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Inflammatory Pseudotumor of the Temporal Bone: A Case Series

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Introduction

Inflammatory pseudotumor (IPT) is a rare, idiopathic, inflammatory process that is histopathologically benign albeit locally invasive. IPT was first described by Gleason and Busse in 1903¹, and then further characterized by Birch-Hirschfeld in 1905². In 1954, Umiker and Iverson referred to this disease as “inflammatory pseudotumor” because the aggressive clinical course is similar to malignant tumors³. Since then it has also been referred to as plasma cell granuloma, plasma cell pseudotumor, inflammatory histiocytic tumor or inflammatory myofibroblastic tumor. IPT can present as a single lesion or multiple inflammatory masses. It is most commonly found in the lung but can occur anywhere in the body. Approximately 5% of cases involve the head and neck with the orbit being the most common site⁴. Involvement of the temporal bone is uncommon and is likely under-represented in the literature^{4,5-7}.

The etiology of IPT is still largely unknown. It is histologically characterized as polymorphous inflammatory cell infiltrates with variable amounts of fibrosis, necrosis, granulomatous reaction, and myofibroblastic spindle cells. Although benign, IPT can be locally invasive to surrounding nerves, bone, and soft tissue. Its aggressive nature and

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debilitating manifestations are often mistaken for a neoplastic or infectious process. Some authors suggest it may be post-infectious in nature or a local manifestation of a systemic inflammatory process such as IgG4 related sclerosis⁴.

While reports of IPT of the temporal bone have been increasing over the past 10 years, this pathologic entity is still relatively obscure. Diagnostic and management algorithms have yet to be standardized, which can lead to delay in appropriate care. This is a review of seven patients diagnosed and treated at a tertiary care center which describes our recent experience in the management of IPT of the temporal bone with an average 17.8 month follow up. The goal of this report is to emphasize the clinical findings, diagnostic tools and treatment modalities for IPT, to raise awareness of this entity, and to facilitate thorough and expeditious care.

Materials and Methods

A retrospective review of 7 patients diagnosed with IPT of the temporal bone from January 1, 2014 to January 1, 2016 was performed at a single tertiary medical center. The following data were extracted: patient demographics, presentation, associated operative reports, imaging, histopathology, treatment, follow-up, recurrence, and final outcome of disease.

Results

Demographics, Symptoms & Signs

There were a total of five males and two females (n = 7) with a diagnosis of IPT of the temporal bone. The mean age at presentation was 41 years old, with a range from 8 years to 75 years old. The most common presenting symptom was hearing loss (7/7, 100%), headache (4/7, 57%), otorrhea (4/7, 57%), otalgia (2/7, 29%) and disequilibrium (2/7, 29%). Tinnitus was less common (1/7, 14%). Clinical findings included an inflammatory aural polyp that was present (4/7, 57%), followed by facial nerve paralysis (2/7, 29%) and purulent external auditory canal exudate (2/7, 29%). (See Table 1). Reported levels of IgG4 were 20 cells/hpf for patient #5, 12 cells/hpf for patient #2, 85 cells/hpf for patient #1 and 65 cells/hpf for patient #6.

Radiographic Findings

Multi-planar high-resolution computerized tomography was obtained for 7/7 patients and magnetic resonance imaging was acquired for 6/7 of the patients. All patients demonstrated involvement of the temporal bone. Diffuse dural thickening, enhancement and marked T2 hypo-intensity of abnormal soft tissue were found in 5/6 patients (Figure 1a). 2/6 (33%) displayed extension into the internal auditory canal (Figure 1b). Leptomeningeal enhancement and thickening were present in 2/6 (33%) of cases (Figure 1c). Mastoid effusions were present in 3/6 (50%). Some sort of osseous sclerosis or erosion occurred in 4/7 (57%) (Figure 2a). Significant mass extension included one case that extended along the eustachian tube into the nasopharynx, one that encased and constricted the sigmoid sinus (Figure 3), and another that surrounded the internal carotid artery and jugular vein circumferentially and spread into the jugular foramen (Figure 2b).

