Case Report

Central-part laryngectomy after laryngotracheal separation to manage pharyngocutaneous fistula: A case report and retrospective analysis of 12 cases

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ABSTRACT

A 15-year-old girl presented with a 3-year-history of continuous outflow of saliva from a pharyngocutaneous fistula, located at 5 mm superior to her tracheal stoma. She was diagnosed with Miller-Dieker syndrome at birth. At 2 years of age, pediatric surgeons at our institution carried out laryngotracheal separation to prevent aspiration pneumonia. At the age of 12 years, she developed continuous saliva discharge from the fistula. We performed central-part laryngectomy and resection of the pharyngocutaneous fistula, which relieved her from the continuous saliva discharge. Central-part laryngectomy is less invasive and easier to perform than total laryngectomy. We hereby present a case and retrospective analysis of 12 patients, who underwent central-part laryngectomy.

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1. Introduction

Patients with chronic conditions such as congenital disease, neuromuscular disease, and cerebral palsy are more likely to experience intractable aspiration. Total laryngectomy, glottic closure, and laryngotracheal separation are surgical procedures to control aspiration [1–3]. Central-part laryngectomy (CPL) is less invasive as compared with conventional total laryngectomy [4]. Herein, we present the case of a patient with history of laryngotracheal separation, who underwent CPL and resection of pharyngocutaneous fistula to control aspiration.

2. Case presentation

A 15-year-old girl who was diagnosed with Miller-Dieker syndrome (MDS) at birth presented to our institution with a 3-year-history of saliva discharge from a pharyngocutaneous fistula. Miller-Dieker syndrome is a contiguous gene deletion syndrome characterized by lissencephaly, psychomotor retardation, and seizures [5]. The patient had aspiration pneumo-
nia in the neonatal period, and pediatric surgeons performed gastrostomy and laryngotracheal separation at ages 10 months and 2 years. The pediatric surgeons used an Endo-GIA stapler to separate the patient’s trachea and larynx. Nine years after laryngotracheal separation, when the patient was 11 years of age, she underwent a laryngeal foreign body extraction under general anesthesia. The foreign body consisted of a tooth, which naturally dislodged and obstructed the larynx without the guardians’ notice. The patient’s pharyngocutaneous fistula was already identified at the time of the foreign body removal procedure. At the age of 13 years, aspiration pneumonia recurred, requiring her parents to change gauze dressing on the pharyngocutaneous fistula every 30 min to prevent the flow of saliva into the tracheal stoma. Neck computed tomography (CT) scan performed a month before CPL revealed a laryngeal foreign body; however, her guardians failed to identify it on occurrence (Fig. 1a,b). When the patient was 15 years of age, we performed CPL with foreign body removal and resected the pharyngocutaneous fistula to prevent saliva-flow into the tracheal stoma (Fig. 2a,b). Visual images of the central-part of the larynx and the pharyngocutaneous fistula resection (before and after incision) resection are shown in Fig. 3a–c. At the 6-month follow-up, the patient had neither risk of aspiration nor postoperative complications.

3. Discussion

Aspiration prevention surgeries are indicated in patients with history of recurrent aspiration pneumonia, resistant to rehabilitations and medications. Most patients eligible for aspiration prevention surgeries have severe respiratory dysfunction, malnutrition, and poor performance status [6].
Central-part laryngectomy was first reported by Kawamoto et al. [4]. The characteristics of CPL are narrow-field laryngectomy, minor invasion, and minor complications.

A unique point in this case is the development of pharyngocutaneous fistula 9 years after laryngotracheal separation. The pediatric surgeons used an Endo-GIA stapler to separate the trachea from the larynx, and a minor leak from surgical site to skin gradually developed. In addition, the tooth that obstructed the patient's larynx at 11 years of age possibly played a pivotal role in worsening the pharyngocutaneous fistula. Laryngotracheal separation is an effective technique for intractable aspiration management [7]; however, Zocarro KB reported that 43% (24/60) of patients developed postoperative complications, such as pharyngocutaneous fistula, infection at the surgical site, and tracheostomal stenosis [8]. The Endo-GIA is essential for current thoracic and tracheal surgeries [9,10] and other surgical fields. The Endo-GIA is also indispensable in pediatric surgery, and the stapled anastomosis was reported to provide equivalent results as hand-sewn anastomosis [11,12]. In our case, a foreign body was in the patient's larynx (Fig. 1b); and pathological findings could not reveal its nature, and the parents/guardians did not recognize stranguation by the foreign body. The pathologists revealed that the foreign body was an artificial object and not a tooth, suggesting that the foreign body was a piece of toothbrush because the guardians reported that the patient frequently bit a toothbrush by mistake. This chronic, continuous mechanical stress might have induced inflammation and leakage of the surgical site. Sharp dissection with a #15-blade scalpel was needed to separate the pharyngocutaneous fistula from the esophagus, indicating that severe adhesions existed around the fistula.

