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Fibroinflammatory Pseudotumor of the Ear

A Locally Destructive Benign Lesion

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We describe three cases of a fibroinflammatory pseudotumor (tumefactive fibroinflammatory lesion) of the middle and inner ear. The patients presented with total deafness in the affected ear and no response to caloric stimulation. The computed tomographic pattern showed destruction of inner ear structures and a typical widening of parts of the labyrinth. Magnetic resonance imaging performed in all three patients showed an extension greater than expected based on computed tomographic images of both areas of destruction, as well as areas of radiologic normality. An enhancing mass was seen in the inner ear with a characteristic extension into both the internal auditory canal and the middle ear. A transotic approach or subtotal petrosectomy was used to remove the tumor in all three cases. Although histologically benign, these tumors are locally destructive and, as such, behave like a neoplastic lesion. They are composed of fibrovascular tissue admixed with chronic inflammatory cells. To our knowledge, this is the first report on pseudotumors of the middle ear, inner ear, and internal auditory canal. *Inflammatory pseudotumor* used to be a somewhat confusing term for a recognized entity of unknown origin. It is likely that infection is an important contributing factor in the development of these lesions. Although surgical removal seems to be the treatment of choice, no clear judgment of its prognosis can be made owing to the rarity of this tumor.

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Fibroinflammatory pseudotumors are defined as histologically benign, locally destructive lesions composed of fibrovascular tissue admixed with chronic inflammatory cells.¹ Pseudotumors of the middle and inner ear have not, to our knowledge, been described previously. Moreover, the fact that these lesions may mimic a malignancy clinically justifies the attention drawn to this histopathologic entity.

Three patients were diagnosed as having a fibroinflammatory pseudotumor in the ear in the Department of Otorhinolaryngology–Head and Neck Surgery, Radboud Hospital, Nijmegen, the Netherlands, between June 1991 and October 1993. These patients subsequently underwent surgery for treatment of this condition. Herein, we pre-

sent these three cases and describe the radiologic and histologic findings, as well as the treatment used.

REPORT OF CASES

CASE 1

A 39-year-old man presented with hearing impairment of 3 months' duration and otalgia in his left ear. A diagnosis of otitis media with effusion was made, and a myringotomy was performed. Immediately thereafter, his hearing deteriorated to total deafness and a violent vertigo developed that forced the patient to stay in bed for 2 days. One week later, the vertigo had gradually subsided but the total hearing impairment in his left ear remained. On examination, the caloric response was absent, and the patient was referred to our hospital.

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CASE 2

Except for the deafness there were no significant complaints. Otoloscopic examination revealed fluid behind the left tympanic membrane that simulated otitis media with effusion. Facial nerve function was undisturbed. Audiometry revealed total deafness in the left ear, while the hearing in the right ear was normal. High-resolution computed tomography (CT) in the axial plane with coronal reformatted images revealed that the contour and diameter of the internal auditory canal were normal, although the fundus of the canal seemed to be enlarged. The contours of the geniculate ganglion were normal. The ossicles were present and in their normal anatomic position. The bony contours of the vestibule were indistinct. The superior semicircular canal showed a widened lumen and had sclerotic margins. The whole tympanic cavity was filled with soft tissue, and the pneumatized mastoid cavity was not aerated. Magnetic resonance imaging (MRI) of the ear before and after gadolinium-diethylene-triamine-pentaacetic acid contrast enhancement using a surface coil showed an enhancing mass in the tympanic cavity and inner ear and also a mass in the internal auditory canal.

