Factors Influencing Surgical Intervention in Adult Cystic Fibrosis Patients

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ABSTRACT

Purpose: To identify characteristics of adult cystic fibrosis patients that predict surgical intervention with endoscopic sinus surgery (ESS).

Methods: Patients were identified in a tertiary sinus center by ICD-9 code 277.00-277.03. Charts were reviewed for cystic fibrosis gene mutation, Lund-Mackay Score (LMS), SNOT22 score, number of previous ESS, and need for ESS after presentation. Fisher's exact test was used to compare the need for ESS based on presence or absence of previous ESS. A t-test was used to compare mean LMS and SNOT22 between the groups of previous ESS or no previous ESS. Analysis of variance was used to compare mean LMS, SNOT22, and need for ESS, between mutation groups, homozygous for delF508 mutation, heterozygous for delF508, or other mutations.

Results: 100 patients met the inclusion criteria for the study. 43/100 of patients had undergone previous surgery, 29/100 underwent subsequent surgery after initial presentation to the sinus center (of those 11 had not undergone surgery previously), and 46/100 never underwent surgery. Patients that had undergone previous ESS were more likely than patients that had not to require an additional ESS after presentation (p=<0.01), even though LMS was not significantly different between the groups of previous ESS and no previous ESS (9.6 and 11.3, p=0.11), and SNOT22 score was not significantly different between the two groups (37.5 vs 31.8, p=0.22). Status of the F508 deletion did not impact Lund-Mackay Score for homozygotes, heterozygotes and other mutations (10.8, 9.6, 8.7 respectively, p=0.45). Delta F508 status did predict the need for further ESS after presentation with 25% of homozygotes, 3% of heterozygotes and 20% of other mutations requiring ESS (p=0.04).

Conclusion: Surgical intervention in adult cystic fibrosis patients is predicted by previous ESS, and status of F508 deletion, but not LMS or SNOT22 score.

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INTRODUCTION

Chronic rhinosinusitis (CRS) is a common medical condition that is reported to occur in up to 15% of the general population and accounts for significant healthcare burdens and decreased quality of life. The Sinonasal Outcome Test -22 is an outcome measure that correlates with quality of life, and has been validated as a patient reported outcome measure for CRS.

Cystic fibrosis (CF) is an uncommon disease with a high prevalence of CRS due to viscous mucous and stasis leading to bacterial overgrowth. Previous authors have reported on the use of validated questionnaires to diagnose CRS in the CF population.

In the current study the authors sought to determine the impact of genetic mutation, Lund Mackay Score (LMS), SNOT22 score, and previous endoscopic sinus surgery (ESS) on the likelihood of needing further ESS.

METHODS AND MATERIALS

A retrospective chart review was performed after IRB approval (Massachusetts Eye and Ear protocol 778969) at a tertiary care institution. Patients were included in the study if they presented to the institution's Sinus Center, were treated by the senior author (STG) between January 2001 and January 2015, and carried an ICD-9 code of 277.00-277.03 (Cystic Fibrosis). Patients were excluded if the chart did not include a diagnosis of CF. Charts were reviewed for CF gene mutation, SNOT22 score, LMS, number of previous ESS, and need for ESS after presentation.

Fisher's exact test was used to compare the need for ESS based on presence or absence of previous ESS. A t-test was used to compare mean LMS and SNOT22 between the groups of previous ESS or no previous ESS. Analysis of variance was used to compare mean LMS, SNOT22, and need for ESS, between mutation groups, homozygous for delta F508 mutation, heterozygous for delta F508, or other mutations.

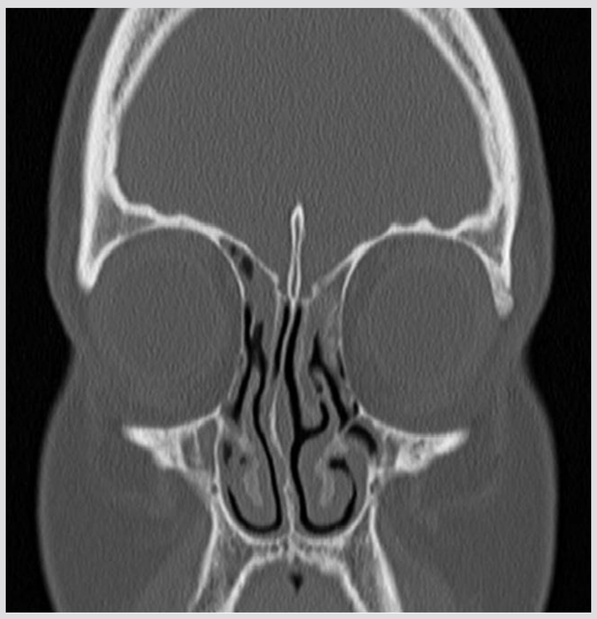

Figure 1. Coronal CT scan or the sinuses in a patient with CF

RESULTS

100 patients met the inclusion criteria for the study. 43/100 of patients had undergone ESS prior to presentation at our institution, while 57/100 had not undergone previous ESS. 29/100 underwent subsequent ESS after initial presentation to the Sinus Center, and of those 11 had not undergone ESS previously at another institution. 46/100 patients never underwent ESS at any institution. Patients that had undergone previous ESS were more likely than patients that had not to require ESS after presentation (p=<0.01), even though LMS was not significantly different between the groups of previous ESS and no previous ESS (9.6 and 11.3, p=0.11), and SNOT22 score was not significantly different between the two groups (37.5 vs 31.8, p=0.22) (See Figure3).

