Relapsing polychondritis (RP) is an uncommon disease with an unknown cause. However, antibodies against type II collagen fibers may play a role in the pathogenesis of the disease, which is characterized by recurrent inflammation of cartilaginous tissues, including in the ears, nose, peripheral joints, and laryngotracheo-bronchial tree. In a report by McAdam et al., nearly 50% of patients had laryngotracheo-bronchial involvement; however, only 12% of them had initial airway involvement.

RP is a systemic disease and can induce cardiovascular disorders, such as aortitis, aneurysm, valvular heart disease, and pericarditis. Patients with RP can also have other hematologic, rheumatic, or autoimmune diseases, including thyroid disorders. Here, we present a case of RP that initially manifested with upper airway involvement. Typical auricular and nasal chondritis, and scleritis, developed later. Ascending aortic aneurysm and hyperthyroidism with thyroid goiters were also found. We hope this case will facilitate increased awareness about early airway involvement in RP.

Case Report

A 58-year-old female patient had been well until April 2002, when she began to have an intermittent nonproductive cough and shortness of breath. At that time, there were no fevers, chills, night sweating, orthopnea, or lower-leg edema. Hoarseness and choking ensued 6 months later. Dyspnea became worse, and stridor developed intermittently thereafter. The patient was treated for bronchial asthma, without upper airway involvement. Typical auricular and nasal chondritis, and scleritis, developed later. Ascending aortic aneurysm and hyperthyroidism with thyroid goiters were also found. We hope this case will facilitate increased awareness about early airway involvement in RP.
any significant improvement. She visited Chung-Hsin Municipal Hospital in April 2003, where bronchoscopy was performed. Subglottic stenosis was found, and the patient experienced further dyspnea, so emergency tracheostomy was performed that night.

Three months after tracheostomy, swelling, tenderness and erythematous changes in both ears were noted. Inflammation of the pinna persisted, despite treatment with prednisolone 20 mg/day. Under the impression of RP, the patient was admitted to Taipei Veterans General Hospital for further management. On admission, physical examination disclosed “cauliflower” ears (Figure 1), saddle-nose, periorbital lid edema, increased ocular discharge, and scleritis. Fine rales over both lower lung fields were heard. There was no heart murmur, arthritis or hearing loss. Laboratory data revealed hemoglobin of 12.1 g/dL, white blood cell count of 9,600/mL, and platelet count of 4.5 × 10^5/mL. The serum level of C-reactive protein was 1.34 mg/dL (normal, < 0.5 mg/dL), and erythrocyte sedimentation rate was 134 mm/hour. Renal and liver function tests, and immunoglobulin (Ig) levels, including IgM, IgG and IgA, were all within normal limits. Antinuclear autoantibody, rheumatoid factor, and Venereal Disease Research Laboratory test of serum were negative. Computed tomography (CT) of the chest showed marked swelling of cartilages of the trachea and bilateral main bronchus (Figure 2), severe stenosis above the tracheostomy (Figure 3), ascending aortic aneurysm, and bilateral enlargement of the thyroid gland. Bronchoscopy found that the lumen of the trachea above the tracheostomy was nearly collapsed. The patient’s thyroid function profile disclosed hyperthyroidism, with a thyroid stimulating hormone level of 0.03 µIU/mL (normal, 0.25–4 µIU/mL), and free thyroxine level of 2.23 ng/dL (normal, 0.56–1.47 ng/dL).

The patient was treated with intravenous methylprednisolone 30 mg every 6 hours, and azathioprine 100 mg/day. The swelling of the ears and scleritis of both eyes then improved slowly. The dosage of prednisolone was tapered according to clinical response. The patient has been taking prednisolone 5 mg/day for more than 9 months without any flare-up. However, complete laryngeal collapse has persisted, and the deformed pinnae have shown no improvement.

Discussion

RP is not a common disease, and the cause remains unknown.1–6 The diagnostic criteria for RP proposed...
Laryngotracheal involvement in relapsing polychondritis

by McAdam et al in 1976 comprise the following: 1) recurrent chondritis of both auricles; 2) non-crossee inflammatory polyarthritis; 3) chondritis of nasal cartilages; 4) inflammation of ocular structures, including conjunctivitis, keratitis, scleritis/episcleritis, and/or uveitis; 5) chondritis of the respiratory tract involving laryngeal and/or tracheal cartilages; 6) cochlear and/or vestibular damage manifesting as neurosensory hearing loss, tinnitus, and/or vertigo. When 3 or more of these features are present, even without biopsy confirmation, the diagnosis of RP is definite. The initial presentations in our patient included cough, shortness of breath, and nonspecific respiratory symptoms, so RP was not considered because the patient did not fulfill the abovementioned criteria. Three months after tracheostomy, auricular and nasal chondritis, and scleritis ensued, so a definite diagnosis of RP was then established.

The pathogenesis of RP remains to be elucidated, although autoantibodies against type II collagen fibers may play a role. Indeed, patients with RP have demonstrable serum levels of such autoantibodies, and the autoantibody titer correlates with disease activity. Histopathologic findings in affected cartilage specimens have revealed loss of normal cartilage and inflammation of the perichondrium, which is infiltrated by neutrophils, eosinophils, lymphocytes, and plasma cells.

The most commonly involved organs in RP include the ears (85%), nose (62%), eyes (54%), and joints (50–75%). The airways are affected in about 50–70% of RP cases, but airway involvement as the initial presentation is rare: such initial involvement was noted in only 12% of cases in the report of McAdam et al. As in our case, airway manifestations of RP, such as cough, hoarseness, stridor, wheezing, and choking, can be misdiagnosed as bronchial asthma or other airway diseases. Without adequate treatment, respiratory tract involvement can be complicated by airway collapse, which is the main cause of death in RP, accounting for almost 30% of mortality.

CT studies of the airway in RP can reveal circumferential thickening or a “worm-eaten” appearance in affected cartilage; such changes suggest deformity or edema in the tracheal mucosa. In the late stages, a narrowed, irregular and distorted airway lumen will develop.

The incidence of cardiovascular involvement in RP is about 25–52%. Aortic aneurysm is a rare complication in RP (about 2% of cases), and occurs mostly in the ascending aorta. Other cardiovascular findings may include valvular heart disease, conduction disturbances, arrhythmias, and pericarditis. About 30% of patients with RP have concurrent vasculitis, or autoimmune, hemolytic, or connective-tissue disease. In the study by McAdam et al, thyroid disease had a higher frequency associated with RP (about 17%).

A standard treatment protocol for RP remains to be established; however, corticosteroids are the agents of choice for active inflammation. Methotrexate, cyclosporine, azathioprine, and cyclophosphamide, have all been used to treat RP, either because of their steroid-sparing effects or for use in patients with life-threatening disease. Recently, infliximab was reported to have been used successfully to treat a refractory case of RP. If collapse of laryngeal or tracheal cartilaginous rings or airway obstruction occur, tracheostomy is indicated. If airway obstruction is not severe, the placement of stents may be feasible.

In summary, when a patient has recurrent cough, dyspnea and stridor that do not respond to traditional treatment for upper airway infection or bronchial asthma, RP should be considered, even in the absence of typical auricular, nasal, ocular or articular manifestations. High doses of corticosteroids, or emergency tracheostomy, should be performed immediately if severe airway obstruction has developed. We hope that this report will raise alertness about potential early upper airway involvement in RP.

References

