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Case Report

Recurrent Esophageal Stricture in a Child Post Steven-Johnson Syndrome: A Case Report

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Abstract:

Background: Esophageal stricture is an abnormal narrowing of the esophageal lumen, resulting in dysphagia. Despite its rarity, this condition could be caused by various etiologies, including Steven-Johnson Syndrome (SJS). In some cases, stricture could recur, which complicates the management. This case report presented a rare case of refractory esophageal stricture in children with Steven-Johnson Syndrome.

***Case*:** A 5-years-old boy with a prior history of SJS presented with dysphagia for one month. The patient experienced choking, blood vomiting, stomatitis, swelling on the lips, and difficulty in swallowing solid food. Barium meal and EGD test confirmed the diagnosis of esophageal strictures. The patient then underwent dilation using bougie dilator. However, he continued to experience dysphagia, resulting in a total of 15 serial dilation sessions.

Discussion: Esophageal dysphagia is observed in patients who experience difficulty swallowing solid food. SJS can contribute to the development of esophageal dysphagia by causing inflammation of the esophageal mucosa, resulting in lesions and strictures. In patients with esophageal strictures, two types of dilation methods are available: bougie dilator with wire guidance (Savary-Gilliard) and balloon dilator, with the current consensus for dilation procedures supporting the rule of three. For patients with refractory strictures, other modalities such as mitomycin-C injection and stent placement are also available. Esophageal replacement surgery is considered as the last resort for refractory stricture patients who have not responded to previous treatments.

***Conclusion*:** Steven-Johnson Syndrome is a rare cause of esophageal strictures. The management of refractory esophageal stricture requires a comprehensive subspecialty care and long-term monitoring.

Keywords: dysphagia, esophageal stricture, steven-johnson syndrome, refractory

Introduction

Esophageal stricture is a rare cause of dysphagia in children. It is defined as an abnormal narrowing of the esophageal lumen due to various etiologies. The most common etiology is the ingestion of corrosive substances. Other etiologies include radiation-related injury, post-anastomosis stricture, and eosinophilic esophagitis.[1] Inflammation of esophageal mucosa in Steven Johnson syndrome is an atypical cause of esophageal stricture.[2] Additionally, management of esophageal stricture remains challenging. Balloon dilation or bouginage is the initial management for esophageal stricture.[3] However, in some cases, evaluation after several dilation sessions revealed a recurrent or refractory esophageal stricture. Administration of steroids or Mytomycin-C has become one of the treatment choices for those experiencing refractory stricture.[4] Other alternatives, such as stent placement also reported to be successful in some refractory cases. This case report presented a case of refractory esophageal stricture in children with Steven-Johnson Syndrome.

Case

A boy aged five years and eight months old came with a chief complaint of difficulty swallowing for the past month. A month prior, the patient had a choking episode, followed by vomiting reddish vomit mixed with food. He also experienced stomatitis and swelling on the lips (**Figure 1**). The symptoms occurred after the administration of acetylsalicylic acid. He was then hospitalized for three days. Two weeks after the admission, the patient had another episode of vomiting. He could only eat porridge and was unable to eat solid food. Upon eating solid food, the patient felt something stuck in his throat, causing him to vomit. During this period, the patient lost 3 kg of body weight. Physical examination showed no abnormalities in the mouth and tonsils.

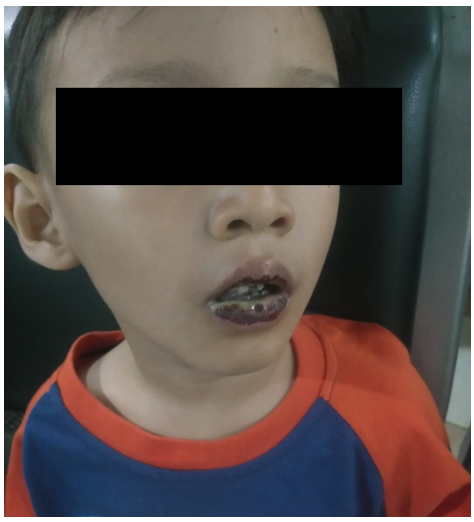

Figure 1. Stomatitis dan Swelling presented on the patient

Rhinopharyngolaryngoscopy (RFL) test revealed laryngopharyngeal reflux, adenoid hypertrophy, and mechanical dysphagia. Further examination with flexible endoscopic evaluation of swallowing (FEES) showed good swallowing movement in the pharyngeal phase but also noted reflux from the esophagus. The barium meal test indicated an esophageal stricture from thoracic vertebra 5 to 8, measuring 4.5 centimeters in length (**Figure 2**). The result of esophagogastroduodenoscopy (EGD) also demonstrated a 5-centimeter esophageal stricture, 15 centimeters from the incisors, and a presentation of gastroduodenitis. No furrowing was found in the esophagus (**Figure 3**).

