Symptoms: Painful Ear Swelling

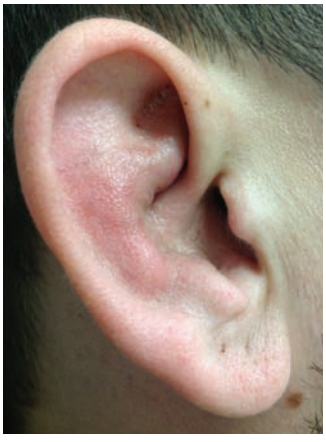
By Hamid R. Djalilian, MD

A patient comes into the office with painful ear swelling, stating that the pain has been increasing gradually from the time it began two weeks earlier. He was started on antibiotics by his primary care doctor, but that did not help, nor did another antibiotic started by his ENT. He previously has been seen in the office for tinnitus.

He states that his ear otherwise had never had swelling, though he has had a few occasions when his nose became very erythematosus

The patient says he did not have an insect bite or other trauma to his ear, and he has not recently traveled outside California. He states that his ear otherwise had never had swelling, though he has had a few occasions when his nose became very erythematosus. He took antibiotics for that, and, while they did not help, the nose problem gradually subsided. The figure to the right shows an image of his ear.

What is your diagnosis? See p. 18.



In this image of our patient's right auricle, note the erythema and edema, with loss of contour of the antihelix. The lobule is spared.

Diagnosis: Relapsing Polychondritis

By Hamid R. Djalilian, MD

Continued from p. 17

The most common cause of a painful red auricle is an infectious etiology—generally an insect bite or the extension of an otitis externa, which often occurs in adults. Otitis externa starts in the external auditory canal skin and, without treatment, can gradually migrate to the pinna and involve the entire auricle. The infection tends to spread anteriorly and inferiorly, affecting the facial and neck skin but rarely involving the scalp.

If not treated, the infection eventually will spread to the auricular cartilage and its overlying perichondrium, resulting in perichondritis. Patients with this condition have redness of the entire skin of the auricle, including the lobule. Those with auricular cellulitis, a skin infection not involving the cartilage, have ear discomfort but not severe pain. The most common bacterial cause of these infections is *Pseudomonas spp.* or *Staphylococcus aureus.* Once the cartilage is involved, intravenous antibiotics are the only option.

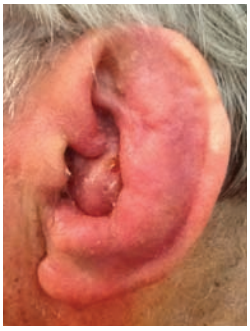
Polymicrobial infections of the auricle are caused by insect bites, usually mosquito or spider bites. These infections tend to occur a few days after the initial bite, by which time the patient has forgotten about it. The redness of the skin spreads slowly to the entire auricle. When the infection is caught early in a patient with a normal immune system, oral antibiotics can be used. In those who have diabetes, are otherwise immunocompromised, or have an advanced infection involving the face, treatment will require intravenous antibiotics.

The patient in this scenario had relapsing polychondritis, an autoimmune condition affecting hyaline cartilage, which is found in the ears, nose, larynx, and trachea, and multiple systems in the body. The inflammatory reaction of the cartilage, or chondritis, leads to many of the manifestations seen clinically. The diagnosis depends on the presence of at least three of the following six criteria:

- Auricular chondritis.
- Non-erosive seronegative polyarthritis.
- Nasal chondritis.
- Ocular inflammation.
- Respiratory tract chondritis.
- Audiovestibular damage.

Other diagnostic criteria include chondritis in two of three sites (auricular, nasal, or laryngotracheal), or chondritis in one of these sites plus two other features, including ocular inflammation, audiovestibular damage, or seronegative inflammatory arthritis.

The manifestations of this condition in the ear are the most common clinical signs. Inflammation of the auricular cartilage can be unilateral or bilateral, and it occurs in 83 percent of patients. The condition usually presents with acute or subacute pain, swelling, and redness of the ear. The inflammation characteristically does not involve the lobule, as the lobule does not contain cartilage.



This patient had a skin cancer removed from his ear, and the site subsequently became infected, leading to perichondritis.

Conductive hearing loss can occur due to edema of the ear canal or Eustachian tube dysfunction and possible serous otitis media. Hearing loss also can be sensorineural, resulting from inflammation of the internal auditory artery. Chronic auricular cartilage inflammation can cause loss of the auricular structure and lead to a floppy or deformed ear. Some patients may manifest a cauliflower ear in the long term.

Conductive hearing loss can occur due to edema of the ear canal or Eustachian tube dysfunction

Musculoskeletal manifestations include joint pain, especially in the chest joints, costochondral joint (between the ribs and sternum), and sternoclavicular joint (between the sternum and the collarbone). Arthritis can occur in other joints and is usually asymmetric, only affecting one side, unlike osteoarthritis, which usually affects both sides in a symmetric fashion.

Chondritis of the nasal cartilage occurs in 61 percent of patients, with symptoms of nasal obstruction, crusting, drainage, and bleeding. Over time, if the nasal chondritis continues, it can destroy the nasal septum and cause the nose to collapse. When the inflammation involves the larynx, trachea, or bronchi, the problem could be life-threatening because of airway narrowing. Patients can have hoarseness, loss of voice, wheezing, cough, or shortness of breath.

These patients generally are treated by a rheumatologist, who will check multiple blood studies to evaluate for the presence of other autoimmune conditions that can occur in conjunction with relapsing polychondritis. The prognosis depends on the aggressiveness of the disease. Patients are given immunosuppressive agents for prevention or treatment of an acute attack.

Dr. Djalilian is the director of neurotology and skull base surgery and an associate professor of otolaryngology and biomedical engineering at the University of California, Irvine.