Based on the total deafness in the left ear, the lack of response using ear, nose, and throat recordings for caloric testing, and the destruction of the labyrinth with an erosion of the internal acoustic canal suggesting the presence of a vascularized tumor, it was decided to perform a modified transotic approach to remove the tumor and to preserve facial nerve function. The middle ear was filled with mucus, and a thickened mucosa covering a distinct tumor was found in the middle ear, mastoid cavity, and semicircular canals. The tumor extended medially to the horizontal and descending part of the facial nerve into the basal coil of the cochlea, and had invaded the internal auditory canal. The tumor was found to be firm but had a clear plane of cleavage in relationship to its surroundings. The internal auditory canal was sealed off with fascia and fibrin glue. The cavity was filled with abdominal fat, and a blind sac closure was made to the external auditory canal. The final histologic diagnosis was consistent with a fibroinflammatory pseudotumor.

A 38-year-old woman, who had previously undergone six surgical procedures in her right ear because of chronic otitis and cholesteatoma and a radical mastoidectomy in 1983 after which otorrhea persisted, was referred to our hospital. Otoloscopic examination showed a large polyp near the promontory. The right ear was totally deaf, and the left ear showed a mainly sensorineural hearing loss with a mean of 20 dB at 0.5, 1, and 2 kHz. The vestibular test showed no response using ear, nose, and throat recordings for caloric testing on the right side. High-resolution CT scans in the axial and coronal planes showed normal contours and a normal diameter of the internal auditory canal. The fundus of the canal was enlarged. The geniculate ganglion was normal, but the windings of the cochlea, the vestibule, as well as the contours of the semicircular canals could not be detected. The vestibule was totally replaced by a soft-tissue mass. The mastoid cavity was detected, but no parts of the ossicular chain were visible. Magnetic resonance imaging showed an enhancing mass in the mastoid cavity and inner ear extending into the internal acoustic canal and the cochlea.

A modified transotic approach, including removal of the promontory and cochlea, exposing the internal auditory canal, was performed. A reddish tumor was removed in toto. The lesion extended into the internal auditory canal and had also eroded the sheath of the carotid artery and the facial nerve. The tumor was dissected from the facial nerve, which was preserved. The tumor was further removed, leaving a large cavity in the petrous bone, which was filled with abdominal fat. A blind sac closure of the external auditory canal was made. The histologic diagnosis was consistent with a fibroinflammatory pseudotumor.

CASE 3

A 50-year-old woman presented with a chronic otitis with otorrhea, otalgia, and hearing impairment in her right ear. She underwent an exploratory tympanotomy. Postoperatively, severe dizziness was present.