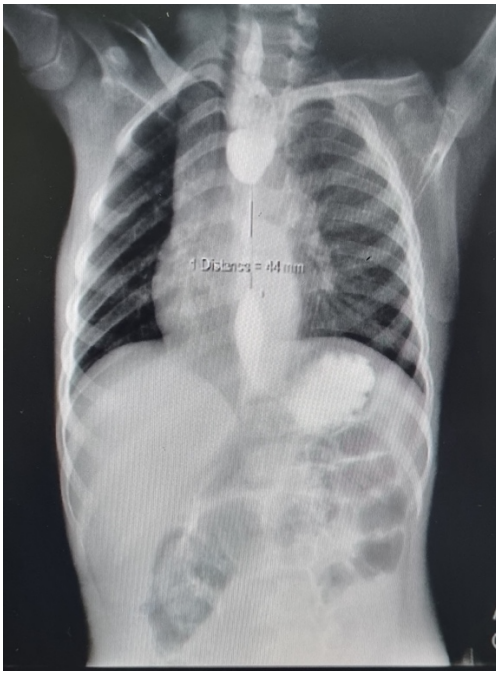

Figure 2. Barium Meal Test

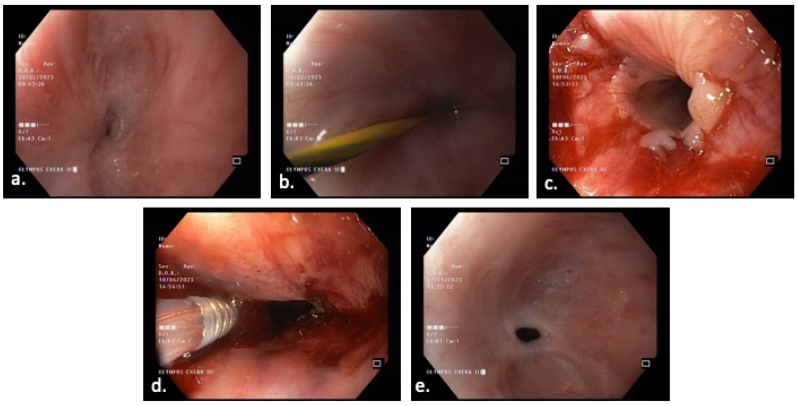

Figure 3. EGD Results. a. Esophageal stricture pre-dilation; b. First dilation; c. Laceration post-dilation; d. Triamcinolone injection post-dilation; e. The diameter of lumen after multiple dilation sessions

The patient then underwent dilation using Savary-Gilliard bougie number 7 and nasogastric tube insertion with the assistance of a nasal scope. Biopsies from the duodenum, antrum, and distal esophagus revealed no eosinophils was found in any tissues. A second EGD was conducted one month after the first procedure, with the result of an 8-millimeter stricture. Dilation procedures were then repeated using bougie no. 8, 9, and 10.8, until a 9.2 mm scope could enter the stomach and duodenum. The examination showed strictures were still present at 6cm, 14 cm, and 20 cm from the incisor. During the follow-up evaluation three weeks after the procedure, the patient still reported difficulty swallowing solid food. Triamcinolone 20 mg was then injected at several laceration points during the fourth to sixth dilation sessions. Despite the intervention, the patients continued to experience dysphagia 3-4 weeks post-dilation, leading to a total of 15 serial dilation sessions.

Discussion

Swallowing is a complex process that occurs sequentially. The swallowing process can be categorized into 3 phases: the oral phase, pharyngeal phase, and esophageal phase. Dysphagia can occur in any of these stages, either acutely or chronically. Acute inflammation along the swallowing pathway may cause dysphagia, including retropharyngeal abscess, diphtheria, acute epiglottitis, or Steven-Johnson Syndrome (SJS). Acute dysphagia occurring in the esophageal phase could be caused by swallowing a foreign body, swallowing a caustic substance, or esophageal perforation.[1]

The complaint of difficulty swallowing experienced by the patients one month prior indicated an acute dysphagia. History-taking that should be assessed for this condition includes the presence of ingested foreign objects, pain during swallowing, pain related to eating, fever, skin or mucosal lesion, voice disturbance (dysphonia), acute neurological deficit, and whether swallowing difficulty is limited to solid food or also occurs while consuming liquid food. Dysphagia related to eating might result from gastroesophageal reflux disease (GERD). The presence of fever suggests an infectious etiology. Furthermore, dysphagia occurring only due to solid food indicates esophageal dysphagia, while dysphagia due to both solid and liquid food suggests an esophageal motility disorder such as achalasia.