Between April 2016 and July 2021, we performed CPL in 12 patients to manage intractable aspiration at our institution. The clinical characteristics of these patients are shown in Table 1. We did not evaluate loss of voice because none of our patients could speak before surgery. Moreover, not all patients had functional oral intake. The median surgical time was 173 min even though seven distinct surgeons performed CPL. This time was significantly shorter than that of conventional total laryngectomies (Supplementary Table 1) to manage intractable aspiration between June 2004 and January 2011 (p = 0.0007) (Fig. 4), indicating that CPL is easier than conventional total laryngectomy. In addition, no surgical complication was reported in our 12 cases (Table 1). The main

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**Table 1.** Characteristics of 12 patients who underwent CPL. In our study, no patients had speaking ability before surgery.

<table>
<thead>
<tr>
<th>No.</th>
<th>Age</th>
<th>Gender</th>
<th>Disorders causing aspiration</th>
<th>Preoperative patient’s conditions</th>
<th>Surgical time (minutes)</th>
<th>Bleeding (mL)</th>
<th>Surgeon</th>
<th>Surgical complication</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>46</td>
<td>Male</td>
<td>CP</td>
<td>Done</td>
<td>200</td>
<td>277</td>
<td>A</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>36</td>
<td>Male</td>
<td>SSPE</td>
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<td>207</td>
<td>207</td>
<td>A</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>26</td>
<td>Male</td>
<td>CP</td>
<td>No</td>
<td>132</td>
<td>76</td>
<td>B</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>57</td>
<td>Female</td>
<td>Brainstem infarction</td>
<td>Done</td>
<td>187</td>
<td>87</td>
<td>A</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>18</td>
<td>Male</td>
<td>CP</td>
<td>No</td>
<td>161</td>
<td>0</td>
<td>C</td>
<td>None</td>
</tr>
<tr>
<td>6</td>
<td>64</td>
<td>Male</td>
<td>ALS</td>
<td>No</td>
<td>168</td>
<td>47</td>
<td>D</td>
<td>None</td>
</tr>
<tr>
<td>7</td>
<td>32</td>
<td>Male</td>
<td>CP</td>
<td>No</td>
<td>174</td>
<td>0</td>
<td>E</td>
<td>None</td>
</tr>
<tr>
<td>8</td>
<td>64</td>
<td>Male</td>
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<td>No</td>
<td>150</td>
<td>49</td>
<td>F</td>
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</tr>
<tr>
<td>9</td>
<td>70</td>
<td>Female</td>
<td>ALS</td>
<td>No</td>
<td>173</td>
<td>60</td>
<td>A</td>
<td>None</td>
</tr>
<tr>
<td>10</td>
<td>49</td>
<td>Male</td>
<td>CP</td>
<td>No</td>
<td>213</td>
<td>0</td>
<td>C</td>
<td>None</td>
</tr>
<tr>
<td>11</td>
<td>22</td>
<td>Female</td>
<td>Traumatic brain injury</td>
<td>No</td>
<td>154</td>
<td>56</td>
<td>A</td>
<td>None</td>
</tr>
<tr>
<td>12</td>
<td>15</td>
<td>Female</td>
<td>MDS</td>
<td>Done</td>
<td>173</td>
<td>26</td>
<td>G</td>
<td>None</td>
</tr>
</tbody>
</table>

limitation of this study was the limited number of patients. Further studies are required to elucidate the benefits of CPL to manage intractable aspiration and pharyngocutaneous fistula in a larger population.

4. Conclusion

We presented the case of a patient with MDS who underwent CPL 13 years after laryngotracheal separation to manage pharyngocutaneous fistula. Central-part laryngectomy is a superior technique to control intractable aspiration because it is less invasive and easier than conventional total laryngectomy.

Ethical statement

This study followed Helsinki Declaration principles. We obtained from written informed consent from the patient’s guardians.

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Declaration of Competing Interest

All authors declare that there are no conflicts of interest.

Supplementary materials


CRediT authorship contribution statement


References