ABSTRACT

Title
Bilateral second branchial cleft fistula in an Adult

Educational Objectives
The observer should be able to recognize the anatomy of the second branchial cleft and its relation to clinical presentation as well as to understand work up, treatment and possible genetic basis.

Study Design
Case report.

Methods
Patient chart review and review of the literature.

Results
A 72-year-old Caucasian female presented with a one-year history of a right neck mass. She underwent work up with imaging and FNA to exclude malignancy. She underwent excision of right neck mass that revealed fistula tract from the right neck to ipsilateral tonsillar fossa. During resection the tract demonstrated characteristic course overlying the glossopharyngeal and hypoglossal nerves. The fistula was lined by squamous and respiratory mucosa in keeping with reported histology of fistulae (6,7).

Conclusions
Though exceedingly rare, bilateral second branchial cleft fistulae may present in elderly individuals. Bilateral presentations appear to have a familial component, which should arouse clinical suspicion for underlying genetic disorders such as branchio-oto-renal syndrome (BOR).

INTRODUCTION

Case: 72 year old female non-smoker presented for right neck mass of one year duration. Exam concerning for metastatic disease prompting PET scan showing increased uptake in right tonsil. Direct laryngoscopy and pharyngoscopy failed to show any evidence of malignancy but did reveal active expressible purulence from right pharynx inferior to tonsil (Figure 3). Definitive surgery was performed transcervically and transorally with fistula tract excised in continuity with preservation of all vital structures (Figure 4). Findings at time of surgery were consistent with second branchial cleft fistula. The patient does have a history of remotely excised left neck lesion and a family history of similar neck lesions that raises the suspicion of a genetic basis in the form of a branchio-oto-renal syndrome spectrum of disease. Pre-operative contrasted CT was useful in characterization and histologic findings of both respiratory and squamous mucosa were confirmatory of branchial fistula. Recurrence rates are reported at 8.6 to 22% in the literature. Our patient had no complications and is has no evidence of recurrence at eight months.

METHODS AND MATERIALS

Retrospective chart review and case report. Surgical, radiologic and histologic reports reviewed.

Reviewed English language peer-reviewed journals for background, diagnosis and various surgical techniques used to address lesions of second branchial cleft using PubMed.

RESULTS

Surgical pathology (Figures 1,2)
Branchial cleft cyst and fistula tract with marked acute and chronic inflammation and abscess. No dysplasia or carcinoma identified.

Fistula tract lined by squamous and respiratory mucosa with abundant acute and chronic inflammation and lymphocytic aggregates.

Soft tissue, intraoral fistula tract, excision: Squamous mucosa and fibrovascular tissue with chronic inflammation.

DISCUSSION

Second branchial cleft anomalies account for 90% of developmental abnormalities of the branchial apparatus (8). Fistulae normally present by 5 years of age and cysts or sinuses between the second and fourth decades and occur when both a branchial cleft and a pouch fail to obliterate (5). Second branchial fistulae (SBF) follow a typical course superficial to both the glossopharyngeal and hypoglossal nerves, under the posterior belly of the digastric and sometimes, but not always, coursing through the carotid bifurcation (1,5,7,8). The typical site for the distal eruption of the fistula is at the lower one third of the anterior border of the sternocleidomastoid. The proximal, or mucosal, aperture is usually located at the level of the palatine tonsil. Several studies have evaluated the role of pre-operative imaging highlighting that CT, MRI and fistulogram combined with CT can all effectively outline the course of a SBF (6) with the latter providing highest sensitivity if a distal opening is able to be cannulated.

In our patient, the presence of bilateral SBF does raise the suspicion of branchio-oto-renal syndrome spectrum of disease. BOR is an autosomal dominant disease with hallmark signs of hearing loss, preauricular pits, second branchial cleft anomalies, and renal issues. Interestingly, our patient does have mild to moderate mixed hearing loss bilaterally but no renal issues making branchio-oto-syndrome a possibility for which genetic testing could be confirmatory. In any case of SBF the treatment is surgical with an emphasis on careful dissection to at least the level of the hyoid bone prior to ligation and amputation. In the pediatric population concomitant tonsillectomy has been evaluated and shown to not improve recurrence rates (4).

CONCLUSIONS

Neck mass in an adult immediately raises suspicion for malignancy. This is an unusual case presentation of complete branchial fistula in an adult with a possible underlying genetic disorder. Pre-operative imaging is a useful adjunct in surgical planning with aberrations noted in the literature for the course of fistula tract for which the surgeon should be wary. The mainstay of treatment remains meticulous surgical dissection with preservation of vital structures.

REFERENCES