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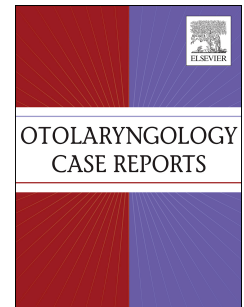
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Title: A case of allergic fungal otomastoiditis with aural polyps

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Abstract: While common in the paranasal sinuses, the presence of allergic mucin within the middle ear is rare. Here we describe a case of what has been termed allergic fungal otomastoiditis (AFOM), an unusual immunologic response to the presence of fungal hyphae within the middle ear, with a unique presentation of aural polyps. A 25-year-old woman presented with recurrent right-sided aural polyps and chronic otorrhea, despite two previous tympanomastoidectomy surgeries performed in India. CT scan of the temporal bones demonstrated complete opacification of the tympanic cavity and mastoid. Canal wall down tympanomastoidectomy was performed, revealing thick otorrhea and polyps filling the external auditory canal and middle ear, eroding the entire tympanic membrane and obstructing the antrum. Pathology revealed glandular and squamous metaplasia, marked inflammation with eosinophils, as well as allergic-type mucin with non-invasive fungal hyphae highlighted by GMS stain. The histopathologic findings of AFOM are reminiscent of allergic fungal rhinosinusitis (AFRS) and suggest a similar pathophysiology. Optimal treatment in these cases may require a combination of surgical debridement and medical management. AFOM should be considered in the differential diagnosis of chronic otitis media, particularly in the presence of aural polyps and thick otorrhea.

Keywords: allergic fungal, otomastoiditis, mucin, aural polyps, middle ear

INTRODUCTION:

Allergic mucin is commonly found in the paranasal sinuses as allergic fungal rhinosinusitis (AFRS); however, the presence of allergic mucin within the middle ear is rare.¹ Allergic fungal otomastoiditis (AFOM) was first described by Chen and Chiang in 2013 in a 65-year-old patient with chronic otorrhea who was found to have thick allergic mucin with fungal elements in the middle ear [1]. Since then, only one additional case of AFOM has been described in a child, but no mention of aural polyps was made in either of these two cases [2]. Here we present a unique case of AFOM with aural polyps, and in doing so, hope to give greater awareness to this entity and its spectrum of disease.

CASE PRESENTATION:

A 25-year-old woman who recently moved from India presented to our office with chronic right-sided otorrhea and hearing loss despite two previous tympanomastoidectomies at ages 19 and 23 for presumed cholesteatoma. She had no significant medical history other than allergic rhinitis. On exam, she was noted to have thick, purulent otorrhea and polyps filling the right ear canal. She did not have any facial weakness or vertigo. An audiogram was obtained showing profound right-sided sensorineural hearing loss of unclear etiology. CT scan of the temporal bones revealed complete opacification of the right external auditory canal, tympanic cavity, and mastoid, with erosion of the ossicles and facial canal but an intact tegmen and inner ear (Figure 1).

Canal wall down tympanomastoidectomy was then performed. Intra-operatively, polyps were found to be filling the external auditory canal, eroding the tympanic membrane, and filling the entire middle ear into the antrum. The mastoid was filled with thick pus, but no cholesteatoma was seen. The malleus and incus were partially eroded and wrapped with polyps. The polypoid disease was removed and sent to pathology, which showed squamous and glandular metaplasia with marked infiltration of eosinophils. Pathology also demonstrated “allergic-type” mucin with fungal hyphae highlighted by GMS stain, without evidence of tissue invasion (Figure 2).

Post-operatively, she was treated with two weeks of oral prednisone, along with several courses of topical corticosteroids. She had a good initial response with cessation of drainage and no recurrence of aural polyps. However, about one year after surgery, she began to develop recurrent aural polyposis along with thick otorrhea. *In vitro* allergy testing was also performed, which showed positivity to multiple molds, including *Aspergillus fumigatus*. Interestingly, CT scan of her sinuses showed no significant sinus disease. Her disease has remained stable over the last four years with regular in-office debridements and multiple courses of topical steroids.