From the history taking, the patient experienced difficulty swallowing, lesion and swelling on the lips after drug consumption (acetylsalicylic acid). The patient could not swallow solid food but could take liquid food, suggesting esophageal dysphagia. Dysphagia due to solid food is usually caused by esophageal inflammation resulting from GERD, eosinophilic esophagitis (EoE), or esophageal stricture.[5] He then underwent esophageal contrast meal and esophagogastroduodenoscopy with histopathological examination.[6]

The history of lip swelling and bloody vomit after taking acetylsalicylic acid indicated that the cause of dysphagia was esophagitis due to SJS. Steven-Johnson syndrome could cause complications in many organ systems, including gastrointestinal. Among the SJS patients who were suspected of having gastrointestinal complication and underwent endoscopy, 11% of them had abnormalities such as ulceration or stricture, primarily in esophagus.[2]

Barium meal examination confirmed the occurrence of esophageal stricture, which was also visualized using EGD. One of the potential etiologies of esophageal stricture that needs to be considered is EoE. However, the endoscopic findings typically associated with EoE, such as exudate, trachealization, and furrowing, were not observed in this case. Furthermore, the histopathological result indicating EoE, which would be presented as significant eosinophils (≥15 cells/LPB) in the esophagus, were also not found in this case.[7]

Two dilation techniques are available to treat esophageal stricture: bougie dilator with wire guidance (Savary-Gilliard) and balloon. Balloon dilation has better safety profile and lower rate of failure compared to the bougie dilation.[3] However, bougie dilation is a safe and effective procedure; thus, it is still commonly practiced in hospital with limited resources. The experience of the hospital and healthcare provider performing one of the dilation techniques also determined the success of therapy.[8] Bougie dilation was used in our case. Currently, there is still no guideline regarding the time and frequency of dilation for children with esophageal stricture. Research comparing the dilation on esophageal atresia per three weeks with on-demand revealed that the on-demand group underwent fewer dilations with the same efficacy and safety.[9] The current general consensus is the rule of three: maximum dilation up to three times the diameter of stenosis, maximum three dilations per session (with an increase of 1 mm per dilation) after resistance, and a minimum interval of three weeks between dilation sessions.[9] In infants under three months of age, optimal dilation could be achieved with 8-10 diameter dilation, while in older children, it can be achieved with 10-15 mm diameter dilation.[10] Long-term management of our patient was challenging, mainly due to the recurrent stricture after dilation sessions. Refractory esophageal stricture occurs due to scarring or fibrosis in the lumen, causing the recurrence of narrowing without inflammation. Refractory stricture is diagnosed if the expected esophageal diameter cannot be achieved after five sessions with maximum four-week intervals, or if the expected diameter could not be maintained for four weeks after the diameter is achieved.[11] In our case, the patient had undergone 15 times of dilation, establishing the diagnosis of refractory stricture. Currently, there is still no standard therapy for refractory stricture. However, several modalities that can be used for refractory stricture.

The application of mitomycin-C at the mucosal lesions post-dilation has been reported to be beneficial in reducing the total dilation sessions. Mitomycin-C is an antifibrotic and cytostatic agent anthracycline group that inhibits fibroblast proliferation and reduces scar formation. For esophageal strictures, mitomycin-C is administered by applying a cotton swab soaked in a 0.1 mg/mL mitomycin-C solution directly to the mucosa following dilation. Mitomycin-C was shown to be safe and effective for refractory strictures.[4]

Management using plastic or nickel-titanium alloy stents could also be considered as they are removable and able to handle angulation in the esophagus. The use esophageal stent in children increased with the presence of self-expandable metal or plastic stents. There is still no guidance regarding the duration of stent insertion, causing varying duration from 1 to 24 weeks. A study in pediatric patients demonstrated an effective use of stents in 52% of the cases without further intervention needed.[12] Additionally, intralesional injection of triamcinolone has been reported to be effective for refractory stricture in adult patients. The main advantage of triamcinolone injection lies in its relatively safe administration technique and its ability to reduce the frequency of dilation sessions. The dosage of triamcinolone administered to patients varied across different centers, typically ranging from 20 mg to 40 mg per lesion. In pediatric patients, the use of intralesional triamcinolone has been reported to yield varying outcomes, with some showing success while others do not.[13]

Esophageal replacement surgery is the last choice of treatment for refractory stricture. Several organs were reported eligible for esophagus substitution, including colon segments, stomach, and small intestine segments, but none can perfectly replace esophageal function. The long-term prognosis for colon interposition following pediatric esophagectomy also did not exhibit promising results with high morbidity and multiple organ disorders, including gastrointestinal symptoms (85%), respiratory symptoms (58%), and eating difficulties (50%). Furthermore, the majority of patients experienced failure to thrive.[14]

Conclusion

Steven Johnson Syndrome is a rare cause of esophageal strictures. The management of refractory esophageal strictures requires comprehensive subspecialty care and long-term monitoring. In this case, the treatment plan for refractory stricture includes the application of mitomycin-C or stent placement. Consultation with pediatric surgeon for esophagus substitution therapy should also be considered if dilation results are not optimal, despite the high morbidity.

Conflict of Interest

None declared

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