CASE REPORT

Improvement in Reflux Gastroesophagitis in a Patient with Spinal Muscular Atrophy after Surgical Correction of Kyphoscoliosis

A Case Report

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Received: 25 January 2011 / Accepted: 30 August 2011 / Published online: 5 October 2011
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Abstract

Background Scoliosis, a three-dimensional deformity, has secondary effects on the gastrointestinal system. Reflux gastroesophagitis with hiatus hernia in patients with scoliosis is difficult to manage. We present a patient in whom primary correction of a spinal deformity was associated with resolution of symptoms of reflux.

Case Description A 15-year-old girl with severe thoracolumbar kyphoscoliosis visited our scoliosis research institute complaining of back pain, positional imbalance, intermittent

respiratory tract infections, and gastrointestinal discomfort such as pain, dysphagia, and heartburn for several years. On preoperative CT, her abdominal organs were in a deviant position, and esophagogastroduodenoscopy revealed severe reflux gastroesophagitis, Los Angeles classification (LA) Grade D, and a sliding hiatus hernia. After kyphoscoliosis correction, the patient's truncal balance and pain improved. Postoperatively, the patient reported abdominal pain and dysphagia that gradually subsided after 3 weeks. At 1 year, the patient had no abdominal complaints secondary to reflux gastroesophagitis, and episodes of recurrent respiratory tract infections were substantially reduced. Postoperative evaluation showed the reflux gastroesophagitis had improved to LA Grade A. Postoperative CT showed the abdominal cavity had expanded and the abdominal organs were more centered.

Literature Review The association between scoliosis and reflux gastroesophagitis is well documented. However, the secondary effects of scoliosis correction on gastrointestinal symptoms caused by reflux gastroesophagitis have not been investigated in detail.

Purpose and Clinical Relevance This patient illustrates the relationship between spinal deformity and gastrointestinal symptoms. Postural balance correction resulted in the alleviation of reflux gastroesophagitis symptoms secondary to hiatus hernia.

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All ICMJE Conflict of Interest Forms for authors and *Clinical Orthopaedics and Related Research* editors and board members are on file with the publication and can be viewed on request. Each author certifies that his or her institution approved the human protocol for this investigation, that all investigations were conducted in conformity with ethical principles of research, and that informed consent for participation in the study was obtained. This work was performed at Korea University Guro Hospital, Seoul, Korea.

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Introduction

The association between neuromuscular scoliosis and hiatus hernia is documented [2, 10]. Axial deviation of the vertebral column at the level of the hiatus results in decreased intraabdominal volume and stretching of the esophageal sphincter [6]. The increased intraabdominal pressure causes hiatus hernia and reflux gastroesophagitis [8].

Symptoms of reflux gastroesophagitis vary from simple abdominal discomfort, such as heartburn or dysphagia, to recurrent lung infection and respiratory distress. When not controlled by pharmacotherapy, this condition requires surgery [11, 12]. However, in patients with neuromuscular scoliosis, surgical correction can reduce the intraabdominal pressure, improve postural balance, and reduce the symptoms of gastroesophageal reflux disease (GERD). Therefore, if the main cause of reflux gastroesophagitis with hiatus hernia is spinal deformity, correction of the deformity might alleviate the reflux symptoms. To the best of our knowledge, no previous studies have reported improvements in hiatus hernia and reflux gastroesophagitis in patients surgically treated for scoliosis.

Case Report

A 15-year-old girl with severe thoracolumbar kyphoscoliosis visited our scoliosis research institute complaining of back pain, positional imbalance that required her to use her upper extremities to support her body (Fig. 1A), intermittent respiratory tract infections, and gastrointestinal discomfort, such as abdominal pain, dysphagia, and heartburn for several years. On physical examination, the patient's height and weight were 138 cm and 26 kg, respectively. Plain radiography revealed severe kyphoscoliosis (Fig. 1B). Cobb's angle, which was measured from the third thoracic vertebra to the fourth lumbar vertebra, was 162° and the kyphotic angle, which was measured from the first to twelfth thoracic vertebra, was 106° . CT showed the abdominal organs deviated to the convex side of the patient's torso and the chest wall was distorted along