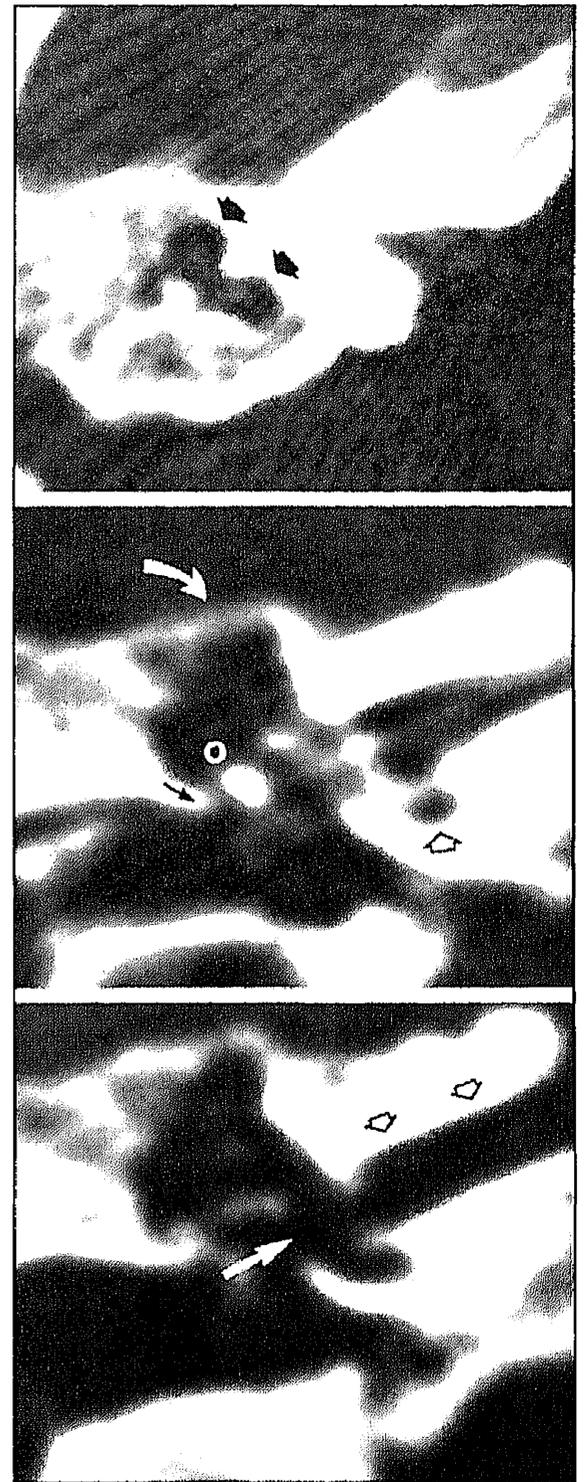


Figure 1. Case 3. High-resolution computed tomographic scan of the right temporal bone. Top, A transverse plane, widening of the superior semicircular canal (arrows). Center, Direct coronal plane, soft-tissue mass in the external auditory canal, tympanic cavity, and inner ear with destruction of the vestibule. Normal scutum (thin arrow), ossicles (O), tegmen (curved white arrow), and basal turn of the cochlea (open black arrow). Bottom, Coronal image, more posterior than that seen in the center illustration with normal internal canal (open arrows), but with widening and destruction of the vestibule (white arrow).

A long-lasting history of sinusitis was also present. An otoscopic examination revealed abundant otorrhea on the right side obstructing the view of the tympanic membrane. No other ear, nose, or throat abnormalities were found. Audiometry showed a deaf ear on the right side and a mean sensorineural loss of 35 dB at 0.5, 1, and 2 kHz on the left side. The right vestibulum did not show any function on electronystagmography. High-resolution CT scans in the axial and

