which occur less frequently, are found anteriorly. AHD is unusual and may be associated with pulmonary hypoplasia, resulting in progressive respiratory failure and death of the neonate. Late presentation of AHD is exceedingly rare. Our patient is the oldest reported case in the English-language literature.

AHD results from nondevelopment of the embryologic origins of the diaphragm, including the septum transversum, dorsal mesentery of the esophagus, innermost thoracic wall, and pleuroperitoneal membranes. It is left sided in 88.2% to 97% of cases.

Tzelepis et al. reported absence of the left hemidiaphragm in an asymptomatic adult. Although they stressed that the large pleuroperitoneal communication in such a defect allows for free movement of the intestines, making intestinal strangulation unlikely, the present case demonstrates that such a complication may still occur.

Of the three previously reported cases of “agenesis” of the hemidiaphragm in adults, two were left sided and were repaired by suturing a synthetic mesh to diaphragmatic remnants; given the presence of these remnants, these cases therefore did not represent true agenesis. The remaining case, which was right sided, was managed conservatively without adequate investigation to confirm true agenesis.

In the elective setting, repair of the diaphragmatic defect with prosthetic mesh is optimal. In this case, however, the presence of large bowel obstruction and the need for colonic resection increased the risk of infection of any prosthesis and was therefore not performed. Resection of the mobile transverse colon reduces the likelihood of hernia recurrence by reducing redundant colon. In the event of future symptoms, elective diaphragmatic mesh repair would be considered.

Postoperative CT and MRI of our patient demonstrated absence of the left hemidiaphragm, and intraoperative examination failed to find any diaphragmatic remnants, thus suggesting that this case is the first reported case of an adult survivor with complete true AHD.

REFERENCES

6 Travaline JM, Cordova F. Agenesis of the diaphragm. Am J Med 1996; 100:585

Conservative Treatment of Postsurgical Lymphatic Leaks With Somatostatin-14*

Jean-Marie Collard, MD; Pierre-François Laterre, MD; Freddy Boemer, MD; Marc Reynaert, MD; and Robert Ponlot, MD

Key words: fistula; somatostatin; thoracic duct

Abbreviation: TPN = total parenteral nutrition

Successful management of lymphatic leaks by continuous IV administration of somatostatin was first reported by Ulibarri and coworkers in Spain,1 and more

*From the Departments of Surgery (Drs. Collard and Ponlot) and Intensive Care (Drs. Laterre and Reynaert), St-Luc Academic Hospital, Brussels, and Department of Internal Medicine (Dr. Boemer), Queen Astrid Hospital, Malmedy, Belgium.

Manuscript received March 23, 1999; revision accepted September 7, 1999.

Correspondence to: J.M. Collard, MD, Department of Surgery, St-Luc Academic Hospital, Hippocrates Avenue, 10, B–1200 Brussels, Belgium; e-mail: Collard@ehir.ucl.ac.be

Copyright 

Figure 2. Chest radiographs 1 year postoperative. Top: posteroanterior chest radiograph demonstrates the expanded pulmonary volume and the stomach margins (arrow) in the left hemithorax. Bottom: lateral chest radiograph demonstrates the gastric margins in the left hemithorax.
recently by authors from Italy and Switzerland. The present article reports the clinical history of two patients in whom postsurgical lymphatic leak was successfully treated after the administration of either somatostatin-14 alone (case 1) or combined somatostatin-14 and total parenteral nutrition (TPN; case 2). Although further pathophysiologic studies are needed for the elucidation of its mechanisms of action, somatostatin-14 seems to be an intriguing therapy against postsurgical lymphatic leaks that may make potentially risky transthoracic reoperation unnecessary.

REPORT OF CASES

Case 1

A 54-year-old man with multiple mediastinal masses had been admitted for pretherapeutic check-ups in the Department of Internal Medicine. He underwent conventional mediastinoscopy through a small suprasternal incision in the neck. Several macrobiopsies of the masses were taken for histologic examination. A polyvinyl drain was left in the anterosuperior mediastinum for fasting and being maintained on TPN, injury to the thoracic duct was suspected as drainage outflow progressively increased (Fig 1). Therefore, it was decided to clip the lower segment of the thoracic duct thoracoscopically. Under general anesthesia, four trocars were inserted through the right chest wall, one for the optical device, one for a lung retractor, and two for working instruments or a clip applier. The thoracic duct was identified, lying between the right azygous vein and the descending aorta just in front of the spine. After division of the duct between metal clamps, a chest tube was left in the right chest cavity for drainage. The day after, fluid outflow through the cervical drain dropped from 940 to 330 mL/d without any further decrease during the next days (350 mL/d 7 days after the thoracoscopy). Persistent weeping was ascribed to injury to paraesophageal lymphatic vessels draining pulmonary-in-origin lymphatic fluid. Therefore, somatostatin-14 (UCB S.A. Pharma sector; Braine-L’Alleud, Belgium) was administered by continuous IV infusion at the dosage of 6 mg/d. Cervical drain outflow dropped from 400 to 90 mL/d within 36 h, and it reduced progressively to 3 mL/d within a 5-day period. Somatostatin therapy was maintained for an additional 6-day period, ie, at the dosage of 6 mg/d for 4 days and of 3 mg/d for the last 2 days, after which the drug was discontinued, and the cervical drain was removed. Measurement of the glucose blood level four times per day did not show any major disturbance in the glucose regulatory mechanisms.

