable in the treatment and follow up of the disease.

References

1. Cummings C.W. Cummings otolaryngology-head&neck surgery. In: Cummings CW, Fredrick JM, Harker LA, editors. Chapter 127. Philadelphia: Elsevier-Mosby Press; 2005. p. 2881-2905.

2. 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.

3. Trentham DE, Le CH. Relapsing polychondritis. Ann Intern Med 1998; 129:114-22.

4. Lang B, Rothenfusser A, Lanchbury JS, Rauh G, Breedveld FC, Urlacher A et al. Susceptibility to relapsing polychondritis is associated with HLA-DR4. Arthritis Rheum 1993; 36:660-4.

5. Kent PD, Michet CJ Jr, Luthra HS. Relapsing Polychondritis. Curr Opin Rheumatol 2004; 16:56-61.

6. Thompson LD. Relapsing polychondritis. Ear Nose Throat J 2002; 81:705.

7. Letko E, Zafirakis P, Baltatzis S, Voudouri A, Livir-Rallatos C, Foster CS Relapsing polychondritis: a clinical review. Semin Arthritis Rheum 2002; 31:384-95.

8. Jawad AS, Burrel M, Kim KL, Scott DG. Erosive arthritis in relapsing polychondritis. Postgrad Med J 1990; 66:768-70.

9. Tillie-Leblond I, Wallaert B, Leblond D, Salez F, Perez T, Remy-Jardin M, et al. Respiratory involvement in relapsing polychondritis. Clinical, functional, endoscopic, and radiographic evaluations. Medicine (Baltimore) 1998; 77:168-76.

10. Kumakiri K, Sakamoto T, Karahashi T, Mineta H, Takebayashi S. A case of relapsing polychondritis preceded by inner ear involvement. Auris Nasus Larynx. 2005; 32:71-6.

11. Isaak BL, Liesegang TJ, Michet CJ Jr. Ocular and systemic findings in relapsing polychondritis. Ophthalmology 1986; 93:681-9.