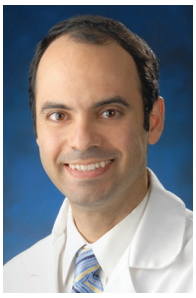


CLINICAL CONSULTATION

Symptoms: Ear Infections and Joint Pain

By Hamid R. Djalilian, MD

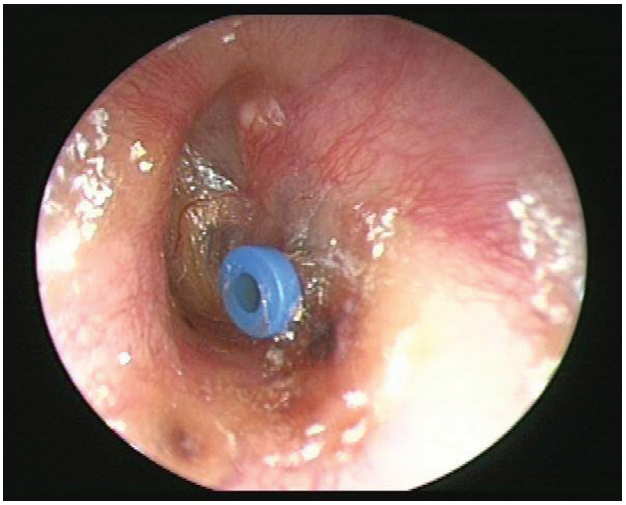
A 45-year-old physician presents with a history of recurrent ear infections that started four months prior to the visit. Since then, he has had significant problems with joint pain, to the point where he has not been able to work or perform surgery.

The patient also has experienced intermittent difficulty standing and walking because of severe knee pain. In addition, he has had two subsequent infections, both of which have resolved. Otherwise, the patient does not have a history of ear infections as a child or of any unusual travel, he said.

Another otolaryngologist had placed a tube in the patient's ear. An examination revealed the tube in place, with serous drainage from the middle ear.

What is your diagnosis? See p. 18.

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



An otoscopy of the patient's tympanic membrane showed serous drainage from the ear tube.

Diagnosis: Granulomatosis with Polyangiitis

By Hamid R. Djalilian, MD

Continued from p. 16

The presentation of acute otitis media in an adult who does not have previous ear disease or Eustachian tube dysfunction should alert the clinician that the patient has something more than just acute otitis media.

Acute otitis media generally is a disease of childhood. It is caused by Eustachian tube dysfunction, which occurs primarily during this stage because of children's anatomy and inflammatory conditions.

A child's Eustachian tube is shorter and has a more obtuse angle with the nasopharynx. In addition, children experience frequent upper respiratory infections, which further increase their likelihood of acute otitis media.

Among adults, on the other hand, infections are less common, and the Eustachian tubes are larger and longer. The middle ear is positioned more superiorly, which leads to a much lower likelihood of acute otitis media development in the adult population.

Also, the adenoid is atrophic and much smaller in adults compared with children. In children, the adenoid harbors the bacteria that cause otitis media. That is why children with recurrent otitis will have an adenoidectomy as part of a pressure-equalization (PE) tube placement the second time the tube procedure is performed.

Our patient had seen a rheumatologist for the joint pains and was told that he had rheumatoid arthritis. Immunosuppression was to be started shortly after that diagnosis.

The patient primarily had presented to us to get his ears checked and make sure that the infection had settled before starting immunosuppressive therapy. The diagnosis of rheumatoid arthritis had been made in the absence of supportive laboratory testing for the condition.

CONSIDER SYSTEMIC CONDITIONS

In this patient and other adults who have recurrent acute otitis media in the absence of Eustachian tube dysfunction or otitis history, an investigation for a systemic condition as the cause of the otitis must ensue.

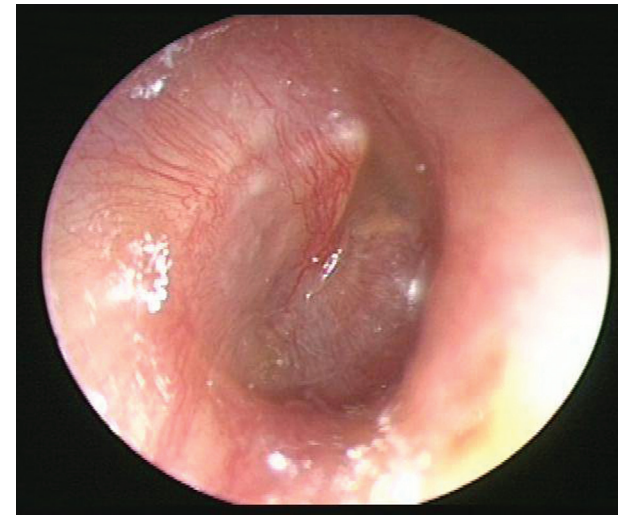
Laboratory studies for tuberculosis; granulomatosis with polyangiitis, formerly known as Wegener's granulomatosis; sarcoidosis; HIV; and systemic lupus erythematosus should be conducted. If travel history warrants it, a test of Lyme disease should be performed.