Histopathology

All patients underwent biopsy. Histopathological analysis was consistent with dense, chronic inflammation with numerous giant cells in the setting of hyalinized fibrosis with storiform features with in all 7/7 patients (Figure 4a, 4c). The chronic inflammation was composed of lymphocytes, plasma cells, and macrophages. A stain for IgG4+ plasma cells was positive in 4/7 (57%), although the number of was significantly increased in only 2/7 patients (Figure 4b).

Treatment

The most frequent treatment was a combination of systemic steroids, antibiotics, and cortical mastoidectomy (5/7), with two of these patients also receiving Rituximab. One patient was treated with steroids and mycophenolate mofetil. One patient received steroids and palliative radiation therapy. Six out of seven patients were maintained on chronic oral steroid therapy, with 1/7 on topical steroids. At the conclusion of this study six patients did not necessitate therapy and one patient was maintained on low dose oral steroids. All patients have had no progression of symptoms. Mean follow up was 17.8 months. (See Table 1)

Discussion

Inflammatory pseudotumor of the temporal bone is a rare and incompletely understood entity. Most of our knowledge of IPT arises from extra-temporal case studies and systematic reviews that have emerged over the past several decades. (See Table 3). One review in 2013 identified only 29 cases⁶. We are only able to speculate as to why our institution has had a high population of patients with the diagnosis of IPT over the past several years despite no objective data to support a long-term, significant increase in diagnosis of IPT within the head and neck or other areas of the body. While this may be purely incidental, a more likely explanation is a raised index of suspicion thanks to close communication with our radiology and pathology departments, which helped to identify this pathology. This is consistent with the overall low and delayed recognition of IPT in the absence of a heightened awareness to the disease. Further investigation into possible risk factors, either genetic or environmental, will continue as more data is obtained. We hope to improve current diagnostic and therapeutic protocols through our own experience with these seven patients.

There is growing evidence that IPT is a subtype of IgG4-related disease, defined by elevated concentrations of IgG4 antibody in tissue and serum. The first case of IgG4-related disease was discovered in sclerosing pancreatitis⁸. Near 90% of patients with sclerosing pancreatitis have high serum IgG4 level.⁹ This pattern of IgG4 up-regulation and the presence of IgG4 associated plasma cells have also been recognized in cases of IPT (Figure 4b), as demonstrated in several of our patients, although the exact correlation with IgG4 is poorly understood.¹⁰ Reporting of IgG4 levels have been inconsistent in our study. The highest level reported, 85 cells/hpf, did correlate with the most aggressive, recalcitrant clinical course.

The presenting symptoms of IPT are non-specific, and the differential diagnosis includes a wide range of infectious, inflammatory, neoplastic, granulomatous and autoimmune

disorders. Initial treatment ultimately depends on the acuity of the clinical presentation. We found that most of our patients had been evaluated several times in the primary care setting prior to our diagnosis. Symptoms of IPT are usually recalcitrant to initial forms of therapy including topical or oral antibiotics. Persistent symptoms should raise suspicion and initiate escalation of care and diagnostic interventions.

Appropriate imaging is critical in the evaluation and diagnosis of IPT. High resolution, multi-planar, contrast enhanced computerized tomography and magnetic resonance imaging are often necessary.

While the radiologic features of IPT are variable and nonspecific, certain features have been consistent across our patient population. Lesions may present with low, equal or high attenuation compared with the surrounding tissue on CT, depending on the varying degree of inflammation and fibrosis. Osseous erosion can also occur and may suggest a more aggressive inflammatory process. Magnetic resonance is more specific, and will often demonstrate iso- or hypointense soft tissues on T1- weighted images, and marked hypointensity on T2-weighted images, which may reflect the fibrotic nature of these lesions^{5, 11}. Adjacent dura also may thicken and enhance. In contrast to the hypointensity of IPT, primary malignant tumors such as chordoma, nasopharyngeal carcinoma invasion, and chondrosarcoma are usually hyperintense on T2¹².

