CASE 10

A 53-year-old Thai woman from Nakornpathom

Chief complaint:

Bilateral painful swollen ears

Present illness:

She had had swelling, redness, and mild to moderate pain of both auricles for 5-6 months. She denied having hoarseness and joint pain.

Past history:

No underlying diseases

Family history:

Nil

Physical examination:

Right ear: mildly swollen erythema of the small area of the upper part of auricle.

Left ear: swollen, tender, mildly erythema, and loss of cartilaginous frame from top part down to middle part of auricle.

Other systems were normal.

Investigation:

CBC : Hb 12.8 g/dL, Hct. 38.8%, WBC 7.95 k/uL, N 64%, L 22%, Mo 11%, Eo 2%, B 1%, Platelets 366 k/uL

Urinalysis: normal

ESR 42 m/hr (1-20), CRP 14.10 mg/L(-3)

ANA: negative, ANCA : negative

Anti-HBc: positive, Anti-HCV: negative, Anti-HIV: negative

Histopathology: (S06-01174)

Degeneration of marginal chondrocyte with vacuolized and pyknotic nuclei

Dense inflammatory cell infiltrate in the perichondrium

Fragmentation of cartilaginous necrosis

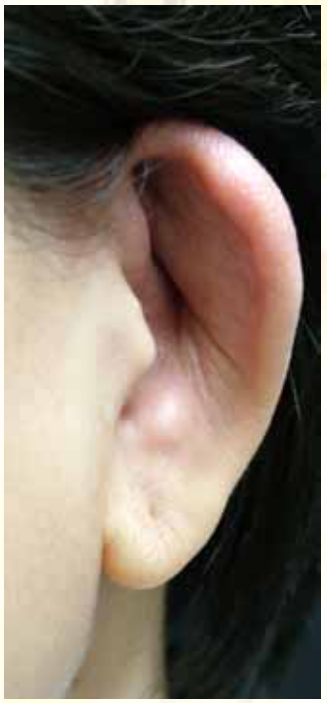

Fig. 10.1

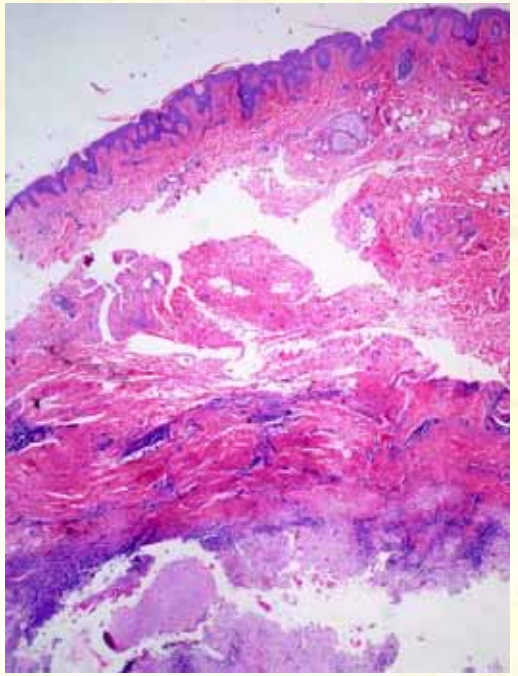

Fig. 10.2

Diagnosis: Relapsing polychondritis
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Discussion:

Relapsing polychondritis (RP) is an uncommon inflammatory disorder with a suspected autoimmune origin. The etiology is unknown, but there are several lines of evidence accumulated to support the role of the immune system:

1. Frequent association with other autoimmune diseases.
2. $CD4^+$ T lymphocytes, plasma cells, immunoglobulin and complement are detectable in tissues lesions.
3. Presence of autoantibody to type II, IX and XI collagens, and other cartilage proteins, including matrilin-I and cartilage oligometric matrix protein (COMP).

4. Detectable cellular immune responses to cartilage constituents.
5. Association with HLA class II DR4.
6. Animals genetically modified and/or immunized with antigens develop symptoms resembling human polychondritis.

RP is characterized by recurrent episodes of inflammation and progressive destruction of cartilaginous structures throughout the body, as well as proteoglycan-rich structures, such as eye, ear, nose, joints, and respiratory system. Auricular chondritis is the most frequent and unique presenting sign, causing pain, redness, and swelling of the ear, sparing the lobule. Inflammatory course generally last a few days or weeks and may subside spontaneously or upon treatment, later, recurrent or persistent inflammation results in cartilage destruction. Most patients have significant disabilities during later stages, including impairments in hearing and vision, as well as cardiorespiratory problems.

Criteria for diagnosis of relapsing polychondritis include three or more of the following features: bilateral auricular chondritis, nonerosive seronegative inflammatory polyarthritis, nasal chondritis, ocular inflammation, respiratory tract chondritis, cochlear and/or vestibular dysfunction or cartilage biopsy confirmation of a compatible histologic picture.

Infectious chondritis, traumatic chondritis, Wegener's granulomatosis, and congenital syphilis may mimic the cartilage destruction seen in RP. Of all these conditions, Wegener's granulomatosis may have the greatest overlap with RP, particularly with regard to dermatologic manifestations and respiratory tract involvement; the former, however, does not have auricular involvement. The two conditions may be distinguished by the presence of vasculitis of small- and medium-sized vessels and anti-proteinase 3 (C-ANCA) serology in Wegener's granulomatosis. Infectious chondritis may involve both the nasal septum and the auricular cartilage, but should lack the other signs and symptoms of RP. Serum antibodies to type II collagen are of limited practical value because they can occur nonspecifically in other rheumatologic disorders.

The primary treatment is prednisolone (30-60 mg./day initially), which improves acute flares and decreases the number and severity of recurrences; NSAIDs may be employed to decrease fever, auricular chondritis, and arthralgia. Sulfones, in particular dapsone (50-150 mg./day as tolerated), can also be used. Other immunosuppressive agents have been tried alone or in conjunction with corticosteroids with variable responses. Anti-TNF agents (infliximab and etanercept) have also been tried successfully inducing remission. Surgery is sometimes performed to repair damaged cartilage.

References:

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