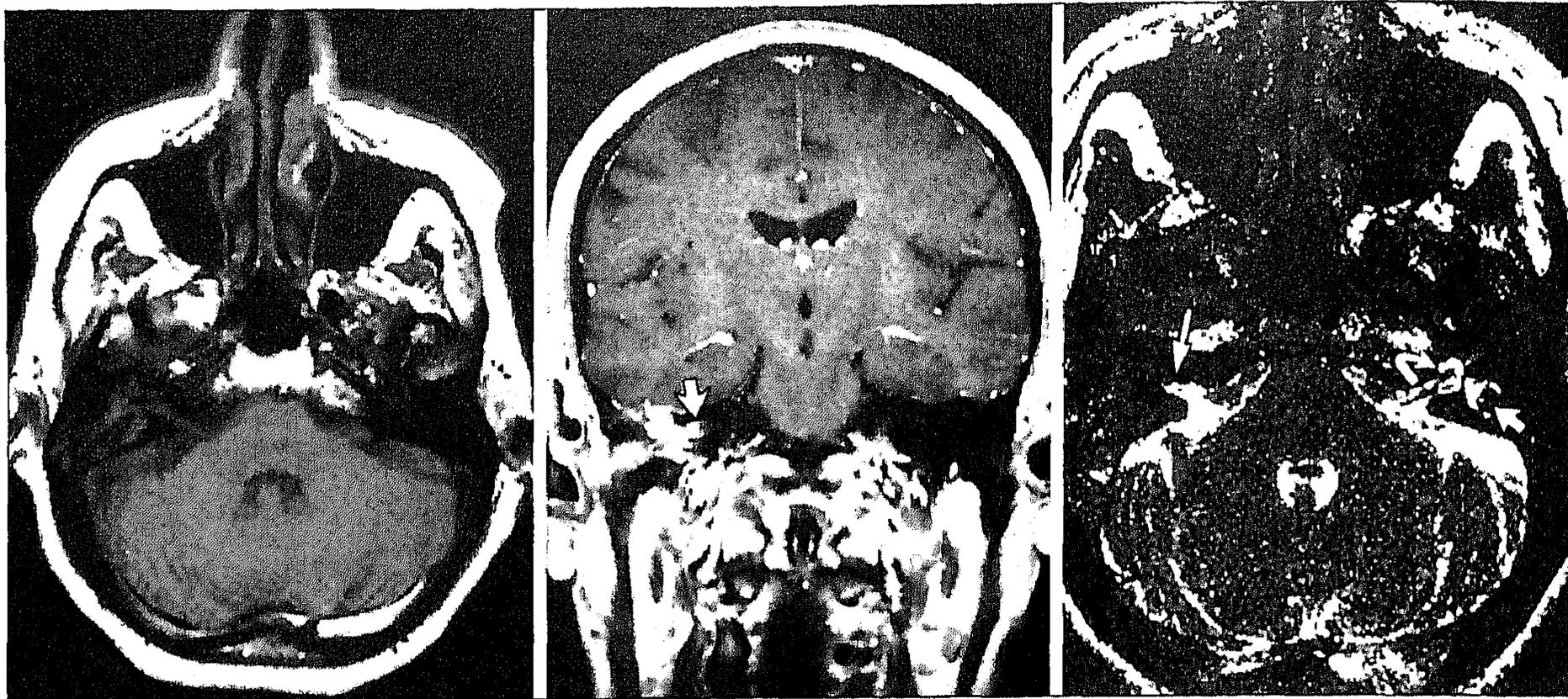


Figure 2. Case 3. Magnetic resonance imaging of the skull base. Left, T_1 -weighted image in the transverse plane showing pathologic soft tissues filling the right inner ear structures and mastoid. Center, T_1 -weighted image in the coronal plane after gadolinium-diethylene-triamine-tetraacetic acid contrast enhancement showing a strongly enhancing mass with extension in the internal auditory canal (white arrow). Right, T_2 -weighted image. Normal signal intensities on the nonaffected left side in the cochlea (curved white arrow) and labyrinth (thick white arrow) and total signal loss on the affected right side. The fundus of the internal canal on the right side is filled with a mass (long white arrow).