While characteristic imaging may be the first identifiable feature of IPT, an aggressive or invasive infectious process must be ruled out. Depending on the extent of the disease, empiric antibiotics may be employed after obtaining a complete blood count, blood cultures and possible lumbar puncture for cerebrospinal fluid culture. All potential sources of infection must be considered. Consultation with infectious disease may also be warranted. In our experience, systemic antibiotics were not beneficial after the diagnosis of IPT and should be used judiciously during long-term management of this disease, as there may be a co-existing infection within the mastoid.

Biopsy is the gold standard for diagnosis of IPT as malignancy must be excluded for all suspicious lesions¹³. Current histological features used for the diagnosis of IPT show dense aggregates of fibroblastic and myofibroblastic cells mixed with chronic inflammatory cellular components (Figure 4c). Although some of the features of IgG4 -related disease, e.g. storiform pattern of fibrosis, and increased numbers of IgG4 plasma cells can be seen, the complete pattern of IgG4 -related disease, including the characteristic obliterative phlebitis, is often not identified. Characteristic features of Inflammatory Myofibroblastic Tumor (IMT) such as intranuclear eosinophilic inclusions may be present. A true malignancy, like spindle cell squamous cell carcinoma or Inflammatory Myofibrosarcoma, is ruled out by the typical immunohistochemical and morphological features of these entities.

A thorough autoimmune evaluation should also be considered on any lesion concerning for IPT. This includes a standard serological panel to rule out Wegener's granulomatosis, Sarcoidosis, Rheumatoid Arthritis, Sjogren's Syndrome and Systemic Lupus Erythematosus. Consultation with rheumatology or immunology may also be indicated.

Treatment of IPT of the temporal bone largely depends on location, degree of local invasion, and amenability to surgical resection. Previous authors have noted that these lesions tend to be more aggressive when compared with pseudotumor in other locations¹⁴. Surgical resection, i.e. mastoidectomy, in combination with steroid therapy has been the most common treatment of choice amongst these patients¹⁵. Of the six patients in our series, three of them underwent complete mastoidectomy with biopsy. Subsequently, these three patients were managed with a course of antibiotics and chronic steroid therapy without the need for radiation or further immunotherapy. Surgery has generally been required for diagnosis, but awareness of these lesions may prevent any unnecessary radical procedures in specific cases. We hypothesize that due to the fibrotic nature of IPT, aggressive mesotympanic resection may not be indicated as this may result in irreversible damage to the sound conduction mechanism or produce excessive post-op fibrosis. Such an outcome can potentially be avoided by limiting the resection to the mastoid and allowing the mesotympanic disease to be better controlled with medical therapy. A persisting tympanic membrane perforation, if present, can be repaired in the future once the disease process has been controlled. Currently there is no role for radical surgical resection secondary to morbidity. IPT lesions tend to be very responsive to corticosteroid treatment. Prior studies have demonstrated an initial response rate of approximately 80%¹⁵. In our case study, the patients with IPT extension past the temporal bone required adjuvant therapy, suggesting that intracranial involvement may be a predictor of the necessity of adjuvant therapy.

Due to the chronic nature of this disease, recurrence rates have been documented in greater than 20% of IPT of the head and neck and complete remission rates around 40-50% after initial therapy¹⁶. To prevent relapse, prolonged administration of steroid therapy has been recommended although IPT may recur during steroid taper. In this setting, other therapies may be necessary to prevent further progression. As exhibited by several of our patients, treatment modalities such as radiotherapy, the monoclonal anti-CD20 antibody, Rituximab, and the inosine monophosphate dehydrogenase inhibitor mycophenolate mofetil (Cellcept®), may also play a role in the treatment of IPT. In our study, patient #1 received Rituximab for recalcitrant disease after several courses of high dose steroids. Patient #5 also received Rituximab due to intolerance and complications from chronic steroid therapy. Patient #3 received mycophenolate mofetil as a recommendation from rheumatology service. He developed recurrent disease in the orbit and had poor tolerance of chronic steroids. Patient #4 underwent radiation therapy for recurrent symptoms with inability to wean high dose steroids. (See Table 2)

