CASE REPORT

Fibroinflammatory Pseudotumor of the Temporal Bone

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ABSTRACT—Chronic inflammatory tumor-like lesions of the temporal bone represent a difficult clinical task for the skull base surgeon. Their osteolytic aggressiveness endangers vital structures and may not be controlled by surgery alone. We present the course of four cases of fibroinflammatory pseudotumor of the temporal bone which were treated by a combined approach of skull base surgery and chemotherapy. Three patients were deafened by the disease and underwent several operative measures. One patient was lost, most likely due to an arrosive bleeding of the internal carotid artery. The chronic and recurrent process could only be stopped by petrosectomy, followed by antiproliferative chemotherapy. Two patients were subsequently provided with a cochlear implant. The differential diagnosis, diagnostic, and operative options of this rare but severe disease are discussed.

Fibroinflammatory pseudotumor (FIPT) is a clinically malignant but histologically benign lesion of unknown etiology. Its manifestation has been described in several locations including the lung,[1,2] the lymph nodes,[3–5] the salivary glands,[6,7] the paranasal sinuses,[8,9] the trachea,[10] and the skin.[11] While some authors consider FIPT as a process reactive to chronic external stimuli,[1] others see it as a local manifestation of an otherwise systemic disease group including sclerosing cholangitis, Riedels' thyreoditis, and peritoneal fibrosclerosis.[7,12] Besides FIPT, several synonyms may be found in the literature, all describing the same entity, e.g., plasma cell granuloma, histiocytoma complex, xanthomatous pseudotumor, fibrous xanthoma, inflammatory myofibroblastic tumor, and inflammatory myofibrohistiocytic proliferation. The typical histological picture of inflammatory pseudotumor consists of proliferating spindle cells of fibroblastic, histiocytic, and myo-fibroblastic origin with a mixed inflammatory cell infiltrate. Plasma cells may be found predominantly. This highly reactive lesion grows infiltratively, thereby destroying the surrounding tissue architecture. Dependent on the localization of the tumor, vital structures may be involved and damaged. This is especially true for FIPT of the temporal bone where the integrity of the labyrinth, the brain, the carotid artery, and the facial nerve is endangered. As this aggressive, self-perpetuating lesion tends to recur, a combined surgical and systemical, i.e., chemotherapeutical, approach may be adequate. We report on four cases of FIPT of the temporal bone that we treated with radical skull base surgery and postoperative chemotherapy.

CASES

Case 1

A 39-year-old female was admitted to our clinic with a 4 year history of bilateral chronic middle ear disease. She had undergone ten middle ear operations on the left and seven on the right during which she was deafened in both ears. She complained of bilateral otalgia, recurrent left otorrhea and unsystematic vertigo. Otoscopy showed sclerosing granulation tissue in both middle ear cavities (Fig. 1). A computed tomography

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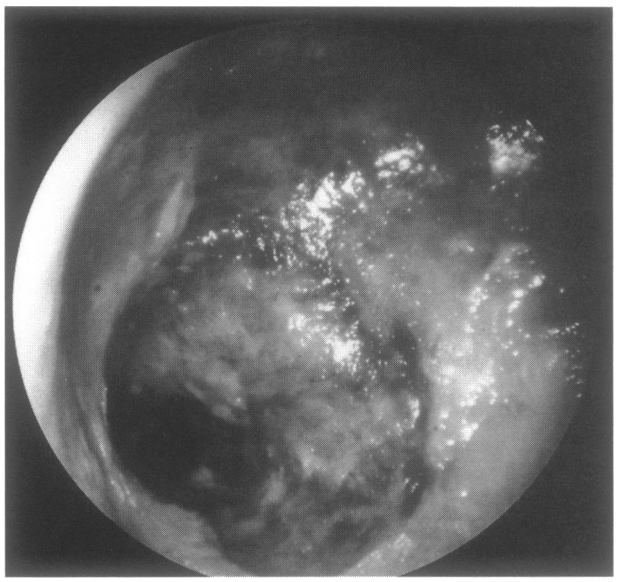

Figure 1. Otoscopic image of FIPT (patient 1). A firm, sclerosing granulation tissue of gritty consistency fills a large portion of the middle ear cavity.

