Bilateral Radical Mastoidectomies as a Treatment for Recalcitrant Otorrhea due to Kartagener Syndrome

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ABSTRACT

Educational Objective: At the conclusion of this presentation, the participants should be able to understand the usage of a radical mastoidectomy to manage persistent otorrhea due to Kartagener syndrome.

Objectives: To describe a case of bilateral otorrhea due to Kartagener syndrome which resolved with radical mastoidectomies.

Study Design: Case report.

Methods: A retrospective review of the medical record of a patient with Kartagener syndrome and otorrhea was performed.

Case Report: A 27 year-old female with Kartagener syndrome presented with years of bilateral otorrhea. She had previously undergone bilateral canal wall down mastoidectomies and had tubes in both tympanic membranes. She had chronic bilateral otorrhea unresponsive to prolonged therapy with topical, oral or intravenous antibiotics. She underwent staged, bilateral radical mastoidectomies, removal of disease mucosa, and skin grafting of the cavities. Her otorrhea has resolved to great patient satisfaction.

Conclusions: In cases of recalcitrant otorrhea in patients with Kartagener syndrome, radical mastoidectomy with removal of diseased mucosa can result in dry cavities free of otorrhea.

INTRODUCTION

Kartagener syndrome is a variant of primary ciliary dyskinesia (PCD) associated with situs inversus, chronic sinusitis, and bronchiectasis. PCD is a rare, genetically mediated dysfunction of ciliary motion that affects 1 in 15,000 individuals. In the United States alone, 15,000-17,000 individuals may have PCD and roughly 50% of those affected have situs inversus. The disorder is transmitted via an autosomal recessive pathway with variable penetrance, and is heterogeneous with multiple mutations causing ciliary dysmotility.

Common head and neck manifestations of PCD include chronic rhinosinusitis and otitis media with effusion (OME). Treatment of OME in patients with PCD frequently includes placement of tympanostomy tubes, which have an increased rate of tube otorrhea in the patient population. Due to this increased rate of tube otorrhea, some authors have recommended against the placement of tubes in patients with PCD. Another study has demonstrated tympanic membrane perforation and calcification of otorrhea in 50% of patients with PCD treated with repeated tympanostomy tube insertion. There is scant literature beyond ventilation tubes and potentially tympanoplasty in cases of tympanic membrane perforation to treat unremitting otorrhea.

We present the case of a 27 year-old female with Kartagener syndrome and bilateral, chronic otorrhea recalcitrant to previous medical and surgical therapy.

CASE REPORT

A 27 year-old female with a history of Kartagener syndrome presented to the otolaryngology clinic with a complaint of bilateral, unremitting otorrhea. She had a classic presentation of Kartagener syndrome with chronic rhinosinusitis, situs inversus (Figure 1), and bronchiectasis. Primary testing for a ciliary abnormality was done at another institution, and results were not available. She noted a history of hearing loss since she was 6 years old, and a history of multiple bilateral ear operations for otorrhea including tympanoplasties for perforated tympanic membranes, and bilateral canal wall down mastoidectomies. She had undergone multiple courses of topical, oral, and intravenous antibiotics without resolution of her otorrhea. She had recently begun the use of hearing aids bilaterally, which were poorly tolerated due to otorrhea.

The patient underwent bilateral revision radical mastoidectomies performed 7 months apart. Her right ear operation was performed initially. The inflamed tympanic membrane, malleus, incus and middle ear mucosa were resected. Her eustachian tube orifice was obliterated with temporalis muscle and bone pâte. Her stapes superstructure was intact, and the mucosa was carefully removed from this structure using a laser. A small piece of tragal cartilage was placed over the stapes superstructure. A temporals fascia graft and split thickness skin graft were used to cover the promontory, obliterated eustachian tube orifice, and cartilage over the stapes.

An audiogram performed 5 months after the operation showed no significant change in hearing. She reported no otorrhea from the operated ear in the 7 months prior to her contralateral left ear operation. A similar left ear operation was performed without complication, and the meatoplasty was revised to allow for better cavity visualization and to prevent debris accumulation.

At last follow-up, eleven and four months after her right and left ear operations respectively, she had been free of otorrhea for the first time in years. She was very pleased with this result. She has been able to continue use of her behind-the-ear hearing aids and has not elected to proceed with surgical implantation of osteointegrated hearing aids.

DISCUSSION

Chronic suppurative otitis media, associated with otorrhea, is known to have a detrimental effect on quality of life. Chronic drainage can lead to embarrassment, withdrawal from social events, absenteeism at work, and the need for recurrent treatment with antibiotics. Using validated questionnaires, it has been shown that surgical treatment of COM can improve disease specific quality of life, and that cessation of otorrhea is a large component of this improvement.

Most of the published literature regarding otologic disease associated with PCD is focused on pediatric patients. There is controversy regarding the optimal management of COM in children with PCD due to the risk of persistent tympanic membrane perforation or chronic, unremitting tube otorrhea. Little data exists regarding the management of patients with tube otorrhea in PCD. One review of 9 children with PCD and a tympanic perforation demonstrated a tympanoplasty success rate of 100%, but a majority of patients developed recurrent COM. This indicates tympanoplasty may be likely to abate otorrhea in most cases.

This case presented a more difficult clinical scenario in which an adult had already undergone extensive surgical manipulation of both ears. She presented after failed tympanoplasties and canal wall down mastoidectomies. Perhaps revision tympanoplasty, potentially utilizing cartilage, may have been an effective treatment of otorrhea. However, given her canal wall down mastoidectomy cavities and history of tympanoplasty failure, it was felt a radical cavity with skin grafting of the middle ear would be more likely to resolve her otorrhea in a single procedure. In the setting of PCD with recurrent otorrhea and COM, a significant hearing improvement in this patient was an unlikely outcome. With dry cavities, she was able to become a more reliable hearing aid user. Osteointegrated hearing aids are a reasonable option in such a patient, but she elected to forego this treatment.

CONCLUSION

In selected patients with unremitting otorrhea due to PCD, radical mastoidectomy with resection of the diseased middle ear mucosa is an effective treatment. Tympanoplasty can also be a successful treatment modality, but in a patient with a history of chronic otorrhea, tube placement should likely be avoided after the graft has healed.

REFERENCES


Figure 1: A posterior-anterior chest radiograph demonstrates dextrocardia, consistent with situs inversus.

Figure 2: Audiometric data.

Figure 2: Axial CT demonstrates bilateral canal wall down mastoidectomy defects. The middle ear spaces are opacified bilaterally, consistent with fluid.