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What is your diagnosis?

Tanınız nedir?

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A 55-year-old male patient presented to the dermatology outpatient clinic with the complaints of redness and swelling of the right ear for the past two weeks. He stated that his ear was painful but did not itch or suppurate. There was no history of trauma. In the medical history, he had cough for one month and was treated with multiple oral and intramuscular antibiotics for the presumptive diagnosis of lung infection with no benefit. His left eye became red two days after cough onset. His eye did not get any benefit from topical cyclopentolate and various antibiotic eye drops either. He also had hemorrhagic nasal drainage and was diagnosed with sinusitis two weeks ago. He was again treated by various antibiotics with no clinical benefit for this complaint. Additionally, he had arthralgia affecting both knees. His medical history revealed that he had hypertension and diabetes and was using metformin and gliclazide. He never smoked. His family history was unremarkable. Dermatological examination showed erythema, swelling and tenderness of the right ear (Figure 1). Needle aspiration revealed no drainage or suppuration. There was also conjunctival hyperemia in his left eye (Figure 2). Laboratory examinations demonstrated a white blood cell count of 13200 /µL (with an increased neutrophil ratio of 71.8% and decreased lymphocyte ratio of 19.5%), a sedimentation rate of 82 mm/hr, and a C-reactive protein value of 72.0 mg/L. Test results for ANA, ANCA and HLA-B27 were all negative. His chest X-ray revealed an increased bronchial shadow and this was nonspecific (Figure 3). Histopathological examination of a punch biopsy specimen obtained from the right ear revealed a predominantly lymphocytic inflammatory cellular infiltrate in the subcutaneous tissue and cartilage.

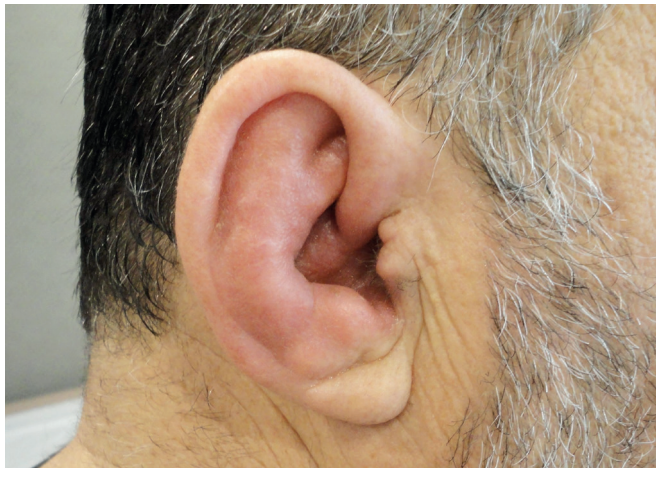

Figure 1. Clinical appearance of the red and swollen right ear of the patient

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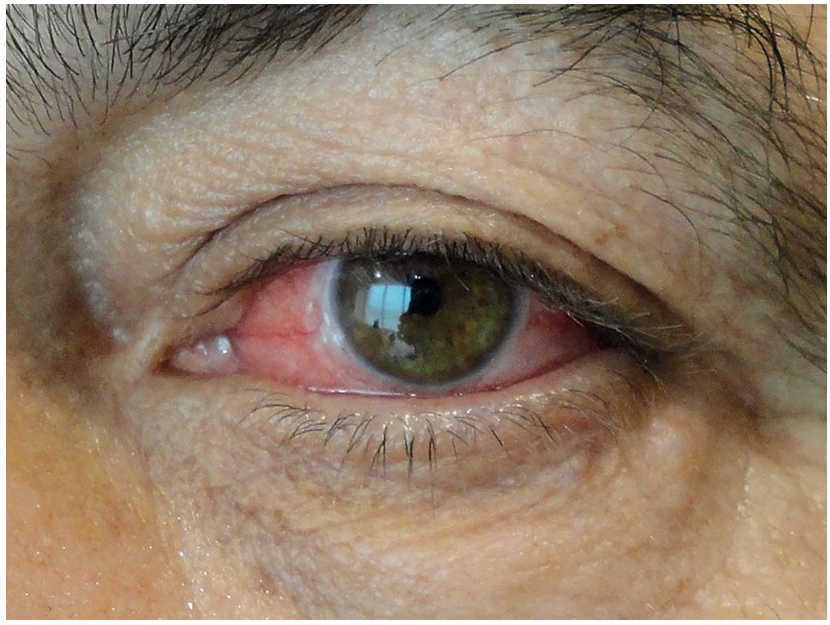

