The Korean Journal of Internal Medicine: 18:238-240, 2003

A Case of Mallory-Weiss Syndrome Complicating Pregnancy in a Patient with Scleroderma

Kyu-Hyun Cho, M.D., Seong-Wook Heo, M.D., Seung-Hie Chung, M.D., Chae-Gi Kim, M.D. Ho Gak Kim, M.D. and Jung-Yoon Choe, M.D.

Department of Internal Medicine, Catholic University of Daegu School of Medicine, Daegu, Korea

The majority of patients with scleroderma have gastrointestinal involvement[1], and a few experience gastrointestinal hemorrhage, however, gastrointestinal hemorrhage due to Mallory-Weiss syndrome is very rare[2, 3].

We report upon a 24-year-old pregnant woman with scleroderma who had gastrointestinal hemorrhage due to Mallory-Weiss syndrome.

Key Words : Scleroderma, Mallory-Weiss syndrome, Gastrointestinal hemorrhage

INTRODUCTION

Scleroderma is a chronic multi-systemic disorder of unknown etiology characterized clinically by thickening of the skin caused by an accumulation of connective tissue, and by the involvement of visceral organs, including the gastrointestinal tract, lungs, heart, and kidneys. The majority of patients show gastrointestinal involvement. However, this seldom results in bleeding[1], and gastrointestinal hemorrhage due to Mallory-Weiss syndrome in patients with scleroderma is very rare.

To our knowledge, the case described here is the first report of Mallory-Weiss syndrome in a pregnant woman with scleroderma in Korea.

CASE REPORT

A 24-year-old woman with a 2-year history of scleroderma with Raynaud's phenomenon visited our rheumatism center in 1998 due to pain and swelling of the joints of both hands and wrists. She had also been suffering from heartburn and dysphagia. Esophageal manometry revealed no peristalsis or contraction of the lower esophagus, and poor relaxation of the lower esophageal sphincter (Figure 1). Omeprazole (Losec®) and cisapride (Prepulsid®) were prescribed with diltiazem (Herben®) and D-penicillamine until 1999.

In 2001, she revisited our hospital at 28 weeks' gestation. And, at 34 weeks' gestation, although her condition remained relatively stable, we noted a decreased fetal movement on the ultrasound scan. A week later, she was admitted to our hospital due to premature rupture of membrane. On examination at admission, generalized pitting edema and albuminuria were found. An intrauterine fetal death was diagnosed by ultrasound scan, and plans were made to terminate the pregnancy.

The day after her admission, intractable vomiting and retching developed, which led to approximately 300 mL of hematemesis. She was pale and moderately dehydrated. though her blood pressure (130/90 mmHg) was within the normal range and her heart rate (106/min) slightly increased. Her hemoglobin level had fallen from 9.7 g/dL to 7.6 g/dL within 12 hours, but the other blood counts, prothrombin time, partial thromboplastin time, electrolytes and liver enzymes were normal.

Despite conservative management and blood transfusion, she continued to bleed. Therefore, an emergency upper gastrointestinal endoscopy was performed, which revealed large blood clots in the distal esophagus and stomach. After removing the blood clots, a Mallory-Weiss tear was discovered

• Received : March, 25, 2003
• Accepted : April, 23, 2003
• Correspondence to : Jung-Yoon Choe, M.D., Department of Internal Medicine, Daegu Catholic University Hospital, 3056-6 Daemyung4-dong, Nam-gu, Daegu, 705-718, Korea Tel : 82-53-650-4027, Fax : 82-53-629-8248, E-mail : jychoe@cataegu.ac.kr

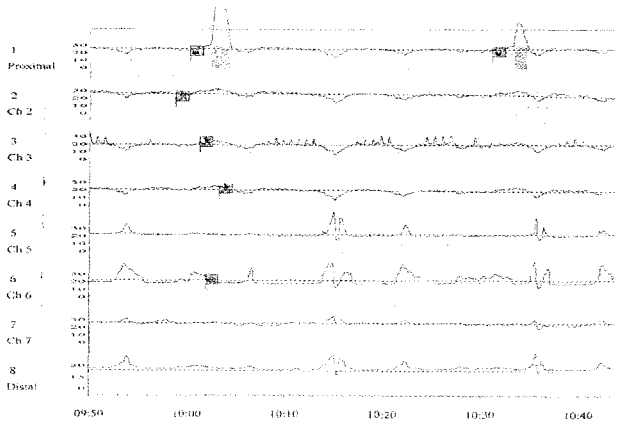

Figure 1. Esophageal manometry showed no peristalsis or contraction of the lower esophagus and poor relaxation of the lower esophageal sphincter.