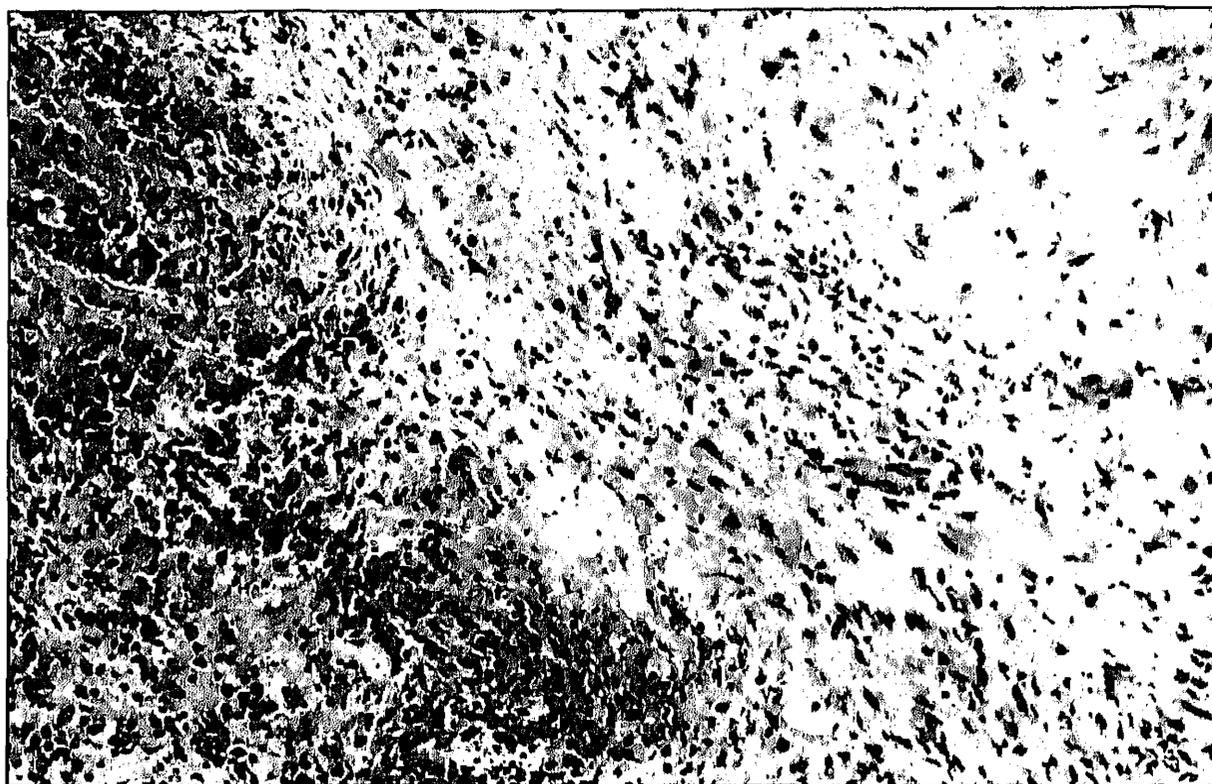


Figure 3. A 6- μ m paraffin section demonstrating a moderately cellular process consisting of loosely arranged plump spindle cells with a patchy mononuclear inflammatory infiltrate (hematoxylin-eosin, $\times 40$).

coronal planes showed that the contours and diameter of the internal auditory canal, fundus, geniculate ganglion, and cochlea were normal. The contours of the vestibule and lateral semicircular canal could not be detected, while the superior and posterior semicircular canals were widened. The ossicles were visible and in their normal anatomic position. Soft tissue was present in the tympanic cavity, and the pneumatized mastoid cavity was not aerated (**Figure 1**). Magnetic resonance imaging showed a strongly enhancing mass in the

middle and inner ear and in the cochlea and internal canal. Pathologic signal loss of the inner ear structures was present on the T_2 -weighted images (**Figure 2**). A subtotal petrosectomy was performed, preserving the labyrinth and cochlea, and a tumor was removed from the internal auditory canal, the cochlea, semicircular canals, and the eustachian tube, which was finally sealed off. A blind sac closure of the external auditory canal was made, and the cavity was filled with abdominal fat. Histologic examina-

tion of the specimen was consistent with a fibroinflammatory pseudotumor.

HISTOPATHOLOGIC FINDINGS

Histologic examination in these three cases showed features consistent with a moderately cellular process, consisting of loosely arranged plump spindle cells (**Figure 3**). Nuclear pleomorphism and mitotic activity were minimal. A conspicuous patchy mononuclear inflammatory infiltrate was present. There was no birefringent material, and additional stainings did not reveal any microorganisms. These tumors were not covered by a distinct epithelial layer. Bone erosion was evident.

