

Relapsing Polychondritis : A case report and review of the literature.

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ABSTRACT :

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Relapsing polychondritis is an uncommon, chronic disorder of the cartilage that is characterized by progressive recurrent episodes of inflammation of the cartilage on ear pinnae, nose, eye, joints, respiratory and cardiovascular system. The cause of relapsing polychondritis is unknown. It is suspected that this condition is caused by "autoimmunity" to various antibodies such as collagen type II, IX, XI, Matrilin-1, and cartilage oligomeric matrix protein. The diagnosis of RP was made by clinical diagnostic criteria.

We reported a rare case with clinical features of Relapsing Polychondritis. The patient presented with auricular and nasal chondritis with no other organ involvements. The symptoms responded to steroid therapy.

Key words: Relapsing Polychondritis

บทคัดย่อ :

สุธนา โพธิ์มนี, ชินมนัส ตั้งจตุรนต์รัศมี รายงานผู้ป่วยโรค RELAPSING POLYCHONDritis วารสารโรคผิวหนัง 2555; 28: 182-186.

สถาบันโรคผิวหนัง กรมการแพทย์ กระทรวงสาธารณสุข

Relapsing polychondritis เป็นโรคที่มีความผิดปกติของกระดูกอ่อนที่พบได้ไม่บ่อย และยังไม่ทราบสาเหตุการเกิดที่แน่ชัด สันนิษฐานการเกิดโรคเชื่อว่าน่าจะมีสาเหตุการเกิดมาจากภาวะแพ้ภูมิตนเองต่อ collagen type 2, 9, 11, matrilin1 และ oligomatrix protein ส่งผลให้โรคนี้มีอาการแสดงทางคลินิกต่อโครงสร้างที่เป็นกระดูกอ่อนเป็นสำคัญ ยกตัวอย่างเช่น จมูก หู ตา ข้อ ระบบทางเดินหายใจและระบบหัวใจ

ในรายงานนี้ได้นำเสนอผู้ป่วยที่มีลักษณะอาการเข้าได้กับโรค Relapsing polychondritis ตามเกณฑ์วินิจฉัยและผู้ป่วยมีการตอบสนองต่อการรักษาด้วยยา steroid

คำสำคัญ: Relapsing Polychondritis

Case report

A 65-year-old Thai male came to the Institute of Dermatology with painful swelling of both ear pinnas and nose deformity for 2 years. The lesions were slowly progressive over time and were not associated with antecedent medications or previous trauma. He denied medical conditions, but complaint about concomitant malaise, arthralgia and hoarseness for 1 year. He also denied other associated symptoms such as cough, dyspnea, vertigo, and hearing loss. None of his family members had the same problems as the patient.

Examination revealed painful, swollen, flesh-colored, soft and floppy earlobes, which looked like “cauliflower” appearance. Saddle nose deformity was observed. The other systemic symptoms were normal (Figure 1).

Histopathology of a lesional skin biopsy showed nodular infiltration of lymphocytes and plasma cells associated with marked fibrosis at the deep dermis; however cartilaginous pattern cannot be identified in the specimen (Figure 2). Systemic work up for hepatitis B and C, and serum antinuclear antibody were negative. Blood chemistry, urine analysis, chest radiograph, electrocardiogram, eye and larynx examination performed to rule out systemic involvement revealed normal findings.

Our patient was treated with oral prednisolone 15 mg twice daily. He has shown some clinical improvement since one month of treatment, while prednisone was slowly tapered to 10 mg once daily. He is still in the follow-up period.

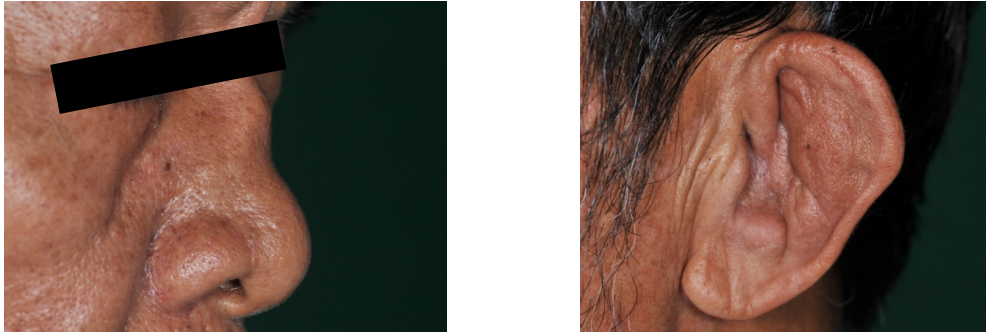


Figure 1 Physical examination revealed (A) Saddle nose deformity.(B) Soft and floppy ear with a cauliflower appearance.

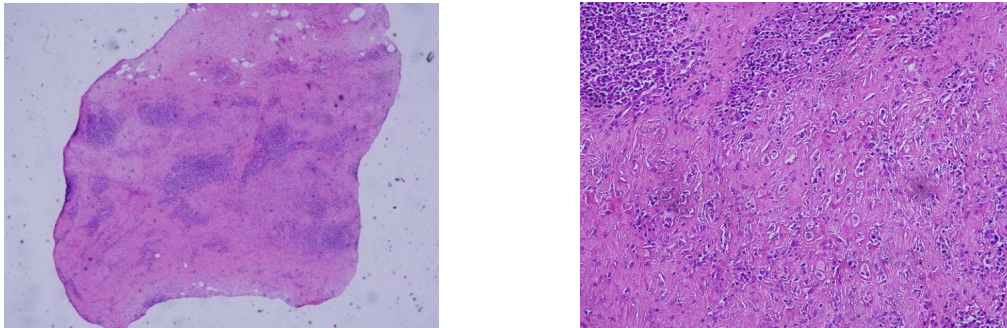


Figure 2 Histopathological examination (A) Magnification 4X (B) Magnification 40X showing nodular and diffuse inflammatory cell infiltration with lymphocytes and plasma cells in thickened fibrous septum.

