Case Report

IgG4 Related Sclerosing Inflammatory Pseudotumor Simulating Cholesteatoma Of The Temporal Bone - A Case Report

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ABSTRACT

Introduction

To study an unusual entity like Sclerosing inflammatory pseudotumor of the temporal bone is a rare fibro-inflammatory, benign albeit locally aggressive pathological entity.

Case Report

We report a case of Sclerosing inflammatory pseudotumor of the temporal bone, simulating a cholesteatoma. A 61-year-old female presented with right otalgia, hearing loss, facial pain and imbalance. Ear microscopy revealed a pulsatile mass in the external auditory canal. HRCT temporal bone and MRI reported soft tissue mass in the right middle ear and mastoid air cells, causing attenuation of the ossicles with erosion through the tegmen and mastoid cortex. The patient underwent a canal wall down tympanomastoidectomy. The histopathology with immunohistochemistry was consistent with IgG4 related sclerosing inflammatory pseudotumor.

Discussion

This case report explores the similarities as well as the clinical differences between cholesteatoma and IgG4 sclerosing inflammatory pseudotumor of temporal bone, along with the diagnostic and treatment challenges. It is often mistaken for neoplasm or cholesteatoma, hence should be considered as a differential diagnosis. A combination of surgical resection and prolonged corticosteroid therapy was advocated as the mainstay of treatment.

Keywords

Inflammatory Pseudo Tumor; IgG4 Related Disease; Sclerosing Inflammatory Pseudotumor

Gleason and Busse in 1903[1] described inflammatory pseudotumor as an unencapsulated benign tumor of unclear etiology commonly found in the lungs. It may be postinfectious in nature or a local manifestation of a systemic inflammatory process such as IgG4 related sclerosis. It has also been referred to as plasma cell granuloma, inflammatory myofibroblastic pseudotumor, inflammatory histiocytoma and inflammatory fibrosarcoma. Approximately 5% of cases involve the head and neck with orbit being the site of preponderance.[2] The involvement of temporal bone is very uncommon. Schiffenbauer first reported an IgG4 related IPT of the mastoid, mimicking recurrent mastoiditis.[2] Common presenting symptoms are otalgia, hearing loss, otorrhoea and vertigo. The diagnostic protocol includes clinical history, examination, radiological tests and histopathology with immunohistochemistry. Standard treatment protocol includes surgical excision and adjunctive steroid therapy.[4]

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Case Report

A 61-year-old female presented with right otalgia, hearing loss, facial pain and imbalance. The complaints were

insidious in onset and gradually progressive over 2 years. She gave a history of ischaemic heart disease, hypertension and pulmonary tuberculosis treated 38 years back. Ear microscopy revealed a pulsatile mass in the right external auditory canal. Fistula and Romberg's tests were negative. Pure tone audiometry reported a right-sided profound sensorineural hearing loss (SNHL).

HRCT temporal bone reported soft tissue filling the right middle ear cavity with extension into mastoid air cells and erosion of the mastoid cortex. The ossicles were attenuated. MRI showed irregular intermediate signal intensity soft tissue, with erosion of tegmen tympani and hyperenhancement of the overlying dura. Post-contrast, the lesion was heterogeneously enhanced causing partial encasement of the right petrous internal carotid artery anteromedially.

The patient was planned for a right canal wall down tympanomastoidectomy. Intraoperatively, friable and vascular granulation tissue was noted in the middle ear and mastoid, eroding its lateral cortex and the tegmen plate. There was presence of a lateral semicircular canal fistula along with dehiscence of the horizontal and vertical segments of the facial nerve. The cochlea and the ossicles were eroded except for an intact stapes footplate. The lesion encased the internal carotid artery and jugular bulb. The intraoperative frozen section reported an inflammatory pseudotumor. Subtotal petrosectomy with wide conchomeatoplasty was done.

Histopathology reported a bone destructive Plasma cell predominant pseudotumor. Sheets of plasma cells with scattered Russell bodies were present. Areas of fibrosis with a storiform pattern were noted. Immuno-histochemistry revealed numerous plasma cells expressing IgG4 and IgG, their ratio being greater than 40%. Rheumatology experts advised PET-CT scan to rule out multisite involvement. On evaluation, it was concluded as an isolated IgG4 related right temporal bone disease. The patient was started on oral Prednisone 30mg/day, tapering over 3 months. One year follow-up showed a healthy mastoid cavity with no recurrence on PET-CT.