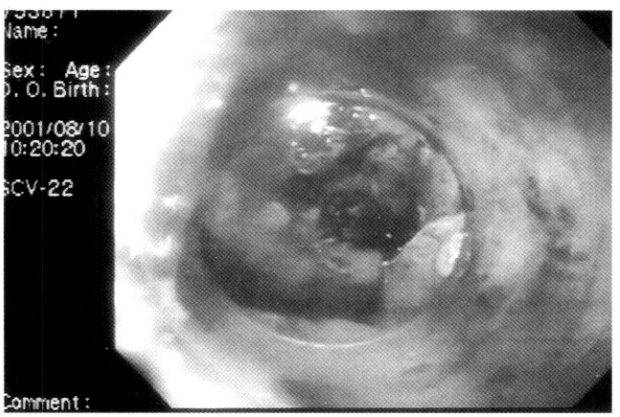

Figure 2. A long broad laceration with active bleeding at the esophagogastric junction (Mallory-Weiss tear).

near the EG junction (Figure 2). The stomach and duodenum appeared normal. Mucosal ligation and hypertonic saline injection were performed for hemostasis.

Two hours after endoscopy, she developed a generalized tonic-clonic type seizure. Her airway was maintained and oxygen was supplied. Diazepam (Valium®) and magnesium sulfate were given intravenously. Her blood pressure increased from 130/90 mmHg to 150/100 mmHg and her pulse rate normalized. An urgent Caesarean section was performed for eclampsia. Post-operatively, she experienced 5 further seizures. Diffuse brain swelling and slightly low-density lesions in the external capsules and in the subcortical white matter were found by CT due to hypertensive encephalopathy. Magnesium sulfate and 20% mannitol were administered by intravenous injection for 3 days.

A total of 10 units of blood were transfused during her 9 day hospital stay. At discharge, there was no more evidence of hemorrhage, and her condition was totally stabilized. She remained stable without further manifestations of gastrointestinal hemorrhage over the following 8 months, when she was lost to follow-up.

DISCUSSION

Scleroderma is an uncommon disease of uncertain etiology, which is refractory to most treatments. The excessive collagen deposition associated with the disease affects the skin and certain internal organs, especially the esophagus. The majority of patients of both diffuse and limited scleroderma have gastrointestinal involvement. Symptoms attributable to esophageal involvement are present in over 50% of patients, these include; epigastric fullness, a burning pain in the epigastric or retrosternal regions, and the regurgitation of gastric contents. These symptoms are due to a reduced tone of the gastroesophageal sphincter and to dilatation of the distal esophagus. Peptic esophagitis frequently occurs and may lead to strictures and a narrowing of the lower esophagus. Chronic esophageal reflux can lead to metaplasia of the lower esophagus (Barrett's esophagus). Ulceration of the mucosa is often present and may be due to either scleroderma or superimposed peptic esophagitis. However, it seldom results in bleeding[1].

This patient developed intractable vomiting, which may have been due to the effects of scleroderma on the esophagus. In 1998, she experienced upper gastrointestinal symptoms, i.e., heartburn, postprandial vomiting and dysphagia, and was prescribed omeprazole (Losec®) and cisapride (Prepulsid®). Hendel et al. reported that long-term treatment with ranitidine (Zantac®) in scleroderma patients provides good gastroesophageal symptomatic relief and healing of the esophagitis[4]. Proton pump inhibitors are more effective at treating erosive esophagitis than H2 blockers. Metoclopramide and cisapride increase gastrointestinal motility but do not significantly improve esophageal motility; however, they increase lower esophageal sphincter tone and can be effective in some patients[1].