Radiotherapy has been evaluated as an option for induction of long-term remission in patients with refractory IPT. Prabhu et al found that seventeen of twenty patients exhibited response to radiotherapy in treating orbital pseudotumor¹⁶. Radiation has the potential to reduce the dose of steroids, or even allow for their discontinuation completely¹⁷. These potential benefits need to be weighed against the risks of radiation induced chronic osteitis of the temporal bone and the potential for damage to the neurosensory epithelium in the otic capsule, neither of which is a problem in cases of soft-tissue IPT. Several studies have also evaluated the role and efficacy of Rituximab for treatment of inflammatory pseudotumor. Although the exact mechanism of the response to Rituximab is unclear, tumor response is most likely due to induction of apoptosis secondary to antibody-dependent cell mediated

toxicity and complement activation. Garcia et al. demonstrated a sustained response to Rituximab of inflammatory pseudotumor of the jaw with perineural, intracranial invasion¹⁸. Another study by Chen et al. evaluated the treatment combinations of Rituximab, systemic corticosteroids, and radiotherapy, which exhibited sustained response in orbital inflammatory disease with continued remission¹⁹. Furthermore, Suhler et al determined that Rituximab has the potential to allow for corticosteroid dose to be reduced by 50% in the treatment of orbital inflammation²⁰.

Immunosuppressive therapies such as mycophenolate mofetil (MMF) (Cellcept®) have also been investigated. Moss et al reported meningeal inflammatory pseudotumor response to mycophenolate mofetil, which eliminated corticosteroid reliance and side effects in both patients described in their case report²¹. Hatton et al also used mycophenolate mofetil as a steroid sparing alternative for idiopathic orbital inflammation in the setting of disease recurrence that was refractory to steroids, in addition to treating relapse in a patient with poorly controlled diabetes mellitus. In this study, mycophenolate mofetil was stopped due to nausea in one patient, although all others (n=4) saw complete resolution of inflammation. They reported that the median time between disease onset and mycophenolate mofetil treatment was 4.5 years.²² Another study demonstrated how MMF was able to control disease within a median time of 3.5 months in 82% of patients with orbital pseudotumor. This allowed for taper of prednisone to less than or equal to 10mg a day²³. Although the data on MMF in the application of IPT of the head neck is still limited to small case studies involving orbital pseudotumor, it is likely an effective modality of adjuvant treatment for IPT in the temporal bone.

Conclusion

Inflammatory pseudotumor of the temporal bone is a rare pathologic entity. Despite its chronic and benign nature, this process can be locally aggressive, and present acutely with significant impact on neurological function and quality of life. It is becoming more evident that IPT is a distinct pathologic process with characteristic imaging and histopathological patterns. Timely recognition and an accurate diagnostic evaluation will lead to more appropriate treatment and expedited care. Maintaining a high index of suspicion and obtaining early surgical biopsy to confirm the diagnosis of IPT and differentiate it from other aggressive pathologies, is necessary in order to avoid long courses of antibiotics in patients with these aggressive lesions of the temporal bone. In our experience, the combination of surgery, and corticosteroids was the mainstay of treatment. Notably, the primary role of surgery is obtaining tissue for histopathological examination. Recalcitrant disease or poor tolerance of chronic steroid therapy may require adjunctive therapeutic interventions. This multi-modal approach may also be warranted in cases with advanced intracranial involvement. As the understanding of the pathophysiology and molecular pathways of inflammatory pseudotumor becomes elucidated, targeted molecular therapies may play an even greater role. Furthermore, we have come to understand that while this disease may present as acute exacerbations, it tends to behave more as a chronic process and will necessitate long term follow up.