DISCUSSION:

Since AFOM shares an identical histologic appearance to AFRS, we can infer that it shares a similar pathophysiology. Fungal antigens are thought to stimulate a type I and/or type III immunologic response, with eosinophilia and inflammatory mediators leading to the production of allergic mucin. Allergic mucin is recognized grossly as thick, dark mucus with “peanut butter” consistency. Histopathology shows eosinophils in a background of mucin and scattered fungal hyphae, along with occasional Charcot-Leyden crystals. Local inflammation of the mucosa leads to outflow tract obstruction, creating stasis and a continued immunologic response. Over time, this leads to expansion and remodeling of the surrounding bone. Because fungus is ubiquitous in the nose and sinuses, it is believed that there are other environmental or genetic factors that predispose one towards developing this type of immunologic response.

The diagnostic criteria for AFRS were described by Bent and Kuhn in 1994, and these are: 1) type 1 hypersensitivity by history, 2) positive *in vitro* or skin testing for fungal antigens, 3) polyposis, 4) eosinophilic mucus without fungal invasion into tissue, 5) a positive fungal stain, and 6) characteristic CT scan findings with heterogeneous soft tissue density and bony expansion or erosion [3]. Our patient fits all of these criteria (with aural polyps instead of nasal polyps), though on her CT scan it was difficult to

¹ Abbreviations: AFRS (allergic fungal rhinosinusitis), AFOM (allergic fungal otomastoiditis)

appreciate any bony changes or soft tissue heterogeneity due to her previous mastoidectomies and remnant ossicles. These signs may be helpful in distinguishing AFOM from cholesteatoma, which typically does not have heterogeneous signal intensity and erodes rather than expands. The presence of polyps and thick otorrhea may also suggest AFOM instead of cholesteatoma.

Our patient's presentation differed from that of the patients described by Chen and Chiang (2013) and Bayer *et al.* (2014) in that she had aural polyps, analogous to the nasal polyps almost universally seen with AFRS [1,2]. There were also two reports of patients with aural polyps and eosinophilic mucin within the middle ear, but no fungal elements were seen in these cases [4,5]. It is possible that fungal elements were simply too scarce or missed in these cases. Regardless, there appears to be a spectrum of disease, ranging from pediatric to adult and from those with aural polyps to those without.

Surgical extirpation of the disease is considered to be the primary treatment for AFRS, with the goal of removing the triggering antigen and improving ventilation and post-operative access for debridement. In the ear, this may require a canal wall down tympanomastoidectomy. As is often the case in AFRS, our patient had recurrence, with return of aural polyps and thick otorrhea about one year after surgery. Topical steroids and periodic debridements appear to have stabilized the disease for the last few years. There is some evidence that anti-IgE therapy may be helpful, but it is unclear whether or not immunotherapy and anti-fungal agents are effective in managing AFOM [2].

CONCLUSION:

AFOM is a rare disease entity, but one that should be considered in the differential diagnosis of chronic otitis media, particularly in the presence of aural polyps and thick otorrhea. A similar histopathology and pathophysiology to AFRS may inform possible treatment options, though there may be a high rate of recurrent disease in certain cases. More cases are necessary in order to better understand this unusual disease process and its optimal management.

Consent: Not applicable; no identifying information or images were included in the case report.

Conflict of Interest Statement: The authors disclose no conflict of interest.

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FIGURE LEGENDS

Figure 1: Pre-operative non-contrast CT of the temporal bones. Axial views at the level of the horizontal canal (A) and the cochlea (B) and a coronal view of the right temporal bone (C) demonstrate complete opacification of the canal wall up mastoidectomy defect, the osseous external auditory canal, and the tympanic cavity with soft tissue density material.

Figure 2: Histopathology from intra-operative biopsy of polyps within the middle ear. **A.** The specimen consisted of polypoid fragments of middle ear mucosa with edema, chronic inflammation, glandular metaplasia, and free-floating acellular debris (top). **B.** In areas, the metaplastic glands were markedly dilated and contained eosinophilic, allergic-type mucin. **C.** The allergic mucin contained numerous intact and degranulated eosinophils along with cellular debris and subtle, hyaline fungal hyphae (arrow). **D.** A special stain for fungi (Gomori methenamine silver) highlighted the non-invasive fungal hyphae within the allergic mucin.