Case 2

A 56-year-old man with cancer arising in Barrett’s esophagus underwent transthoracic esophagectomy and gastric pull-up to the neck. Esophagectomy included resection of the esophageal tube en bloc with the vagus nerves, the thoracic duct, and the right azygous vein. The thoracic duct was firmly ligated just above the cisterna chyli. The immediate postoperative course was uneventful, oral feeding was started on postoperative day 9, and the patient was discharged home 5 days later. Three weeks after discharge, he was readmitted to the hospital for progressive respiratory insufficiency characterized by shortness of breath and dyspnea. Standard chest radiograph showed an extensive left pleural effusion that justified transthoracic needle aspiration. Thirteen hundred milliliters of lipid- and lymphocyte-rich milky fluid was aspirated with subsequent symptomatic relief and discharge. However, routine chest radiographs performed a few days later showed recurrence of the left pleural effusion, which testified to the existence of a chronic lymphatic weeping, probably originating from accessory lymphatic pathways in the posterior mediastinum. After transthoracic needle aspiration of the recurrent effusion, somatostatin-14 was administered by continuous IV infusion at 6 mg/d, while the patient was maintained in the fasting state and given TPN. Somatostatin therapy was maintained for a 14-day period, after which oral feeding was started again without subsequent recurrence of the pleural effusion. No adverse effect of the drug was observed during or after treatment. Six months after the initial operation, the patient takes a normal diet without experiencing any dysphagia, and the standard chest radiograph is unremarkable.

DISCUSSION

Leak of lymphatic fluid is a rare but embarrassing complication of thoracic or cervical surgery. It is indeed difficult to treat, and it results most of the time in an expensive, prolonged in-hospital stay. Moreover, we know from continuous drainage of the thoracic duct in organ transplantation that prolonged loss of proteins and lymphocytes alters immunologic defense mechanisms, a condition that may predispose critically ill patients to systemic infection.

Various therapeutic methods have been used against such lymphatic fluid weepings. Since the first ligation of the thoracic duct at the cervical level by Cushing11 > 100 years ago and at the thoracic level by Lampson12 50 years later, the classic method consists of repeat thoracotomy for ligation of the lymphatic vessel together with all fatty and fibrous tissues lying between the right azygous vein and the aorta just above the diaphragm. More recently, right thoracoscopy has been shown to be an elegant technical modality for approaching the thoracic duct, allowing division of the vessel between metal clips under videoscopic control. A few years ago, Japanese surgeons reported successful management of a thoracic duct injury after pneumonectomy by application of fibrin glue on the site of laceration through the working channel of a bronchosfiberscope that had been inserted into the pleural cavity through a 28F chest tube.15 Tale-induced pleurodesis is an alternative to repeat thoracotomy.16 Likewise, leakage from the cervical segment of the thoracic duct has successfully been treated by local injection of tetracycline hydrochloride.17 In another report, mediastinal radiotherapy has been shown to be effective in resolving chylothorax secondary to neoplastic obstruction of the superior vena cava and thoracic duct.18 Recently, IV administration of an α-adrenergic drug such as etilefrin hydrochloride has been shown to be capable of drying up intra-abdominal lymphatic weeping after resection of the esophagus en bloc with the thoracic duct.19

Case 1 of the current article confirms data from other authors1–3 that the administration of somatostatin-14 may be very effective against thoracic surgery-related lymphatic leaks that do not respond to TPN alone. Within a few days indeed, lymphatic fluid outflow reduced dramatically, and no rebound effect was observed while the dose of the drug was reduced progressively until complete discontinuation. In case 2, the role of somatostatin-14 in drying up lymphatic leakage was less unequivocal than in
case 1, inasmuch as TPN, which was started as the same time as somatostatin-14 therapy was initiated, might have contributed to reducing lymphatic fluid outflow.