COMMENT

The term *inflammatory pseudotumor* is a confusing and possibly ambiguous designation for a recognized entity of unknown origin. For a long time, inflammatory pseudotumor has been confused with plasma cell granuloma or fibrous histiocytoma in the lung.²⁻⁴ Even the first description of two cases of inflammatory pseudotumors in the lung by Brunn⁵ does not correspond with the definition used today. We propose to follow the histologic typing of tumors of the upper respiratory tract and ear,¹ as used in the *International Histological Classification of Tumours*

series, according to the World Health Organization (Geneva, Switzerland) criteria. In this internationally accepted classification, the fibroinflammatory pseudotumor is described in the nasal cavity and paranasal sinuses, nasopharynx, larynx, hypopharynx, and trachea; but, interestingly, the same histologic entity is not described as occurring in the middle and inner ear. Indeed, to our knowledge, the occurrence of these inflammatory pseudotumors in the middle and inner ear, apart from one case involving the facial nerve,⁶ has not been described previously. We did find two reports on plasma cell granulomas in the middle ear,^{7,8} but, as stated before, it was clearly another histologic entity.

Apart from the abdominal organs, inflammatory pseudotumors may be found in the respiratory tract, especially in the lung. In the ear, nose, and throat region, single case reports on inflammatory pseudotumors of the subglottis,⁹ lymph nodes and neck,¹⁰ retropharyngeal and parapharyngeal space,^{11,12} paranasal sinuses,¹³ skull base,¹⁴ and, as was mentioned before, on the facial nerve,⁶ have been published, but, again, in all these publications, the histologic features are not uniform.

The pathogenesis of fibroinflammatory pseudotumors is unknown, although, according to the literature, it seems likely that it concerns a nonspecific inflammatory response to different stimuli, especially chronic infection.

In the lung, the prognosis of previously described inflammatory pseudotumors is excellent following surgical removal. Because these lesions may mimic malignancies clinically, it is obvious that lobectomy is the treatment of choice. Less is known about the follow-up in other sites. Our first patient has been followed up for 2.5 years, and our second patient has been followed up for 1 year. Both are free of complaints. Our third patient underwent surgery 9 months ago. The clinical picture is nonspecific except for the presence of total deafness induced by a myringotomy in one case and by surgical intervention in the other.

In all cases, high-resolution CT scans showed a soft-tissue mass with destruction of parts of the inner ear and contrast enhancement. Therefore, the

differential diagnosis consisted of lesions such as chronic inflammatory diseases, paragangliomas, and granulation tissue, and especially cavernous hemangiomas and facial neuromas.

We noticed a typical widening of the labyrinth and deformation of the vestibule in all our patients. In two patients, the ossicles were still recognizable. Chronic inflammatory diseases of the middle ear cavity showed, in most cases, an enhancing mass on MRI; but, to our knowledge, extension into the labyrinth or internal auditory canal as in our cases has not been described, nor has the typical destruction been observed on CT scan.¹⁵

Diffuse inflammatory infection of the petrous apex does not account for the bony destruction of the inner ear, especially of the labyrinth. The MRI studies revealed an enhancing mass in the middle and inner ear in all cases, showing a larger extension than was presumed on the CT scans. Extension into the internal auditory canal and cochlea could be distinguished particularly well. On T₂-weighted images, the labyrinth and cochlea show a normally high signal intensity, but in these cases signal intensity was reduced or even absent. A strongly enhancing mass could also raise the suspicion of a vascular tumor like a paraganglioma of the glomus tympanicum. However, the absence of the typically permeative bone changes does not fit with this diagnosis. Both CT and MRI gave additional information in all three cases. The pattern of the soft-tissue mass as seen on CT with the very distinctive destruction of inner ear structures and widening of the labyrinth seems to be a typical finding. Extension of the inflammatory changes into areas of destruction and areas of radiologic normality, as revealed by MRI, also seems to be a typical feature of an inflammatory pseudotumor.

In conclusion, fibroinflammatory pseudotumors are benign but locally destructive lesions that have not been described as occurring in the middle ear. Destruction of inner ear structures and widening of the labyrinth were present on CT scans in all our patients. The lesion also shows enhancement with intravenous contrast. At surgery, these tumors are firm and well delineated. Although surgical removal seems to be the treatment of choice, no clear prognosis can

be given regarding lesions in the ear because of the relatively short follow-up periods in these cases.

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