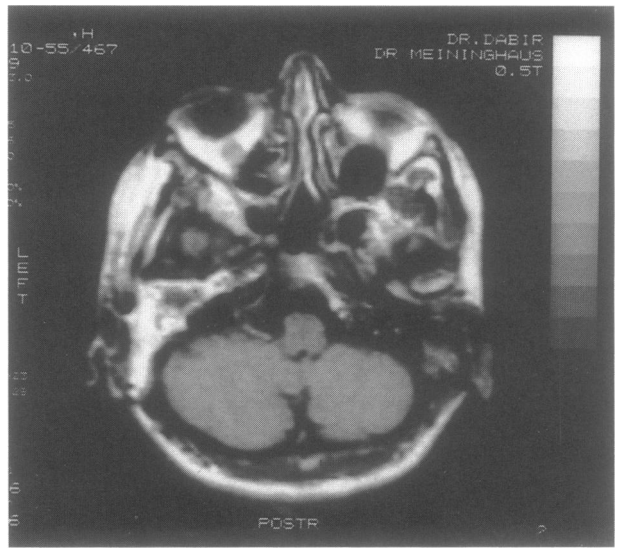

Figure 2. MRI of patient 1 at first presentation. A signal enhancing, soft-tissue mass occupying the left temporal bone is detected.

(CT) scan revealed a destructive process in the left temporal bone which could be confirmed by magnetic resonance imaging (MRI) (Fig. 2). However, it could not be differentiated if the extended bone erosions were caused by the disease or were sequelae of the multiple operative manipulations. Bone and lymphocyte scintigraphy showed an enhancement in the left temporal bone, confirming an active inflammatory process. A subtotal petrosectomy was carried out on the left side and the cavity was obliterated with abdominal fat. The histology showed the typical picture of FIPT with a mixed cellular infiltrate, spindle cells, and a surplus of extracellular matrix (Fig. 3). Postoperatively, a high-dose glucocorticoid regimen was begun; cortisone was tapered after 6 weeks. Four months later, a cochlear implant operation was carried out on the right side, as the osteolytic destruction had left the cochlea intact and a promontory test had given good results. The procedure was uneventful and all electrodes could be placed into the cochlea.

Four months later, the patient experienced massive pain, vertigo, and a swelling behind her right ear. CT scan and scintigraphy showed an active process in her right temporal bone, so a subtotal petrosectomy was carried out, the cochlear implant was explanted, leaving only the electrodes *in situ* (Fig. 4) and a postoperative immunosuppression was begun (intravenous corticoids plus cyclophosphamide), as histology confirmed the diagnosis of FIPT. Ten months later during which the patient had been perfectly well, the electrodes were explanted and a new cochlear implant was inserted (Fig. 5). Rehabilitation was begun and the patient's speech perception was excellent. Four months later she was again admitted to our clinic complaining of an acute and massive otalgia and a fluctuant swelling behind her left ear. She was reoperated and the parotid gland which had been infiltrated by the tumefactive lesion was removed while facial nerve function was preserved. An intense postoperative immunosuppressive regimen was begun (corticoids, cyclophosphamide, α-interferon). Six months after the last operation, the patient is well and the cochlear implant is working appropriately.

Case 2

A 49-year-old female was presented to our outpatient clinic to investigate the chances for a cochlear implant operation. The patient had a 5 year history of bilateral chronic otitis media with subsequent complete hearing loss. The patient suffered from an autoaggressive psychosis during which she had perforated her ear drum several times with a cotton swab. She had undergone middle ear surgery twice on both sides. Her acute complaints were bilateral otalgia and suppurating discharge from both ears. A CT scan showed erosions of both cochleae and a soft tissue mass in the right temporal bone (Fig. 6). A subtotal petrosectomy was carried out on the right side, the defect was obliterated with abdominal fat, and a high-dose cortisone scheme was begun postoperatively. Concurrent intense psychiatric counseling was carried out. Under this regimen the patient was free of symptoms and had no further complaints.