Mutation status was retrospectively identified from review of the record in 80 patients. Using those patients, mutation status did not impact LMS for homozygotes, heterozygotes and other mutations (10.8, 9.6, 8.7 respectively, p=0.45) or SNOT22 score (34, 33.5, 37 respectively p=0.90). Delta F508 status did predict the need for surgery after presentation with 25% of homozygotes, 3% of heterozygotes and 20% of other mutation combinations requiring ESS (p=0.04) (See Figure 4).

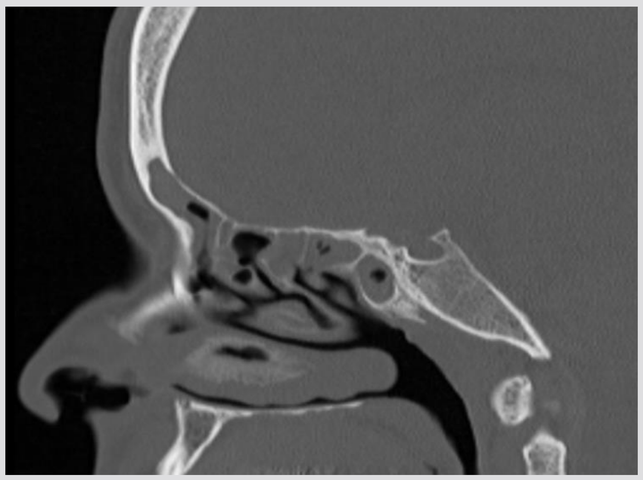

Figure 2. Sagittal CT scan or the sinuses in a patient with CF

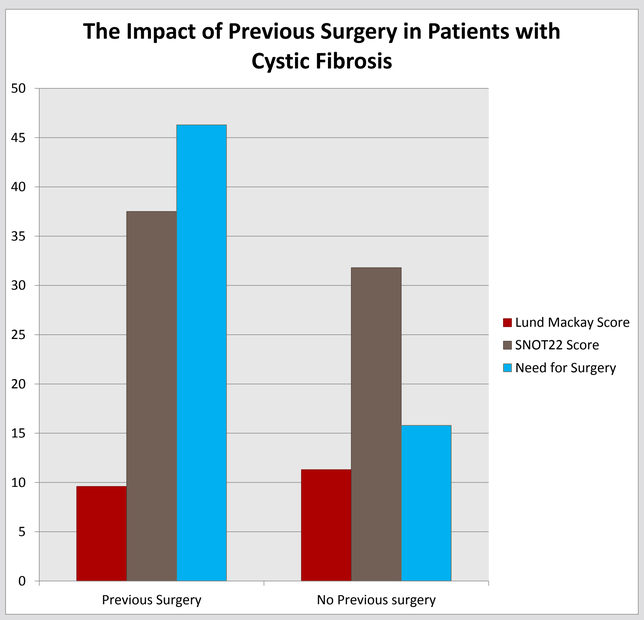

Figure 3. Characteristics of patients that had had previous ESS versus patients that had not undergone previous ESS. The need for ESS was statistically higher in patients who had undergone previous surgery (46.3 vs 15.8%, p=<0.01)

DISCUSSION

CF is an uncommon disease that often presents to the otolaryngologist because of the high rate of CRS in this population. Management of this disease can be difficult due to the underlying disease process and these patients sometimes require surgery for their disease.

Previous work has suggested that more severe mutations, including the most common mutation, delta F508, can lead to more phenotypically severe CRS, with higher LMS, and decreased aeration of the frontal and sphenoid sinuses. The current study suggests that a history of previous surgery and the presence of a delta F508 mutation may predict the need for future or further surgery.

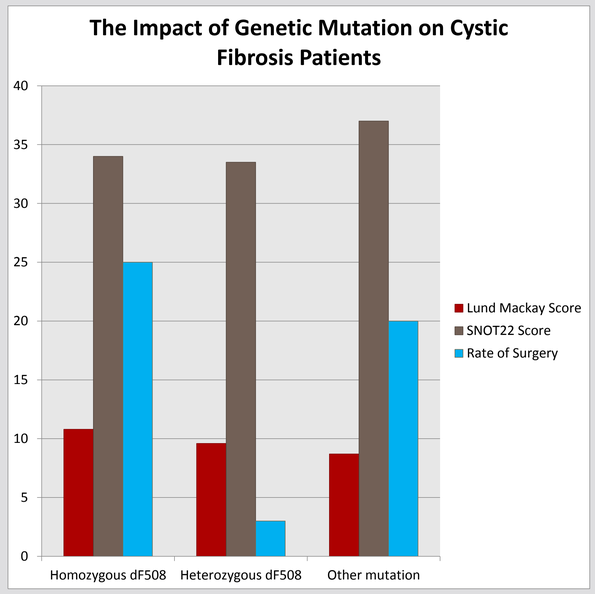

Figure 4. Characteristics of CF patients based on delta F508 mutation status grouped by homozygous, heterozygous and other mutations. The rate of surgery was statistically lower in patients who were heterozygous for the mutation (25 vs 3 vs 20% respectively p=0.04)

CONCLUSIONS

Surgical intervention in adult CF patients is predicted by previous surgical intervention, and status of the delta F508 mutation, but not LMS or SNOT22 score.

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