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Figure 1.

Figure 1a. Axial T2 weighted MRI through the posterior fossa revealing marked T2 hypointense sheet like dural thickening (red arrow) along the dorsal surface of the right petrous temporal bone. Note the presence of mass effect upon the cerebellum (white arrow)

Figure 1b. Involvement of the dura of the IAC

Figure 1c. Leptomeningeal enhancement and thickening

Figure 2.

Figure 2a. Coronal CT image shows soft tissue in the middle ear cavity with dehiscence of the tegmen tympani (arrow).

Figure 2b. Coronal contrast enhanced CT images demonstrate a poorly enhancing soft tissue lesion encasing and constricting the right internal carotid artery

Figure 3.

Axial fat suppressed T1 weighted contrast enhanced image through the skull base reveals smooth dural enhancement constricting the sigmoid sinus (black arrows) and extending into the internal auditory canal (red arrow).

Figure 4.

Figure 4a. (Mag 20×) : Mucosal tissue with dense chronic inflammation and numerous giant cells

Figure 4b. (Mag 20×): Mildly increased IgG4 plasma cells. IgG4 is marked in brown

Figure 4c. (Mag 40×): Chronic inflammation and cholesterol clefts/granuloma; fibrosis with storiform features

Table 1
Patient Demographics, Symptoms and Signs, and Treatment

Characteristic	Patients (n=7)
Age in years, mean +/- SD (range)	41 +/- 23 (8-75)
Male gender, n (%)	5 (71)
Female gender, n (%)	2 (29)
Ethnicity	5 (AA), 2 (Caucasian)
Presenting Symptoms	
Hearing loss, n (%)	7 (100)
Headache, n (%)	4 (57)
Inflammatory polyp, n (%)	4 (57)
Vertigo/ disequilibrium, n (%)	2 (28)
Facial nerve paralysis, n (%)	2 (28)
Otorrhea, n (%)	4 (57)
Otalgia, n (%)	2 (28)
Anatomical presentation of IPT	
Limited to Mastoid bone, n (%)	3 (43)
Mastoid bone, Neck, n (%)	1 (14)
Temporal bone, Intracranial extension n (%)	3 (43)
Treatment modality combinations	
Steroids, Antibiotics, Surgery, n (%)	3 (43)
Steroids, Antibiotics, Surgery, Rituximab, n (%)	2 (29)
Steroids, Antibiotics, Mycophenolate Mofetil, n (%)	1 (14)
Steroids, Antibiotics, Radiation, n (%)	1 (14)
Distribution of Specific Treatment Modalities Distribution	
Steroids (Topical/Systemic), n (%)	5 (71) /6 (86)
Antibiotics, (Topical/ Systemic), n (%)	6 (86) /4 (57)
Mastoidectomy, n (%)	5 (71)
Rituximab	2 (29)
Palliative radiation, n (%)	1 (14)
Mycophenolate Mofetil, n (%)	1 (14)

