Concomitant inflammatory pseudotumor of the temporal bone and lung: A case report

Joo Hyung Lee, MD; Min Kyo Jung, MD; Chang Eun Song, MD; Sang Won Yeo, MD; Hye Kyung Lee, MD; Po Song Yang, MD; Soo Whan Kim, MD

Abstract

Inflammatory pseudotumors are histologically benign but locally destructive lesions that are usually found in the lung, although some cases of temporal bone involvement have been reported. To the best of our knowledge, no case of simultaneous involvement of the temporal bone and the lung has been previously reported in the literature. We describe such a case in a 39-year-old man. The temporal bone lesion was removed in its entirety, and the lung lesion was treated with steroid therapy. At the 2-month follow-up, the size of the lung mass on chest x-ray was significantly reduced, and at 1 year, the patient was asymptomatic.

Introduction

Inflammatory pseudotumors are histologically benign but locally destructive lesions made up of fibrovascular tissue with chronic inflammatory cells. They are usually found in the lung. Several reports of inflammatory pseudotumor of the temporal bone have been published in the literature, but to the best of our knowledge, the appearance of simultaneous lesions of the temporal bone and lung has not been reported previously. In this article, we describe what we believe is the first such reported case.

Case report

A 39-year-old man was referred to the ENT department with a 1-year history of hearing impairment in the right ear and a 1-month history of retrobulbar pain associated with diplopia. His personal and family histories were unremarkable. An otolaryngologic examination detected nothing specific, but an ophthalmologic examination revealed mild palsy of the right lateral rectus muscle. An audiologic study demonstrated a profound mixed hearing loss on the right with poor speech discrimination; the tympanogram was normal. Evoked auditory brainstem responses were absent on the right side.

High-resolution computed tomography (CT) of the temporal bones detected a diffuse soft-tissue lesion in the right middle ear cavity and mastoid antrum (figure 1, A). No evidence of ossicular destruction was identified. Magnetic resonance imaging (MRI) was performed to define the extent of the mass. T1-weighted imaging revealed that the mass was isointense to the gray matter of the brain and that it enhanced relatively well (figure 1, B). On T2-weighted imaging, the mass demonstrated low signal intensity; this image showed that the mass had extended to the right petrous apex.

The lung tumor was found incidentally on chest x-ray, which showed a round, 3 × 3-cm, radiopaque mass in the left upper lung field (figure 2, A). This finding prompted a chest CT, which confirmed the presence of soft-tissue lesions in the left lung (figure 2, B). Findings on hematologic testing were normal except for a high leukocyte count (11,800/mm³) and a high C-reactive protein level (28.5 mg/L).

We performed a simple mastoidectomy to obtain a pathologic specimen. Frozen-section biopsy showed no evidence of malignancy, so we performed an open-cavity mastoidectomy and tympanoplasty to remove as much of the mass as possible. An otolaryngologic examination detected nothing specific, but an ophthalmologic examination revealed mild palsy of the right lateral rectus muscle. An audiologic study demonstrated a profound mixed hearing loss on the right with poor speech discrimination; the tympanogram was normal. Evoked auditory brainstem responses were absent on the right side.

From the Department of Otolaryngology–Head and Neck Surgery (Dr. J.H. Lee, Dr. Jung, Dr. Song, Dr. Yeo, and Dr. Kim), the Department of Pathology (Dr. H.K. Lee), and the Department of Radiology (Dr. Yang), College of Medicine, The Catholic University of Korea, Seoul.

Reprint requests: Soo Whan Kim, MD, Department of Otolaryngology–Head and Neck Surgery, College of Medicine, The Catholic University of Korea, Kangnam St. Mary’s Hospital, 505 Banpo-dong, Seocho-gu, Seoul 137-701, Korea. Phone: 82-2-590-1512; fax: 82-2-595-1354; e-mail: otology@unitel.co.kr
CT-guided biopsy was performed to evaluate the lung lesion. Histopathologic examinations of both specimens revealed proliferation of small vessels and plasma cell infiltration, as well as moderate fibrosis in both the temporal bone and lung (figure 3). Mature lymphocytes and cholesterol clefts were also noted. No atypical plasma cells or Dutcher bodies were seen, but a few Russell bodies were found. Immunohistochemical staining of kappa and lambda light-chain proteins was performed to evaluate the monoclonality of the plasma cells; polyclonality from a mixture of kappa and lambda light-chain proteins was observed.