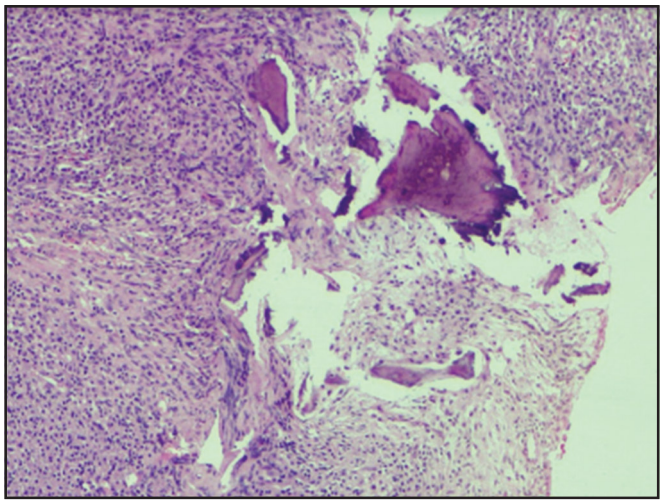

Fig. 1. Bone destructive Plasma cell predominant lesion with background fibrosis; stain: H&E; magnification 40X

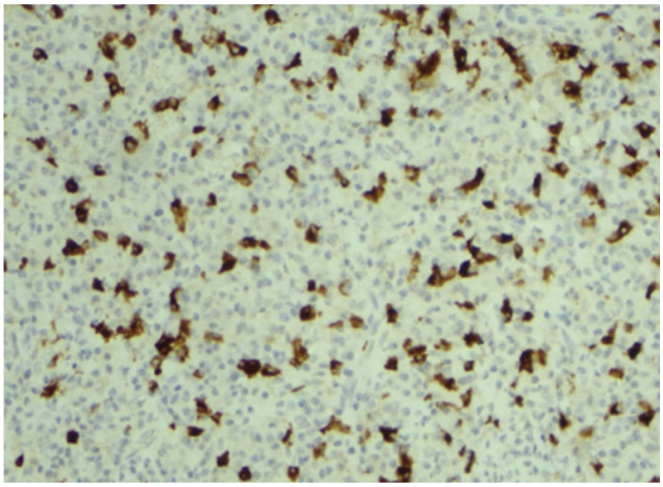

Fig. 2. IHC: Jones Methenamine Stain (JMS) : >50% IgG4 positive plasma cells 100x magnification

Discussion

Sclerosing inflammatory pseudotumor of the temporal bone is rare, usually involving a single ear.[4] It is locally destructive and recurrent in nature. The mastoid and middle ear are the most frequent sites involved. The clinical manifestations are mainly hearing loss, otalgia, otorrhea, facial palsy and vertigo. The lesion may also extend into the surrounding dura, sigmoid sinus, encase the internal carotid artery and/or the jugular vein.

Intratemporal extensions to the otic capsule, facial nerve, petrous apex, and internal auditory canal are also noted.[3]

The diagnosis is one of exclusion based on history, clinical examination, radiological imaging, with histopathology being the gold standard. HRCT Temporal bone shows an erosive soft tissue mass. MRI is more specific demonstrating iso or hypointensity on T1-weighted images and marked hypointensity on T2-weighted images, reflecting the fibrotic nature. Post-contrast the appearance is homogenous.[4] The HRCT temporal bone in our case suggested features likely of a cholesteatoma while the cMRI reported intermediate signal mass, not confirmatory of/ruling out cholesteatoma.

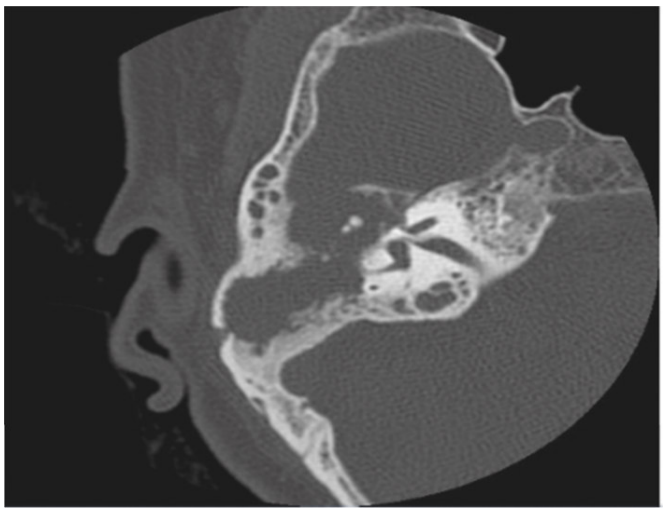

Fig. 3. HRCT Temporal bone, Axial cut : Erosion of lateral SSCC, lateral cortex of mastoid, dehiscent tympanic segment facial nerve, ill defined anterior wall middle ear.

