Esophagopericardial Fistula in a Scleroderma Patient With Peptic Esophagitis

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• **Esophageal perforation from peptic ulceration is rare. A patient with scleroderma who had an esophagopericardial fistula caused by peptic esophagitis is described. The fistula mimicked an acute myocardial infarction.**
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Reflux esophagitis, an uncommon cause of esophageal perforation, rarely results in an esophagopericardial fistula. Perforation of the esophagus typically extends into the mediastinum or pleural space. I report herein what I believe is the first case of spontaneous pneumopericardium resulting from perforation of the esophagus in a scleroderma patient with reflux esophagitis.

REPORT OF A CASE

A 62-year-old woman with a ten-year history of reflux esophagitis secondary to scleroderma was initially seen with sharp substernal chest pain of sudden onset. An esophagram that had been performed a year earlier demonstrated a stricture of the lower esophagus with free gastroesophageal reflux. There was no antecedent history of vomiting, change in esophageal symptoms, or pain. There was, however, a history of congestive heart failure.

On admission, the patient appeared acutely ill. General skin thickening and tightening was apparent. Muffled heart sounds were noted. An ECG was normal. A chest x-ray film disclosed a pneumopericardium (Fig 1). An esophagram was performed with water-soluble contrast which demonstrated free spillage into the pericardial sac (Fig 2). A pericardial window was opened, which immediately drained a large amount of fluid that appeared to contain gastric contents. The fistulous tract was not directly visualized. After aspiration and irrigation of the pericardial cavity, soft, flat drains were left. Supportive treatment and antibiotic therapy was provided, and after several days the patient became afebrile. A week following admission, a repeated esophagram showed apparent closure of the fistulous tract. The patient continued to make a satisfactory recovery and the drainage tubes were removed after ten days. No further surgical intervention was attempted because of the patient's poor overall condition. The patient was discharged on a regimen of a soft diet, antacids, and cimetidine.

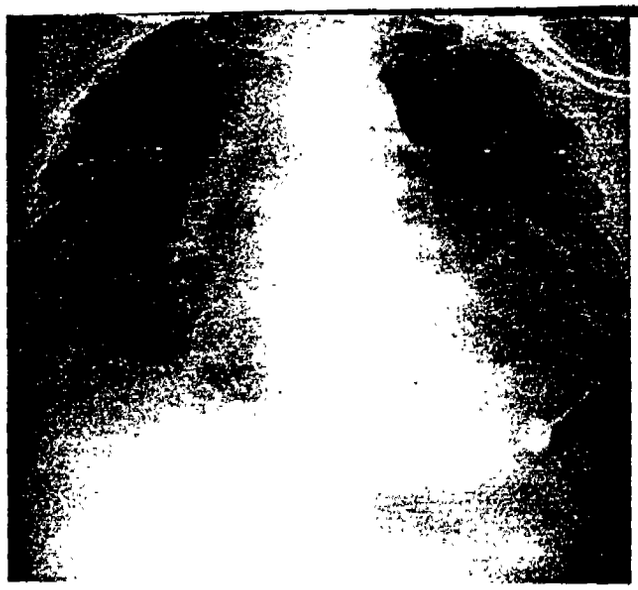

Fig 1.—Portable supine chest roentgenogram demonstrating pneumopericardium.