Discussion

Relapsing polychondritis (RP) is a very rare autoimmune disease characterized by inflammation and degeneration of cartilaginous structures. The disease was first described in 1923 by Jaksch-Wartenhorst under the term of “polychondropathia”¹.

It was reported to occur with equal frequency in both sexes. The average age of the disease onset is around 50, but some reports described the age range of 3 to 86 year-old^{2,3}.

The cause of RP is unknown. There is a positive association with HLA-DR4⁵. It is suspected that this condition is caused by autoimmunity⁵. The role of the

humoral immune response was based on the presence of antibodies to collagen type II⁴, type IX and type XI, matrilin-1⁶, and antibodies to cartilage oligomeric matrix protein.

This condition can affect all types of cartilage such as the elastin cartilage of the ears and nose, the hyaline cartilage of the peripheral joints, the fibrocartilage of the axial skeleton and the cartilage of the tracheobronchial tree. The disease can also affect other proteoglycan-rich structures like the eye, heart, blood vessels and inner ear⁷. The associated diseases include vasculitis, connective tissue diseases and hematologic diseases such as myelodysplastic syndrome^{8,9}.

The patient can present with a wide spectrum of the clinical symptoms and signs. External ear involvement is the presenting sign in almost all patient (85-95%)². Auricular chondritis shows sudden pinna earpain, redness, and swelling sparing the lobule. After several intermittent inflammation, the ear cartilage turn to deformity soft, floppy, and cauliflower-like ear. The second most common clinical finding of RP is seronegative, intermittent, migratory asymmetrical arthritis (52-85%)². The joint involvement often affects metacarpophalangeal, proximal interphalangeal, wrists and knee joints. The third most common clinical finding is nasal chondritis (33-72%)². The nasal cartilage can present with nasal pain, swelling, redness, stuffiness, rhinorrhea and epistaxis. Repeated attacks may cause destruction of nasal bridge creating the characteristic "saddle-nose" deformity².

Other common organ involvements include ocular manifestation (19-51%)¹¹ and respiratory tract chondritis (25-50%) which is the most common cause of death in patients with RP (10-50%)¹⁰.

Dermatological signs are seen in 17-36% of patients with RP¹². The manifestations are non-specific. These signs are frequent especially in association with myelodysplasia. Some reported that oral aphthosis was the most common skin finding. The other manifestations include nodules, purpura, papules, sterile pustules, superficial phlebitis, livedo reticularis, ulcerations and distal necrosis¹².

Cardiovascular (24-52%)¹⁴, renal (5-20%)¹³ and neurological involvement probably resulting from vasculitis can occur¹⁵. Although the renal association is rare, this can indicate the poor prognosis. The cardiovascular complications were found as the second most common cause of death of this disease¹³.

The diagnosis of relapsing polychondritis is based on symptoms. There are no specific laboratory (blood, urine) tests or antibodies to render the diagnosis. The empirical diagnostic criteria for RP have been established by McAdam et al² that included the following 6 features: 1.) Bilateral auricular chondritis 2.) Nasal chondritis 3.) Respiratory tract chondritis 4.) Ocular inflammation 5.) Nonerosive seronegative inflammatory polyarthritis 6.) Audiovestibular damage. At least three criteria, or one criteria with positive histology of the cartilage involvement, or two criteria with response to corticosteroids or dapsone therapy has yielded the disease diagnosis.

Histopathology of affected cartilage shows perichondrial inflammation and loss of the normal cartilaginous basophilia; however biopsy of the affected tissue is not always required.

Prognosis of the disease depends on systemic involvement. Inflammation of the cartilage of the trachea, heart, aorta, and other blood vessels can be fatal. However, the disease is mostly limited and mild. Organ involvement should be ruled out by conventional radiographs and computed tomography of the

respiratory tract, pulmonary function tests, and electrocardiogram.

Prednisone is the drug of choice for RP. Prednisolone (0.3 to 1.0 mg/kg) is used in acute flares and for long-term suppression of inflammation. Continuous treatment with prednisone decreases the disease severity, duration, and frequency of relapses. Pulse intravenous steroids are warranted in severe case of acute airway obstruction, and/or before surgical intervention¹⁶. Nonsteroidal anti-inflammatory drugs, colchicine, or dapsone may be useful for mild cartilaginous inflammation without internal organ involvement¹⁷. There have been some case report describing the efficacy of biologic therapies such as anti-CD4 monoclonal antibodies¹⁸, antitumor necrosis factor agents, and that of autologous stem cell transplantation¹⁹.

We diagnose this patient with two chondritis (bilateral auricular and nasal chondritis) and the response to prednisolone although the skin biopsy showed chronic inflammation and fibrosis, but cannot identify the cartilage. The other symptoms were joint pain, hoarseness which later on revealed as normal findings. The patient did not have systemic involvement as the work up for hearing loss, ocular, cardiovascular, respiratory tract complications all showed negative results. We now continue the treatment with prednisone 10 mg/day and try to maintain the low dose of prednisolone as much as possible.

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