Table 2

Patient Clinical Courses

Patient	Age	Initial Presentation	Initial Treatment	IgG4 Cells/HPF	Chronic Treatment	Interim Course	Clinical Status	Total Follow Up (months)
1	35	Otalgia Hearing Loss Disequilibrium Otorrhea Headaches Aural Polyp	Topical/Oral/IV antibiotics High dose steroids Mastoidectomy Rituximab 900mg per week/4 weeks	85	Steroid Taper	Facial N. Paralysis (House Brackmann 6) Profound Mixed HL	No headache, otalgia, otorrhea No disequilibrium CN 7 House Brackmann 2 Fluctuating hearing loss No continued therapy	26
2	43	Hearing Loss Otorrhea Aural Polyp	Topical/Oral antibiotics Mastoidectomy PE tube	12	Topical steroids	Moderate-Severe Mixed Hearing Loss	No otorrhea or polyp Moderate-Severe Mixed HL No continued therapy	15.5
3	75	Headache Disequilibrium Hearing Loss	Oral antibiotics High dose steroids Mycophenolate mofetil 1000mg bid for 7 months	N/A	Steroid Taper Mycophenolate mofetil	Left orbital IPT Right orbital IPT	No headache or disequilibrium Stable hearing loss No continued therapy	24
4	49	Headache Hearing Loss	Topical/Oral antibiotics PE tube High dose steroids 25Gy radiation therapy	N/A	Steroid Taper	Recurrent headaches Moderate-Severe SNHL	No headaches Stable hearing loss No current therapy	23
5	60	Headache Hearing Loss	IV antibiotics Mastoidectomy High dose steroids Rituximab 800mg/month	20	Steroid Taper Rituximab	Facial N. Paralysis (House Brackmann 6)	No headaches CN 7 House Brackmann 6 Stable hearing loss No continued therapy	12
6	18	Otorrhea Hearing Loss Aural Polyp	Topical antibiotics Mastoidectomy High dose steroids	65	Steroid Taper	Mild -Severe Mixed HL	No otorrhea Resolution of polyp Stable hearing loss.	12
7	8	Otalgia, otorrhea Hearing Loss Aural Polyp	Topical/Oral antibiotics Mastoidectomy High dose steroids	N/A	Steroid Taper	Moderate-Profound Mixed HL	Resolution of Polyp Mild SNHL Continued low dose steroids	12

Table 3
Literature review adapted from Tian et al.⁶

Author	Year	Patients	Treatment	Follow-Up
Yanagihara et al. ²⁴	1991	1	Surgery	NED
Benton et al. ²⁵	1992	1	Surgery, radiation	NED at 1 year
Nam et al. ²⁶	1994	1	Surgery, radiation	NED at 1 year
Wiseman et al. ²⁷	1995	1	Surgery	NED
Mulder et al. ²⁸	1995	1	Surgery	NED
Janicki et al. ¹²		1	Radiation, surgery	Ned at 2 months
Schönermark et al. ²⁹	1996	3	Surgery, steroids, cyclophosphamide Surgery, steroids Surgery, cyclophosphamide, IFN α	NED 6 months NED 30 months NED 2 years
Gasparotti et al. ³⁰	2003	1	Surgery, antibiotics	NED
Williamson et al. ¹³	2003	1	Surgery, steroids	NED 6 months
Cho et al. ³¹	2004	1	Steroids	Remission 3 mon
Cho et al. ³²	2007	1	Surgery	NED 1 year
Lee et al. ³³	2007	1	Surgery, steroids	NED 1 year
Lee et al. ³⁴	2007	2	Surgery Surgery, radiation, steroids	NED 3 years NED 8 years
Strasnick et al. ⁵	2007	2	Surgery, radiation Surgery, radiation	NED 8 years NED 10 years
Coulson et al. ³⁵	2008	1	Steroids	NED 18 months
Lee et al. ³⁶	2008	1	Surgery	NED 6 months
Allona et al. ¹⁴	2009	1	None	Remission 1 year
Ajibade et al. ⁷	2010	1	Surgery, steroids	Relapsed 4 months
Curry et al. ³⁷	2009	1	Surgery, steroids	NED 1.5 years
Santaolalla-Montoya et al. ³⁸	2008	1	Surgery, steroids	Died of disease
Galindo et al. ³⁹	2008	1	None	Remission 3 years
Goh et al. ⁴⁰	2009	1	Surgery, steroids	NED 3 years
Papanikolaou et al. ⁴¹	2012	1	Surgery	NED 6 months
Das et al. ⁴²	2013	1	Mastoidectomy	NED 3 years
Tian et al. ⁶	2013	3	Surgery, steroids, cyclophosphamide Surgery Surgery	3 recurrences. NED at 16 months NED 14 months NED 11 months.
Wang et al. ⁴³	2015	1	Corticosteroids	NED 22 months