Symptomatic gastroesophageal reflux disease is common in pregnant women and is due to a reduced lower esophageal sphincter pressure, which is effected by changes in estrogen and progesterone levels during pregnancy. Intractable vomiting in this patient may have been due to the effects of pregnancy on the esophagus.

The reported outcomes of pregnancies in women with scleroderma have been quite variable. Ron M. and Moshe F. reported that the combination of scleroderma and pregnancy is rare, as female patients with scleroderma have an increased

risk of infertility, spontaneous abortion and stillbirth[5]. Viginia encouraged women with diffuse scleroderma to delay pregnancy until the disease has been stabilized because they have a greater risk of developing serious cardiopulmonary and renal complications early in the disease[6]. According to more recent case reports and case control studies, the risks posed by pregnancy to women with scleroderma or to their infants is not as grave as it was once thought to be[7-9]. Nevertheless, a well-timed pregnancy with careful obstetric monitoring is believed to maximize the likelihood of a successful outcome.

Andrea et al. reported that the involvement of the gastroin- occurrence of profuse hemorrhage. Mucosal telangiectasias were reported to be the most common cause of bleeding (9/22; 40.9%), followed by peptic ulcer disease (7/22; 31.8%) and erosive gastritis (3/22; 13.6%). A case of Mallory-Weiss syndrome has also been reported[2].

Most maternal deaths associated with scleroderma are the result of pulmonary fibrosis with a superimposed infection or renal involvement with hypertension. Although gastrointestinal hemorrhage associated with scleroderma is rare, massive upper gastrointestinal hemorrhage may also account for mortality[2].

Gastrointestinal bleeding due to Mallory-Weiss syndrome can occur in scleroderma patients and lead to death[10]. The syndrome may be induced by not only the fragile esophageal mucosa in scleroderma patients but also by vomiting and retching due to scleroderma or pregnancy[2, 11]. Counseling before pregnancy is probably the best policy in such cases. Furthermore, the possibility of Mallory-Weiss syndrome in a pregnant patient with scleroderma should be considered.

REFERENCES

1) Bruce C. *Gilliland Systemic Sclerosis (Scleroderma) In: Braunward E, Fauci AS, Kasper DL, Hauser SL, Longo DL, Jameson JL eds. Harrison's principles of internal medicine. 15th ed. pp.1937-1945, New York, McGraw-Hill, 2001*
2) Andrea Duchini, Sandra L. Sessoms. *Gastrointestinal Hemorrhage in Patients with Systemic Sclerosis and CREST Syndrome. Am J Gastroenterol 93:1453-1456, 1998*
3) K. A. J. Chin, C. M. Kaseba, J. B. Weaver. *Mallory-Weiss syndrome complicating pregnancy in a patient with scleroderma : diagnosis and management. Br J Obstet Gynecol 102:498-500, 1995*
4) Hendel L., Aggestrup S., Stentoft P. *Long-term ranitidine in progressive systemic sclerosis (scleroderma) with gastroesophageal reflux. Scand J Gastroenterol 21:799-805, 1986*
5) Ron M., Moshe F. *Scleroderma in pregnancy. Obstet Gynecol Surv 44:530-534, 1989*
6) Viginia D. Steen. *Scleroderma and pregnancy. Rheumatic Disease Clinics of North America 23:133-147, 1997*
7) Viginia D. Steen, Thomas A. Medsger, JR. *Fertility and Pregnancy with Systemic Sclerosis. Arthritis & Rheumatism 42:763-768, 1999*
8) Viginia D. Steen. *Pregnancy in Women with Systemic Sclerosis. Obstetrics & Gynecology 94:15-20, 1999*
9) Barbara J. Doss, Suzanne M. Jacques, Maureen D. Mayes, Faisal Qureshi. *Maternal Scleroderma: Placental Findings and Perinatal Outcome. Human Pathology 29:1524-1530, 1998*
10) Jeffrey M. Harris, Jack A. DiPalma. *Clinical Significance of Mallory-Weiss Tears. Am J Gastroenterol 88:2056-2058, 1993*
11) Shiobhan Weston, Miriam Thumshirn, Julie Wiste, Michael Camilleri. *Clinical and Upper Gastrointestinal Motility Features in Systemic Sclerosis and Related Disorders. Am J of Gastroenterol 93:1085-1089, 1998*