Characteristic histopathological features noted are dense aggregates of fibroblastic and myofibroblastic cells admixed with chronic inflammatory cells. There is growing evidence that it represents a subtype of IgG4 related disease. There may be characteristic IgG4 upregulation with the histological presence of IgG4 related plasma cells, storiform fibrosis pattern and obliterative phlebitis.[4] However, it does not correlate with the disease severity. The diagnosis is further confirmed with Immunohistochemistry(IHC), lesion staining positive for kappa and lambda chains suggesting polyclonal nature. The histopathological findings of the reported case were consistent with the above.

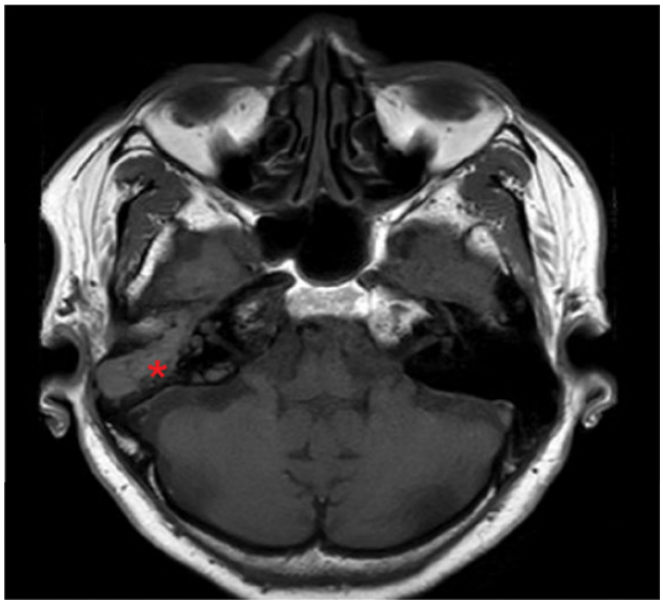

Fig. 4. T1W MRI : Intermediate intensity soft tissue signal in Right middle ear cleft.

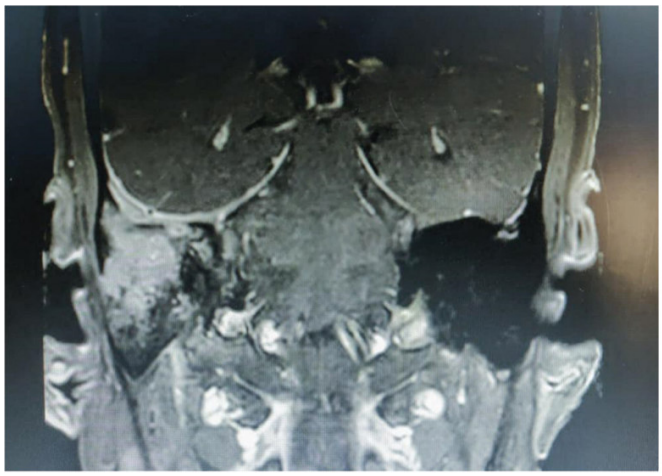

Fig. 5. cT1W MRI - Heterogeneous enhancement of right middle ear soft tissue with enhancement of overlying dura

Treatment for temporal bone pseudotumor can be summarized as subtotal petrosectomy, systemic corticosteroids, radiotherapy, immunomodulators, or any combination of the above depending on the extent and degree of invasion.[5] Subtotal petrosectomy, in combination

with corticosteroid therapy being the most common treatment of choice, was implemented.

A recurrence rate of 22% at 12 months following surgical resection has been reported.[5] Prolonged administration of steroids has been recommended in order to prevent further progression. Adjunctive therapeutic modalities in the form radiotherapy, monoclonal anti-CD20 antibody - Rituximab and the inosine mono-phosphate dehydrogenase inhibitor - Mycophenolate Mofetil (MMF) may play a role in recalcitrant cases and in chronic steroid intolerance.[5]

Conclusion

IgG4 related sclerosing pseudotumor of the temporal bone is a rare benign lesion characterized by its locally destructive nature and chronicity. It is often mistaken for neoplasm or cholesteatoma, hence should be considered as a differential diagnosis. Final diagnosis depends on a careful review of histopathology and immuno-histochemistry. A combination of surgical resection and prolonged corticosteroid therapy has been advocated as the mainstay of treatment. As the disease tends to run a chronic course with a tendency to recur, a long-term follow-up is necessary.

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