Lyme disease is most common in patients exposed to the outdoors of the Midwestern and Northeastern United States. The condition generally does not cause middle ear dysfunction; however, given the arthritis, the patient was evaluated for Lyme disease.

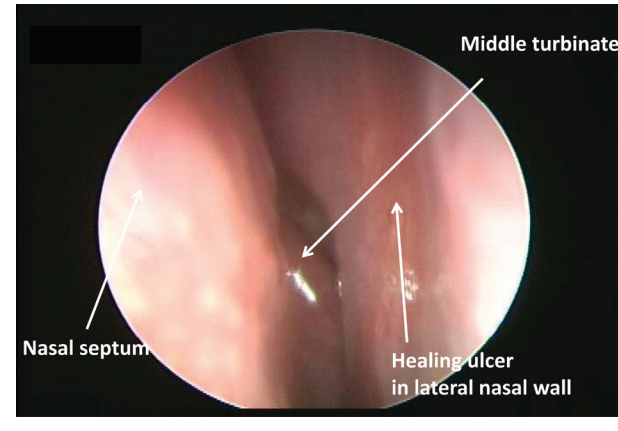
Additional testing such as a chest X-ray and urinalysis may be helpful to look for other signs of sarcoidosis and granulomatosis with polyangiitis, respectively.

In addition to an examination of the ear, the adult patient with recurrent acute otitis should have a nasal endoscopy and nasopharyngoscopy performed to look for signs of nasal inflammation, which could suggest sarcoidosis or granulomatosis with polyangiitis. A laryngoscopy also should be conducted to look for evidence of supraglottic or overt subglottic stenosis.

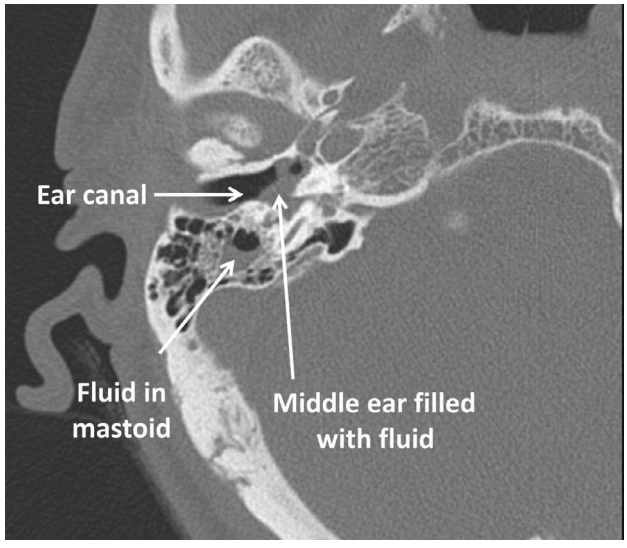
This patient had an abnormal urinalysis and a positive cytoplasmic antineutrophil cytoplasmic antibody (cANCA) test, which indicated the diagnosis of granulomatosis with



The patient's other tympanic membrane shows serous otitis.



This endoscopic image of the left nasal cavity reveals a healing ulcer on the lateral nasal wall.



The axial CT of temporal bones demonstrates fluid in the middle ear and mastoid, without bony destruction.

polyangiitis. He was started on high-dose steroid therapy and later placed on other immunosuppressive agents for this condition by the rheumatologist.

COMPLEX OF SYMPTOMS

Granulomatosis with polyangiitis is a systemic autoimmune condition that primarily affects the upper respiratory tract, lungs, and kidneys, but it can involve any other organ in the body as well. It is a form of vasculitis, or inflammation of the blood vessels, caused by antineutrophil cytoplasmic antibodies, which are circulating autoimmune antibodies against small- and medium-size blood vessels.

The initial signs of granulomatosis with polyangiitis vary significantly, and diagnosis can be delayed given the nonspecific nature of some of the presenting symptoms. The most common presenting signs are nasal drainage and sinusitis symptoms, such as crusting, septal perforation, and saddle-nose deformity.

Otologic symptoms of this condition include conductive hearing loss, which results from middle ear inflammation or serous otitis, and sensorineural hearing loss. Patients may present with rapidly progressive mixed hearing loss.

Patients also can present with progressive kidney dysfunction; subglottic stenosis, or stenosis below the vocal cords; inflammation of the eyes; pulmonary nodules or hemorrhage; blood in sputum; joint pain; and skin nodules, among other symptoms.

While a positive cANCA test helps solidify the diagnosis, the determination of granulomatosis with polyangiitis is clinical and based on the complex of symptoms. If a nodule or lesion can be identified, a biopsy is performed to look for necrotizing granulomas, which are a hallmark of the condition. Subglottic stenosis requires surgical dilation.

Treatment of this condition consists of high-dose steroids and cyclophosphamide to control the symptoms. This patient responded to 80 mg of prednisone daily for a week, followed by a tapered dose.