Antibiotics were prescribed postoperatively. The retrobulbar pain had eased significantly by postoperative day 1, and the lateral-gaze palsy began to gradually resolve. After histopathologic confirmation of inflammatory pseudotumor, oral prednisolone at 60 mg/day was administered for 10 days and then tapered. On postoperative day 12, the lateral-gaze palsy was absent. At 2 months postoperatively, the size of the lung mass on chest x-ray was significantly reduced. At the 1-year follow-up, the patient was asymptomatic.

Discussion
Inflammatory pseudotumors are clinically similar to malignant tumors. While most occur in the lung, some do arise in the head and neck, primarily in the orbit; other head/neck sites include the larynx, pharynx, oral cavity, paranasal sinus, salivary glands, and parapharyngeal space. Our review of the literature revealed no report of simultaneous inflammatory pseudotumors of the temporal bone and lung.

The etiology and pathophysiology of inflammatory pseudotumor are not well understood. Infection with a corresponding immune response has been suggested but not proven as an etiologic factor. In 1954, Umiker and Iverson suggested that the development of inflammatory pseudotumor is associated with a chronic inflammatory state because most affected patients had a history of infection; they reported that plasma cells had a tendency toward polyclonality, which suggests previous inflammation as the cause. Snyder et al reported a translocation and deletion of the chromosomes in inflammatory pseudotumor of the lung, which favors the possibility of an oncogenic origin.

The diagnosis of inflammatory pseudotumor is based primarily on pathologic examination and immunohistochemical confirmation of plasma cell polyclonality. Other causes must be excluded.

The clinical manifestations of inflammatory pseudotumor are varied, as the lesion arises in various organs as a space-occupying mass. In our patient, the lateral-gaze palsy and hearing impairment were probably caused by nerve compression by the mass.

The characteristic pathologic findings of inflammatory

Figure 1. A: Axial CT of the temporal bones shows the soft-tissue lesion in the right middle ear cavity and mastoid antrum. Some bone destruction is seen in the mastoid antrum, but the ossicles are preserved. B: Contrast-enhanced T1-weighted axial MRI shows that the soft-tissue lesion involves the right mastoid and the apex of the right petrous bone.

Figure 2. A: Chest x-ray shows consolidated masses in the left upper lung zone. B: Contrast-enhanced CT (5-mm collimation) of the chest at the level of the aortic arch shows peripheral lobulated masses with relatively good enhancement.
Inflammatory pseudotumor are infiltration of mature lymphocytes and plasma cells and proliferation of myofibroblasts or fibroblasts. Immunoperoxidase staining of plasma cells is a critical step in differentiating inflammatory pseudotumor from monoclonal neoplastic extramedullary plasmacytoma; the appearance of these two entities can be similar under light microscopy. It is known that the more proliferative the myofibroblasts and fibroblasts are, the more infiltrative and recurrent the inflammatory pseudotumor is. In our patient, histopathologic examination showed plasma cell infiltration accompanied by fibrosis (which are typical findings of inflammatory pseudotumor), but proliferation of myofibroblasts or fibroblasts was not so evident.

The natural history of inflammatory pseudotumors is variable. Some of these lesions resolve spontaneously, some remain stable for years, and some continue to grow, occasionally invading the adjacent tissue.

Treatment is not well established. Treatment options include systemic steroid therapy, radiotherapy, and surgical excision—or any combination of the three. The choice of treatment is guided by the location of the lesion, the degree of infiltration into adjacent tissues, the likelihood of achieving a complete excision, and histologic considerations. Bahadori and Liebow reported that steroid therapy was ineffective as a treatment for inflammatory pseudotumor, but most other authors have reported that steroids—either alone or as an adjunct to surgery—are effective in the clinical and radiologic resolution of inflammatory pseudotumor. Radiotherapy might be applied in recurrent cases, in cases when steroid therapy has proved to be ineffective, or when steroids are contraindicated. Surgical excision can be considered when the lesion can be excised without causing injury to major vessels or nerves and when the lesion recurs after steroid treatment.

Our patient exhibited clinical and radiologic improvement in both the temporal bone and lung lesions. We considered surgery initially for the temporal bone lesion because the patient had lateral-gaze palsy, which had probably been caused by compression of the abducens nerve and therefore required immediate decompression. We did not perform petrosectomy because we concluded that removal of the mass had achieved sufficient decompression.

References