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Original Research Paper

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PLASMA CELL GRANULOMA OF MASTOID: A RARE CASE REPORT

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ABSTRACT

Plasma cell granuloma is a rare benign lesion which is found most frequently in the lungs, and rarely in the head and neck.[1] In this study, we present 22year old male with right sided otalgia, tinnitus, hearing loss and swelling over right postauricular region. On otoscopy, tympanic membrane was intact and dull and pure tone audiometry showed right mild conductive hearing loss. HRCT mastoid and MRI brain suggested CSOM with cholesteatoma. Intact canal wall mastoidectomy with debulking of tumour was done. HPE reported plasma cell granuloma (inflammatory pseudotumour) which was confirmed by Immunohistochemistry. Post operative period was uneventful. This case report is being highlighted for its rarity and unusual presentation.

KEYWORDS :

INTRODUCTION:

Plasma cell granuloma is an unencapsulated, benign tumor of unclear etiology. It is also referred to as *inflammatory pseudotumor inflammatory myofibroblastic pseudotumor, inflammatory histiocytoma,* and *inflammatory fibrosarcoma.*[1] The World Health organization defines it "a distinctive lesion composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils".[13] Extra-pulmonary sites are uncommon and head and neck accounts for only 5% of extra pulmonary lesions. The most common location in the head and neck is the orbit, followed by the meninges, paranasal sinuses, infratemporal fossa, and soft tissues.[5] The temporal bone, skull base, and facial nerve are very rarely involved.[2] The most common presenting symptoms are otalgia, hearing loss, and otorrhea. Clinical presentations vary from slow growth with minimal mass effect to bony destruction that can mimic malignancy.[12] Standard treatments include surgical excision, steroid therapy, and radiotherapy, depending on the size and location of the tumor, as well as the patient's age and comorbidities.[5]

In this article, we report a rare case of inflammatory pseudotumor of the mastoid.

Case Report:

A 22year male presented with right sided otalgia, tinnitus and swelling over right post auricular region for four months. He complained of mild decrease in hearing for 1month. it was associated dizziness and nausea. There was no history of ear discharge, vomiting and fever.

On otoscopic examination, patient had dull and intact tympanic membrane on right side and intact tympanic membrane on left. No perforation or discharge seen. Pure tone audiometry showed right sided mild conductive hearing loss.

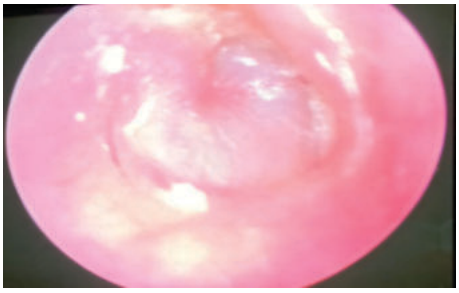

FIG 1: Otoendoscopy of the patient showing intact right tympanic membrane

HRCT mastoid suggested right chronic otomastoiditis with soft tissue density in right middle ear, aditus, common cavity formation and erosion of tegmen, sinus plate and long process of incus with possible intracranial extension.

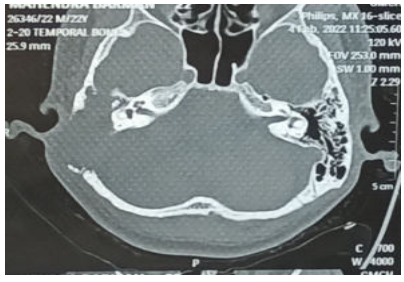

FIG 2: HRCT mastoid of the patient showing soft tissue density and gross bony erosion of right mastoid

To rule out intracranial extension, MRI brain was done. MRI revealed well defined lesion heterogenously hyperintense on T2 and FLAIR and iso to hypointense on T1 within right middle ear and mastoid air cells(4.1cm x 2.6cm x3.4cm) extending upto subcutaneous plane of right post auricular region and no intracranial extension.

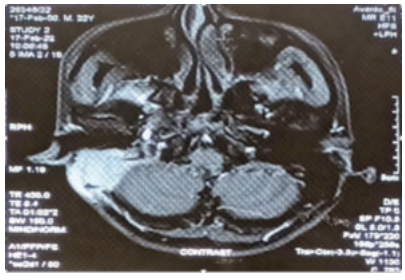

FIG 3: T2 MRI showing hyperintense lesion in right mastoid

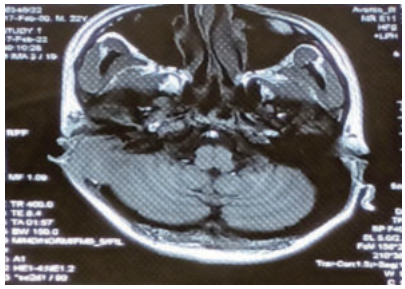

FIG 4: T1 MRI showing iso to hypointense lesion in right mastoid

After carrying out the necessary investigations, patient was prepared for intact canal wall mastoidectomy with debulking of mass under general anaesthesia. Intraoperatively, after making post auricular Wilde's incision and deepening it, lateral wall of mastoid was antrum was dehiscent. Mass was seen in antrum, attic and aditus. The tissue was friable and easily bleeds on touch.3 small sections of mass were excised and sent for frozen section. Intraoperative Frozen section report revealed focal areas of small round cells having hyperchromatic nuclei and scanty cytoplasm and spindle fibroblastic cells with areas of inflammation suggesting Small Round Cell tumour. Debulking of mass done and sent for HPE. sigmoid sinus plate was dehiscent and sinus was exposed. Tympanic membrane and ossicles were intact and mobile.Incision was closed in layers. Post operative period was uneventful.