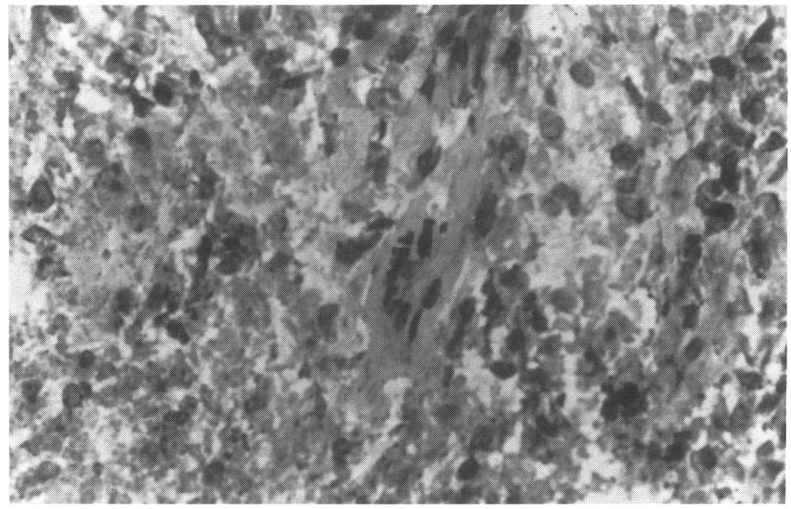

Figure 3. Typical histological picture of FIPT (patient 1, HE staining, 400X): spindle cell nests, formed by myofibroblasts, fibrotic tissue, and a mixed cellular infiltrate composed of plasma cells and neutrophil leucocytes.

Eight months later, a subtotal explorative petrosectomy was carried out on the left side and the remaining granulation tissue was removed. Again, high-dose corticoids were given postoperatively for 6 weeks. Three months after the operation, a cochlear implant was placed in the left cochlea. All electrodes could be inserted. Under close psychiatric monitoring, the rehabilitation phase was successfully passed and the patient developed an excellent degree of speech perception. Thirty months after implantation, the patient shows no signs of recurrent disease and is well balanced.

Case 3

A 35-year-old female patient presented with acute facial paralysis, deafness in her right ear and a profound combined hearing loss on the left. She had been suffering from bilateral otalgia and suppurative otitis media for 3 years. She had undergone multiple ear surgery (six in the right ear; three in the left ear) within this period with no long-lasting improvement. A CT scan showed a soft tissue mass in the right temporal bone with osteolytic destruction of the labyrinth. The left mastoid showed typical postoperative changes but no suspect abnormality. A subtotal petrosectomy was carried out with intraoperative facial nerve monitoring. The facial nerve was intact and could be freed from the tumefactive mass. At histologic diagnosis, fibroinflammatory pseudotumor of the temporal bone was determined. Postoperatively, an immunosuppressive therapy scheme was begun (six courses of cyclophosphamide and subsequent α-interferon under the supervision of the hematology/oncology clinic). With this treatment, facial nerve function returned to normal while the hearing did not improve. The patient shows no recurrence after 2 years.

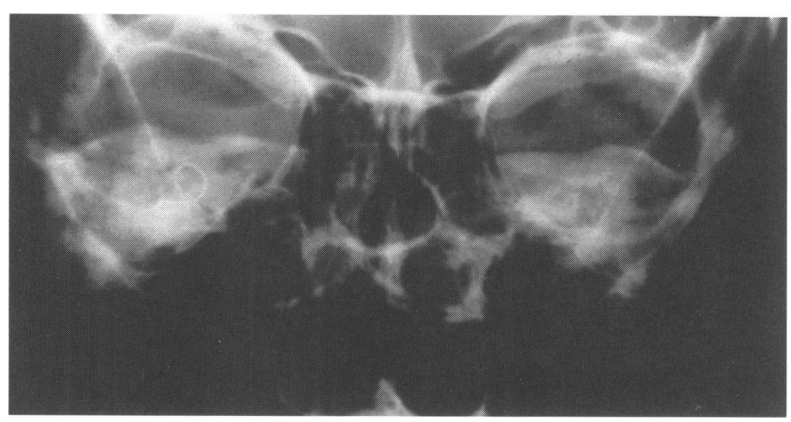

Figure 4. Transorbital X-ray of patient 1 after the partial explantation of the cochlear implant due to the recurrence of FIPT. The intracochlear part of the electrode is still in situ.