the deformed spine and rib cage (Fig. 1C). Thus, the axis between the esophagus and the stomach clearly deviated to the left, compromising the abdominal contents. The patient's forced vital capacity was 21% and her forced expiratory volume in 1 second was 17%. Genetic evaluation revealed a mutation of the SMA gene. In view of the patient's abdominal complaints, a gastroenterologist examined the girl and she underwent endoscopy. Endoscopy revealed severe reflux gastroesophagitis of LA Grade D and a sliding hiatus hernia [4]. Multiple biopsies of the esophageal lining showed inflammation. The patient had been taking histamine-2 receptor antagonists intermittently for her abdominal complaints (heartburn and abdominal pain), but she never experienced complete or long-term relief. In view of her long-standing symptoms and severe reflux gastroesophagitis, the gastroenterologist first suggested a positional change; however, owing to her spinal deformity and back pain, she was unable to sit for as much as half an hour. Therefore, as a second treatment option, the gastroenterologist suggested fundoplication.

Because the patient's main complaints were spinal deformity, back pain, and positional imbalance, we decided to correct the deformity first, and then reexamine her gastrointestinal problem after spinal correction. The spinal deformity was corrected with a posterior-only pedicle screw system. Postoperative radiographs showed a reduction in the Cobb angle from 162° to 62° and of the kyphotic angle from 106° to 13° (Fig. 2). The postoperative CT results for this patient showed the axis between the esophagus and stomach became vertical and almost anatomically normal (Fig. 2B). The intraabdominal space increased after surgery, and the internal organs centrally aligned. Postoperatively, the patient reported abdominal pain and dysphagia, but these

Fig. 1A–C (A) The patient had severe scoliosis, pelvic tilting, and contact between the ribs and pelvis, indicating a decreased abdominal space. (B) Severe scoliosis of the thoracolumbar spine was visible on a plain radiograph. (C) The internal organs of the abdomen deviated from the midline and the cavity was distorted on a coronal CT image.

The gastroesophageal junction, stomach, and liver are indicated by (1), (2), and (3), respectively, and the red line is the axis between the esophagus and stomach. The axis line of the stomach and esophagus deviates to the left, decreasing the space available for the abdominal organs.

Fig. 2A–C (A) Correction of the spinal deformity and some restoration of the abdominal cavity can be seen on this postoperative radiograph. (B) Postoperative coronal CT images showed that the axis (red line) between the esophagus and stomach was almost vertical and anatomically normal. The esophagus, gastroesophageal junction,

stomach, and liver are indicated by (1), (2), (3), and (4) respectively, and the red line is the axis between the esophagus and stomach. (C) Three weeks after the operation, the patient was able to balance herself in a seated position.

Fig. 3A–C (A) Slackness of the gastroesophageal flap valve (white arrow) is evident before surgery. (B) The slackness was improved as observed on followup endoscopy after 7 months (empty block arrow). (C) The flap valve function was well preserved at the last followup endoscopy at 1 year.

symptoms gradually subsided after 3 weeks. She was prescribed regular histamine-2 antagonists for 1 month, but required no additional treatment. An esophagogastroduodenoscopy was performed at 3 weeks. The gastroenterologist reported that insertion of the esophagogastroduodenoscope was much easier after correction of the curve. On esophagogastroduodenoscopy, the sliding hiatus hernia was still visible, but the reflux gastroesophagitis had subsided. After surgery, the patient experienced substantial improvements in postural balance, back pain, and gastrointestinal symptoms. At 1-year followup, she denied gastrointestinal or respiratory symptoms, and reported she could sit for 2 hours, which she could not do before surgery (Fig. 2C). Repeat esophagogastroduodenoscopy revealed the flap valve mechanism of the lower esophageal sphincter had a normal function compared with that of the slack flap valve that was observed preoperatively (Fig. 3).