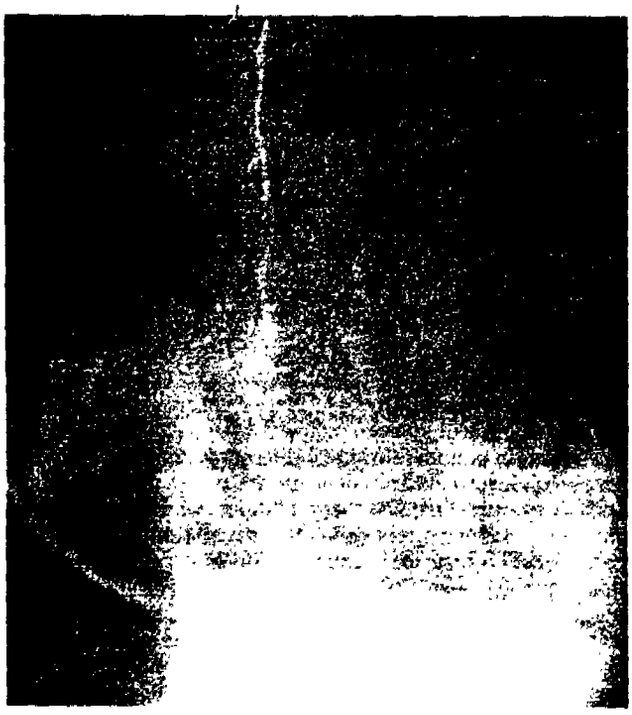

Fig 2.—Esophagram showing free spillage of contrast into pericardium with pooling of contrast on upright film.

COMMENT

An esophagopericardial fistula is a rare complication of esophageal perforation. To my knowledge, only eight patients with peptic ulceration resulting in an esophagopericardial perforation have been described in the literature since 1851.[1-8] And to my knowledge, this is the first reported case of a scleroderma-reflux esophageal perforation resulting in a pericardial fistula.

The esophagus is the internal organ most commonly involved by scleroderma. Up to 80% of patients with scleroderma will demonstrate symptoms of reflux esophagitis.[7-9] Fibrous-tissue replacement of the esophageal smooth musculature results in dilatation and poor peristalsis.[9,10] Gastroesophageal reflux in the patulous distal esophagus leads to subsequent inflammation and reflux esophagitis. Pathologic examination of the esophagus in previous cases of peptic esophagopericardial fistulas demonstrated the esophageal perforation at the site of active ulceration.[2,3,6] It is likely that a similar mechanism caused the fistula in this patient.

Early recognition is critical in the management of esophagopericardial perforations.[4] Clinically, splashing sounds corresponding to systole (*bruit de la roue hydroulique*), tympany over the precordium, and precordial shifting dullness can be detected.[6] A routine chest x-ray film will show air within the pericardium, a universal finding in patients with esophagopericardial fistulas. Water-soluble contrast examination of the esophagus is important to establish the diagnosis and site of perforation. It has been recommended that a second study be performed one hour later if the initial examination is normal and clinical suspicion of a perforation persists.[4] If treatment is initiated within 24 hours of perforation, the patient has a much better chance of survival. One of the two patients described in the literature whose peptic esophagopericardial fistulas were

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treated within 24 hours survived. Of the other seven patients with delayed therapy, only one lived.[1-6] To my knowledge, mine is the only patient to survive following a conservative drainage procedure.

This case demonstrates a rare sequela of reflux esophagitis not previously associated with scleroderma. Although esophagopericardial perforation is an unusual cause of sudden chest pain, suggestions of it can be turned up on clinical examination and easily confirmed by roentgenograms.

James R. Edinger, DO, contributed the contrast studies.

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Thrombotic Thrombocytopenic Purpura During Penicillamine Therapy in Rheumatoid Arthritis

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• Thrombotic thrombocytopenic purpura (TTP) developed in a patient with seropositive rheumatoid arthritis (RA) after 2½ months of treatment with penicillamine. After discontinuation of the drug, plasmapheresis and steroid therapy led to a sustained remission. To our knowledge, no prior cases of penicillamine-induced TTP in RA have been reported.

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Thrombotic thrombocytopenic purpura (TTP) is a rare occurrence in rheumatoid arthritis (RA) and other connective tissue diseases. To our knowledge, there has been only one previous report of TTP as a complication of penicillamine therapy, and no prior reports of penicillamine-induced TTP in RA. We describe the development of TTP in a patient with RA after 2½ months of treatment with penicillamine. Thrombotic thrombocytopenic purpura responded to various therapeutic measures and did not recur after discontinuation of penicillamine therapy.