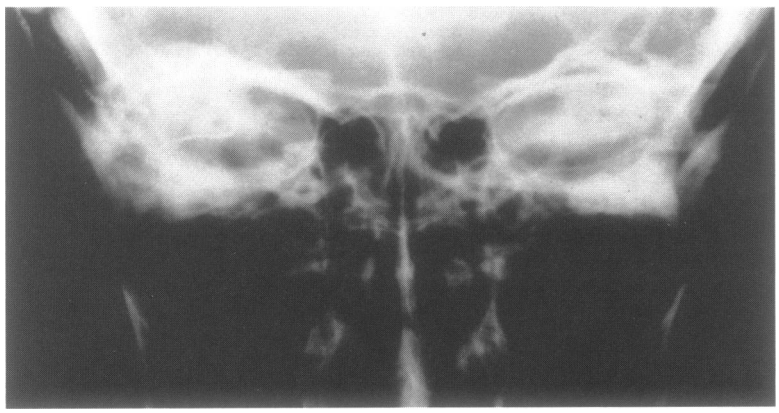

Figure 5. Transorbital X-ray of patient 1, 12 months later. A cochlear implant was reimplanted successfully.

Case 4

A 59-year-old male was admitted to the hospital with a 2 year history of recurrent bilateral otalgia, otorrhea, and progressive hearing loss. He had undergone middle ear surgery of both ears twice before when he had been diagnosed with chronic bilateral mastoiditis. The last operation had been carried out 8 months before admission. Otoscopy showed subtotal defects of both tympanic membranes with polypoid structures in the middle ear. A CT scan revealed a soft tissue mass in both mastoids with focal erosions of the right labyrinth. The patient was deaf in his right ear and had a combined, nearly complete hearing loss in the left. A petrosectomy was carried out in the right ear and a radical mastoidectomy was carried out on the left ear 2 weeks later. The histology showed the typical picture of FIPT on both sides. Postoperatively, a combined immunosuppressive and cytotoxic therapy was begun (30 mg prednisone, 1300 mg cyclophosphamide/day). Three weeks later, the patient experienced a sudden and complete loss of vision in the right eye, a right paresis of the abducent nerve, and a partial paralysis of the seventh cranial nerve on the same side. CT showed a soft tissue mass at the tip of the right orbital funnel which extended to the cavernous sinus (Fig. 7). An open biopsy was carried out which revealed unspecific granulation tissue, resembling FIPT. High-dose corticoids were given intravenously with no clinical improvement. Seven days later, the patient's left vision deteriorated dramatically

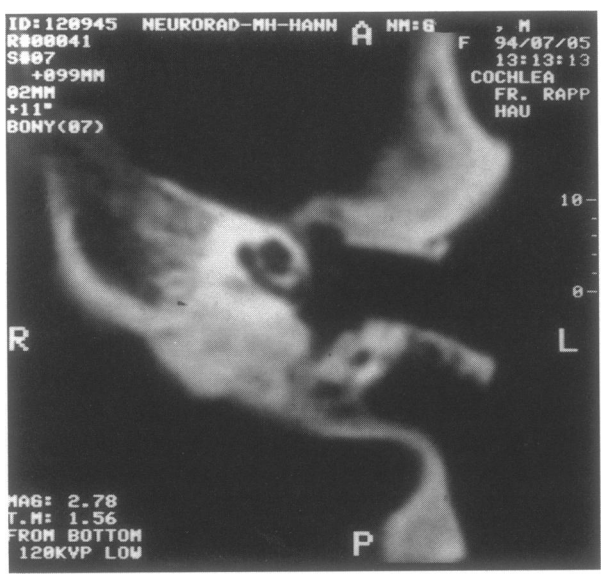

Figure 6. High-resolution CT scan of the left temporal bone (patient 2). FIPT has eroded the basal turn of the cochlea.

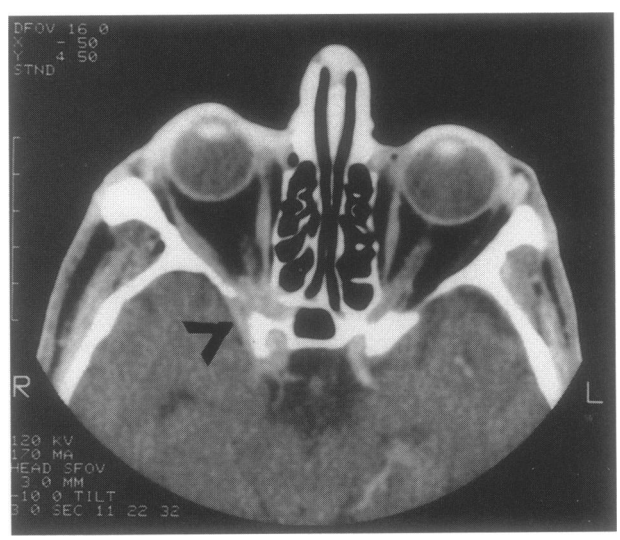

Figure 7. Axial CT scan of patient 4. A soft-tissue mass infiltrates the tip of the right orbital funnel (arrowhead). Deeper CT sections show that the tumor originates from the temporal bone.

and before another radiologic procedure could be carried out, the patient succumbed to the lethal effects of a suspected arrosive bleeding of the internal carotid artery. Unfortunately, the relatives rejected an autopsy.