Figure 2. Conjunctival hyperemia in the left eye of the patient

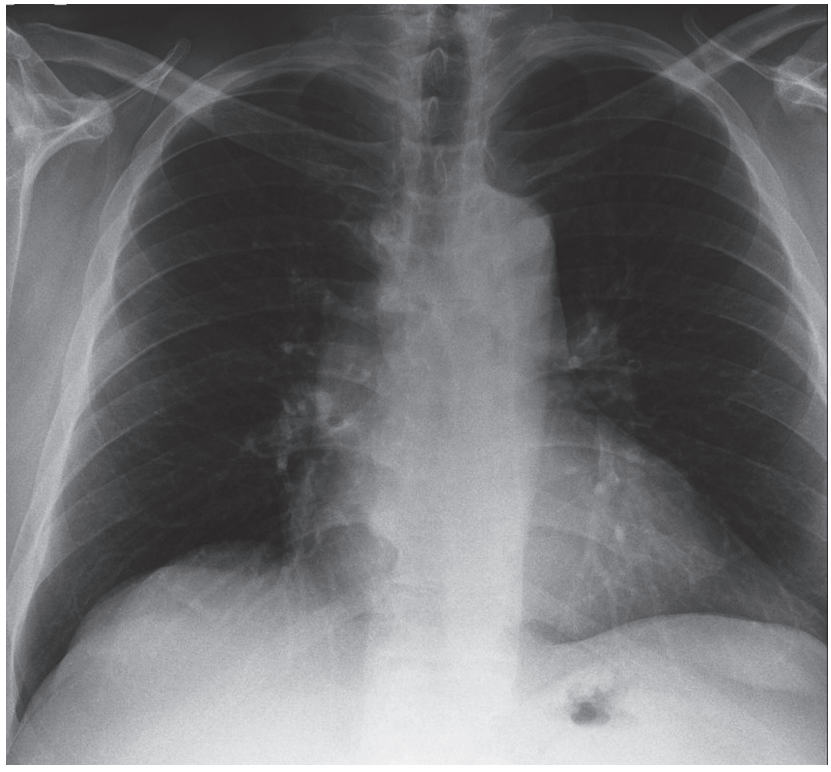

Figure 3. Chest X-ray of the patient

Diagnosis of the case

Diagnosis: Relapsing polychondritis

In the differential diagnosis of a clinical picture characterized by red and swollen ear, red eye and accompanying respiratory findings, we must consider relapsing polychondritis among various other infectious and inflammatory diseases[1,2]. Relapsing polychondritis is a rare inflammatory and degenerative systemic autoimmune disorder in which cartilaginous tissue is converted to fibrous connective tissue[3-5]. Most commonly affected cartilaginous structures are elastic cartilage of the ears and nose, hyaline cartilage of the peripheral joints and tracheobronchial tree, and fibrocartilage of axial skeleton[3]. The eyes are also frequently affected[6]. Structural similarity between microbial heat shock proteins and cartilage is supposed to be important in the etiopathogenesis of this disease[3]. Relapsing polychondritis is supposed to develop as a result of a Th1 cellular immune reaction and abnormal auto-antibody formation against type 2 collagen and/or matrilline-13. The disease is characterized by sudden appearance and a relapsing course[3]. It has been reported that 66.7-73% of patients were misdiagnosed at the beginning and the diagnosis of this disease was delayed consequently[3,6]. The four most common clinical manifestations reported in the literature were ear, nose, peripheral joints and eye involvements[3,4]. There were accompanying disorders, such as systemic vasculitis, connective tissue disorder, autoimmune diseases and hematologic malignancy, in up to 30% of patients with this disease[3]. The diagnosis of relapsing polychondritis is made in the presence of at least three compatible clinical findings and there are no diagnostic laboratory tests available[3,4,6]. Non-steroid anti-inflammatory drugs, colchicium, corticosteroids, various immunosuppressive drugs, such as methotrexate, dapsone and mycophenolate mofetile or biologicals, are used in the treatment[3-5]. Death often occurs due to infectious and cardiovascular problems[3]. Mortality rate is between 30-40% and depends on disease severity and age at disease onset[3].

Ethics

Informed Consent: Consent form was filled out by participant.
Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: B.A., O.E., Concept: B.A., Design: B.A., Data Collection or Processing: B.A., O.E., Analysis or Interpretation: B.A., O.E., Literature Search: B.A., Writing: B.A.
Conflict of Interest: No conflict of interest was declared by the authors.
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