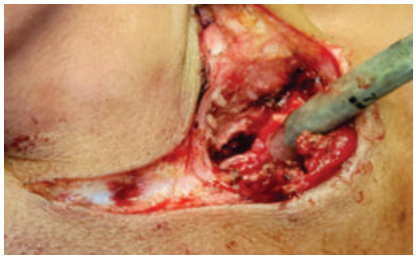

FIG 5: Intraoperative picture showing the mass eroding the mastoid

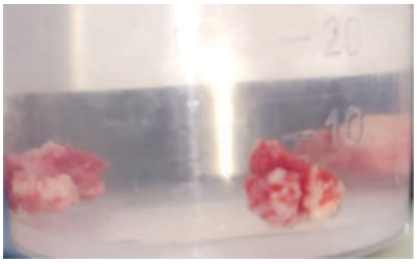

FIG 6: Tissue sent for intraoperative frozen section

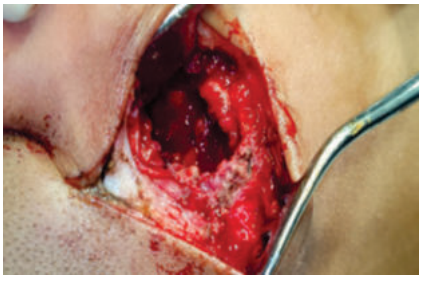

FIG 7: Intraoperative picture after debulking the mass

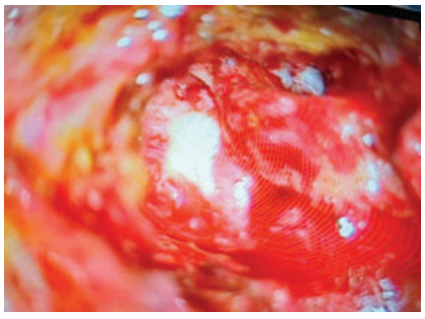

FIG 8: Intraoperative microscopic picture

HPE revealed fibroinflammatory lesion with variable infiltrates of lymphocytes and numerous plasma cells along with myofibroblasts in a fibrocollagenous background. scattered Russel bodies and foamy histiocytes were also seen suggesting diagnosis of Inflammatory pseudotumour (plasma cell granuloma). IHC was done to confirm the diagnosis. on IHC, plasma cells were highlighted by CD138 and MUM1. Both kappa and lambda were equally expressed on these plasma cells.

Prednisone at 60 mg/day followed by a taper was initiated for the treatment of the residual inflammatory pseudotumor. Patient was followed up at 1 week, 1 month The recovery was uneventful. Otoscopy revealed intact tympanic membrane.

DISCUSSION:

Plasma cell granuloma is a relatively rare lesion comprised of polyclonal plasma cells set in a background of fibrosis and spindle cell proliferation. While uncommon, this lesion may occur within any site and should be included in the list of differential diagnoses for plasma cell neoplasms. this entity can be mistaken for a plasma cell neoplasm. Although the etiology of these lesions is unclear, it is generally accepted that they are secondary to an unusual tissue response to injury[3] or may be secondary to infection[4]. Recent literature suggests that this falls within the spectrum of IgG4 related diseases.[1] Unlike in other organ systems where the manifestations of IgG4-related disease may be an incidental radiological finding, plasma cell granulomas in the head and neck most frequently present due to local destruction of the surrounding tissue.[10] Their ability to erode and infiltrate bone leads to easily being misclassified as malignant upon early assessment.

Inflammatory pseudotumor usually demonstrate unusual clinical, radiographic, and pathologic features.[5] While our patient's complaints of otalgia, hearing loss, and no otorrhea might suggest congenital cholesteatoma but not chronic otitis media. He had radiographic findings suggestive of a neoplasm. However, otoscopy revealed intact tympanic membrane and no visible cholesteatoma, retraction pocket or perforation. Audiometry showed mild conductive hearing loss, as expected with. Congenital cholesteatoma or chronic otitis media. Our provisional diagnosis was congenital cholesteatoma. however, the focal area of bone destruction noted laterally through the mastoid cortex was atypical for cholesteatoma. Instead both CT and MRI suggested chronic otitis media and cholesteatoma.

Temporal bone inflammatory pseudotumor have been successfully treated with surgical excision, and high-dose steroids, which we used in our patient. Radiotherapy has been used to treat (1) inflammatory pseudotumor in anatomically inaccessible regions such as the skull base, (2)recurrent or refractory disease, (3)tumors that exhibit a poor response to steroid therapy, and (4)patients who cannot tolerate other treatments, although it is associated with radiation-induced neoplasms.[6,7,8,9] Our patient's relatively young age and otherwise good health supported our decision to encourage steroid therapy rather than radiotherapy after surgical excision.

Because these tumours are unencapsulated, surgical excision with wide margins is necessary to prevent recurrence. A recurrence rate of 22% at 12 months following surgical excision has been reported.[11]

CONCLUSION:

Inflammatory pseudotumor rarely occurs in the temporal bone. Characteristic clinical and radiographic findings distinguish this condition from mastoiditis, chronic otitis media, and cholesteatoma. An accurate diagnosis depends on a careful review of CT and MRI findings and histopathologic analysis.

These pseudotumors are locally destructive, and they have the capacity to erode through bone and invade soft-tissue structures, so they must be treated aggressively. Total surgical excision should be recommended as the initial treatment, with

adjunctive therapy such as steroids and radiotherapy reserved for residual, recurrent, or inaccessible tumors.[3]

Consent:

Written and informed consent was obtained from the patient regarding the use of his clinical findings and reports of the investigations that were conducted.

Disclosure Statement:

No potential conflict of interest was reported by the authors.

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