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REPORT OF A CASE

A 64-year-old man had a 12-year history of deforming, seropositive RA that was treated with nonsteroidal anti-inflammatory drugs. A six-month course of prednisone therapy was discontinued in January 1982, when an upper gastrointestinal hemorrhage developed. After this episode, the patient received naproxen therapy, but he experienced progressive disability.

In March 1982, a physical examination disclosed active synovitis involving the metacarpophalangeal joints, wrists, elbows, and knees. There were flexion contractures of the elbows and right knee, swan neck deformities, ulnar deviation in both hands, and nodules at both elbows.

Laboratory studies disclosed the following values: hematocrit, 31%, with a normal platelet count; ESR (Westergren), 90 mm/hr; and rheumatoid factor and fluorescent antinuclear antibody both positive at a titer of 1:640. Roentgenograms showed erosions and joint space narrowing in the hands, wrists, knees, and feet. Treatment with 250 mg/day of D-penicillamine was started on March 30, 1982. Other medications included 375 mg of naproxen twice a day and 300 mg of cimetidine at bedtime. There was moderate clinical improvement during the ensuing two months and on June 1 the hematocrit reading was 33%, WBC count was 8,500/cu mm, and platelet count was 261,000/cu mm.

In mid June, he was seen by his family physician with a six-day history of fever, malaise, transitory aphasia, and confusion. An examination disclosed a temperature of 38.3 °C and numerous petechiae on his feet; a hematocrit reading was 26% and a platelet count was 45,000/cu mm. Thrombotic thrombocytopenic purpura was suspected and treatment with aspirin, dipyridamole, prednisone, and subcutaneous heparin sodium was initiated. Fresh frozen plasma was given and therapy with D-penicillamine was discontinued.

The patient was transferred to the Baptist Memorial Hospital, Memphis, on June 19, 1982. Physical examination disclosed scattered petechiae on the hands and feet. There was no peripheral adenopathy or hepatomegaly, but mild splenomegaly was noted. He had recurrent, transient episodes of expressive aphasia, but a neurologic examination showed no abnormalities. Laboratory studies disclosed the following values: hematocrit, 20%; WBCs, 15,200/cu mm, with 94% neutrophils; platelets, 56,000/cu mm; and reticulocyte count, 6%. Occult blood was not detected in the stools. Direct Coombs' test was negative. The peripheral blood smear showed numerous schistocytes. Prothrombin time, partial thromboplastin time, thrombin time, and fibrinogen were normal. Chemical profile was normal, except for elevations of the level of lactate dehydrogenase (537 IU/dL), total bilirubin (1.3 mg/dL), urea (31 mg/dL), and serum creatinine (1.3 mg/dL).

A gingival biopsy was negative for microthrombi or vasculitis. The patient responded promptly to plasma exchange, high-dose prednisone therapy, and packed RBC transfusions (Figure). He was discharged from the hospital on June 29, 1982, considerably improved, on a tapering dose of prednisone. Blood chemistry abnormalities had returned to normal. Weekly infusions of reconstituted fresh frozen plasma were given for three weeks, then discontinued. On Aug 6, 1982, he was receiving 20 mg/day of prednisone, the platelet count was 358,000/cu mm, the hematocrit reading was 36%, and there were rare fragmented RBCs. In late October, prednisone therapy had been reduced to 12.5 mg/day. The platelet count was 300,000/cu mm, hematocrit reading was 35%, and RBC morphology was normal.

COMMENT

Thrombotic thrombocytopenic purpura is a clinical syndrome characterized by microangiopathic hemolytic anemia, thrombocytopenia, fluctuating neurologic abnormalities, fever, and renal impairment. More than 500 cases have been reported in the literature, since the initial description in 1925 by Moschowitz.[1] However, only two cases have been associated with well-documented RA[2,3] and both were reported before the introduction of penicillamine. We cannot eliminate the possibility that TTP in this patient was related to the basic illness rather than to penicillamine therapy, since rechallenge was not attempted. However, TTP did not recur during a four-month interval after discontinuation of pencillamine therapy. Gingival biopsy in