Furthermore, the mucosal breaks had healed with scarring, and the size of the hiatus hernia had decreased from 5 cm preoperatively to 3 cm postoperatively (Fig. 4). The patient's LA classification grade improved from D to A.

Discussion

Epidemiologic studies of the natural history of GERD and its complications in older children are scarce. Children with chronic respiratory and neurologic diseases commonly experience recurrent or chronic GERD symptoms. Half of the older children with GERD have chronic relapsing disease [3]. The first line of treatment for combined reflux gastroesophagitis with hiatus hernia is conservative therapy with an emphasis on thickened meals, smaller meal volume, proper positioning, and medicines. Operative

Fig. 4A–C (A) Preoperative evaluation revealed whitish discoloration of the esophageal mucosa with patchy detachment, and the reflux gastroesophagitis was classified as Grade D using the Los Angeles classification system. (B) There was marked improvement in the preoperative patchy detached esophageal lesion 3 weeks after

surgery, and the Los Angeles classification grade was downgraded to B. (C) An ulcer scar was observed at the squamocolumnar junction, but no definite mucosal breaks were present at 1 year; the reflux gastroesophagitis was classified as Grade A using the Los Angeles classification system.

intervention is considered in patients who are refractory to pharmacotherapy or have a large hiatus hernia, long segment Barrett's esophagus, and/or recurrent ear-nose-throat and respiratory symptoms [8, 12]. However, the treatment guidelines for reflux gastroesophagitis combined with hiatus hernia in children with scoliosis are not clear. Surgery in such patients carries an increased risk, particularly from respiratory and nutritional standpoints, and can be life threatening [12]. Therefore, treatment for such patients should be managed by a multidisciplinary team. Important factors that contribute to the development of hiatus hernia in patients with neuromuscular scoliosis are postural imbalance, reduced intraabdominal space, and increased intraabdominal pressure. Correction of the primary spinal deformity helps realign the internal abdominal organs, thereby reducing back pressure and the effects of reflux gastroesophagitis. Postoperatively, the patient's postural balance improved, which also helped to reduce her gastroesophageal reflux symptoms. Correction of spinal deformity could result in better treatment outcomes in patients with combined hiatus hernia and reflux gastroesophagitis [7, 9, 10]. Previous reports recommended surgical treatment to address the combination of reflux gastroesophagitis and hiatus hernia for patients who were unresponsive to pharmacotherapy [5, 7]. Because the main cause of hiatus hernia and reflux gastroesophagitis is spinal deformity, correcting the deformity may improve long-term outcome.

Corroboration of this theory can be found in the report of Dickson and Harrington [1]: they noted the incidence of hiatus hernia was 6.2% before correction of a spinal deformity in patients with adolescent idiopathic scoliosis, but the incidence decreased to 2.1% after surgery. They attributed the reduction in the size of the hernia to reduction of the scoliosis [1]. In their study, no gastrointestinal surgeries were performed, they simply recorded the incidence

rates of hernia before and after correction of the spine but did not document changes in symptoms. For our patient, her reflux gastroesophagitis healed and gastrointestinal symptoms subsided after correction of her spinal deformity. Although the hiatus hernia remained, we found that if the adaptation mechanism for the esophageal hiatus can be activated, the hiatus hernia could be treated nonsurgically.

Our patient underwent esophagogastroduodenoscopy because she had limited mouth movement owing to a temporomandibular joint problem. Furthermore, she could not tolerate a catheter for pH monitoring, and no postoperative pulmonary function test was performed because the patient refused to provide consent. The patient did not have any postoperative respiratory problems. Histologic evaluation allowed us to exclude the possibility of Barrett's esophagus or other diseases.

This case report provides insight into the relationship between gastrointestinal symptoms and spinal deformity, which is understudied and perhaps underreported. We hope the report will stimulate other spinal surgeons to further examine their patients and that it might lead to better understanding and perhaps even case series studies. We have found esophagogastroduodenoscopy is useful for evaluating such patients. However, more patients should be evaluated to determine whether observation and followup with appropriate medical treatment are sufficient to treat hiatus hernia with reflux gastroesophagitis after correction of spinal deformity.

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