Little is known about the mechanism of action of somatostatin regarding chyle production on the one hand and intraluminal pressure in the lymphatic system on the other. Although somatostatin reduces intestinal absorption of fats\textsuperscript{20,21} as well as triglyceride concentrations in thoracic duct lymph\textsuperscript{22}, the question of whether the drug acts on nutrient absorption by direct interference with the transport process in the gut wall, or indirectly by lowering intestinal blood flow\textsuperscript{23} or motility\textsuperscript{24} remains unanswered to date. Nakabayashi and coworkers\textsuperscript{22} have shown in animal studies that somatostatin infusion through either the portal or the femoral vein exerts an attenuating effect on thoracic duct lymph flow, probably in relation to changes in the splanchnic lymph dynamics. The same authors also showed that the attenuating effect was abolished by truncal vagotomy. However, the fact that our second patient had had truncal vagotomy contradicts the second observation of this group.

In any case, although further pathophysiologic studies are needed for the elucidation of the mechanism of action of the drug, somatostatin-14 infusion appears to be an intriguing alternative method for drying up postsurgical lymphatic leaks. Because of the poor general condition of most of the patients concerned, this medical treatment should be attempted before reopening of the chest for lymphostasis and, in any case, in the presence of residual lymphatic weeping after primary ligation of the thoracic duct. However, because somatostatin modulates the blood glucose regulatory system\textsuperscript{25,26} we recommend monitoring of blood glucose every 6 h during treatment. In addition, the standard dosage of 6 mg/d must be reduced to 3 mg/d during at least a 48-h period to prevent any rebound effect.

References


Giant Cell Myocarditis Responding to Immunosuppressive Therapy*

Andrea Frustaci, MD, FCCP; Cristina Chimenti, MD; Maurizio Pieroni, MD; and Nicola Gentiloni, MD

An unusual case of giant cell myocarditis presenting with cardiogenic shock that dramatically responded to conventional dose of steroids and azathioprine is reported. Cardiac recovery was rapid, complete (left ventricular ejection fraction rose to 55% from 10%), and was accompanied by the disappearance of the inflammatory infiltrates including giant cells in the control endomyocardial biopsy. Maintenance of the recovery at 16 months of follow-up on a low dose of azathioprine suggests that giant cell myocarditis might be a heterogeneous disease having either a negative untreatable trend necessitating cardiac transplantation, or a curable substrate responding to immunosuppressive drugs.

(CHEST 2000; 117:905–907)

Key words: giant cell myocarditis; heart failure; immunosuppressive therapy

Abbreviations: EF = ejection fraction; LV = left ventricular; LVEF = left ventricular ejection fraction

Giant cell myocarditis is a progressive disease of unknown cause that has been reported in subjects aged 6 months to 70 years. This entity has a grim prognosis, as conventional immunosuppressive therapy is usually ineffective and affected patients die in a short time unless a heart transplant is rapidly done. Causes of death include progressive heart failure and sudden death. The authors report the unusual case of a young patient presenting with cardiogenic shock due to giant cell myocarditis that responded dramatically to a combination of steroids and azathioprine maintaining a complete recovery for a long period.

CASE REPORT

A 23-year-old girl was admitted because of rapidly worsening shortness of breath. Two months before, she had a flu-like syndrome. On admission, she was stable. Tachycardia was present (heart rate, 130 beats/min) with a gallop rhythm; BP was 130/80 mm Hg. The ECG showed low voltages, left anterior bundle branch block, and diffuse ST-T wave abnormalities. Chest radiograph was normal. Routine laboratory tests including glucose, creatinine, BUN, serum electrolytes, and urinalysis were within normal limits. Erythrocyte sedimentation rate was 108 mm/h at the first hour. C-reactive protein was 187.5 mg/L (normal value, < 5 mg/L). Blood cell count revealed a leukocytosis (13.4 × 10⁹) with 80% neutrophils. The echocardiogram showed normal cardiac dimensions and mild reduction of left ventricular [LV] contractility (ejection fraction [EF] of 45%). Three days later, the patient had a clinical deterioration with worsening of dyspnea and severe hypotension (BP, 70/40 mm Hg). A new echocardiogram revealed LV dilatation of 61 mm, with marked impairment of LV contractility (EF, 10%). The patient underwent an invasive cardiac study including cardiac catheterization, LV (Fig 1, left) and coronary angiography, and LV endomyocardial biopsy with extraction of three good-sized tissue samples that were processed for histology following standard techniques and stains (hematoxylin-eosin, Miller’s elastic Van Gieson, and Masson’s trichrome). Coronary angiography was normal.

*From the Departments of Cardiology (Drs. Frustaci, Chimenti, and Pieroni) and Internal Medicine (Dr. Gentiloni), Catholic University, Rome, Italy.

Manuscript received May 10, 1999; revision accepted September 21, 1999.

Correspondence to: Andrea Frustaci, MD, FCCP, Cardiology Institute, Catholic University, Largo Gemelli, 800168 Roma, Italy.