DISCUSSION

Fibroinflammatory pseudotumor of the temporal bone is a rare but nevertheless important disease as its highly aggressive clinical course may endanger and destroy important or even vital structures. The skull base is a location for this pathology that has only been reported twice thus far.[13,14] The important, common clinical features in the four presented cases are the long history and the high recurrence rate of the disease which lead to multiple ear operations, pain, and the appearance of a soft tissue mass which caused an erosive destruction of the temporal bone and showed considerable signal enhancement in the MRI or scintigraphic imaging techniques. The diagnosis has to be made histologically in concordance with an international pathologic classification of tumor-like lesions (No. 76820).[15] The lesion typically shows atypical, spindle-shaped fibroblasts with pleomorphic nuclei, a mixed cellular infiltrate of plasma cells and neutrophil leucocytes, and areas of fibrosis with poorly defined margins. Besides the careful analysis of the surgical specimen, which should be carried out by an experienced pathologist, diagnostic measures consist of imaging techniques (CT, MRI, bone scintigraphy, and leukocyte scintigraphy) to evaluate bone erosion, intracranial extension and disease activity,[16,17] laboratory analysis (erythrocyte sedimentation rate, white blood counts, C-reactive protein, ANCA, ANA, and complement CH50), and the search for possible causative microorganisms including highly sophisticated methods as the polymerase chain reaction for the detection of mycobacteria.

The differential diagnosis includes specific and unspecific inflammatory diseases, e.g., vasculitis,[18] Wegener's granulomatosis,[19] arachnoid granulations,[20] chronic osteomyelitis,[21] pachymeningoencephalitis,[22] sarcoidosis, cholesterol granuloma, or cholesteatoma;[23,24] malignomas, e.g., adenocarcinoma,[19] squamous cell carcinoma,[25] or chondrosarcoma;[19,26] and more systemic diseases, e.g., histiocytosis X or eosinophilic granuloma,[19,27,28] and lymphoproliferative diseases.[4] Specifically, the diagnosis of infectious ear disease must be questioned when there is a lack of responsiveness to acceptable therapies.[19]

The aggressive and obviously life-threatening course of the disease calls for an aggressive and multidisciplinary clinical approach. Radical surgical measures, which included total petrosectomy in our cases, should be undertaken as a first-line therapy. Nevertheless, facial nerve function and residual hearing should be preserved which can be facilitated by intraoperative monitoring. If the cochlear nerve is preserved, a cochlear implant operation may restore the hearing successfully. We suggest, however, that the implantation should only be carried out after the patient has fully recovered and neither scintigraphy nor laboratory parameters indicate a residual inflammatory lesion. An immediate postoperative, immunosuppressive chemotherapy should be administered. The anti-inflammatory and antiproliferative effects of the drugs block the disease-promoting activity of immune cells and humoral factors (cytokines, growth factors) that are present in high concentrations in normal healing wounds, and which are responsible for the self-perpetuating course of FIPT.[5,11] The medical treatment should be coordinated closely with a hematologist/oncologist. Under combination therapy with high-dose corticoids and cyclophosphamide, followed by α-interferon in two of our four cases, we did not see any recurrence of the disease. However, the wound-healing was retarded in all patients and required intensive local care over more than 2 weeks. The postoperative management includes the relatively liberal prescription of powerful analgetics. As described above, severe and deep-seated pain is a typical feature of the disease.

The long course and the high recurrence rate force the clinician to begin a close and lifelong follow-up.[13] This strategy is even further supported by a recently published report that an inflammatory pseudotumor progressed into a full-blown spindle cell sarcoma after several recurrences.[29] In this case, the gradual transformation into malignancy took a total span of 12 years.

In conclusion, fibroinflammatory pseudotumor of the temporal bone is a chronic, recurrent, and serious disease. Its destructive course endangers all structures of the temporal bone. The adequate treatment is multidisciplinary and consists of radical and consequent surgery, immunosuppressive chemotherapy, and potent analgesia.

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