Inflammatory Pseudotumor of the Inner Ear
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ABSTRACT

Objective: To present a report of inflammatory pseudotumor of the inner ear in a child, discuss radiographic findings, clinical management, and review the current literature on this rare disease.

Design: Case Report

Results: A 2 1/2 year old male with presumed chronic right otitis media presented with otalgia, transient vertigo, and fluctuating facial palsy partially responsive to myringotomy with tube. Work-up for infectious and neoplastic processes was negative. Magnetic resonance imaging (MRI) of the temporal bone revealed diffuse enhancement of the middle ear, mastoid, inner ear and petrous apex with an unusual expansile, erosive appearance to the otic capsule on CT. The patient’s symptoms were transiently responsive to tympanomastoidectomy and eventual inner ear with histopathology after labyrinthectomy revealed changes consistent with pseudotumor.

Conclusions: Inflammatory pseudotumor of the inner ear is extremely rare but should be considered in cases of refractory chronic otitis media with facial palsy particularly when certain changes appear on CT of the temporal bone.

CASE PRESENTATION

An otherwise healthy 2 1/2 year old male, with a history of bilateral myringotomy at 6 months for recurrent otitis media, presented with chronic otitis media and new onset of otalgia and fluctuating facial palsy. He acutely underwent myringotomy and tube placement, and antibiotic therapy was initiated, with resolution of facial palsy and his symptoms. Cultures from the middle ear were negative. His facial palsy recurred within a week. CT was obtained and revealed soft tissue density in the middle ear with a very unusual centrally expanded appearance of otic capsule. The basal turn of the cochlea was normal, but the remainder was dilated, cystic and dehiscent into the middle ear. The posterior semicircular canal was dehiscent into the middle cranial fossa and the lateral canal was enlarged and cystic. The muscles, scutum, and tegmen were intact, and the fallopian canal was enlarged. Audiology revealed profound sensorineural hearing loss on the right despite normal newborn screening, and previously present bilateral distortion product otoacoustic emissions. He underwent tympanomastoidectomy and abundant rubbery, inflamed tissue was found within the mastoid. The lateral semicircular canal was markedly abnormal as described and the facial nerve was clinically dehiscent but structurally intact. Pathology was consistent with chronic inflammation and infection. His facial palsy was resolved postoperatively but fluctuated over the next 3 weeks (at worst HB 3). He was continued on oral and ototopical antibiotics, but on clinical followup granulation tissue developed on the tympanic membrane and a polyp was noted in the external canal. This prompted revision tympanomastoidectomy. The same abnormal tissue was abundant. Cultures were weakly positive for Pseudomonas aeruginosa which led to a diagnosis of possible skullbase osteomyelitis. Directed antibiotic therapy with Cefpime was initiated, and a technetium bone scan was obtained. This revealed mild uptake in the region of the right mastoid. MR imaging was consistent with the CT and showed soft tissue density in the mastoid, semicircular canals, and tympanic segment of the facial nerve on the right. Given the appearance of the tissue there remained a suspicion for tumor. The mastoid was resected and labyrinthectomy performed for tissue diagnosis. The otic capsule was partially eroded with thin, fragile bone and leathery tissue occupying the vestibule. Given his facial paresis, attention was directed to the facial nerve which did not appear to be encased or invaded with tumor, though the tympanic segment was impinged by tumor-like mass. Pathology revealed fibroconnective tissue with abundant plasma cells and lymphocytes. Immunohistochmcal (s-100, CD 1A, CD 68, Vimentin, Desmin) results were nonspecific, showing no evidence of tumor or histiocytosis. The final diagnosis was inflammatory pseudotumor.

He was subsequently treated with a short course of steroids, and is currently 1 year status post labyrinthectomy with complete resolution of the facial palsy. Repeat imaging 4 months after labyrinthectomy revealed no change or growth of the soft tissue lesion.

CONCLUSION

Inflammatory pseudotumor of the inner ear is rare but should be considered in cases of refractory chronic otitis media with complications such as facial palsy, particularly when characteristic changes appear on imaging.

REFERENCES