anti-IL-2 receptor activity (Ortho Diagnostic Systems). Binding of various MoAbs was assessed with indirect immunofluorescence using FITC-conjugated rabbit antimouse immunoglobulin (DAKO) as second antibody. Cells prepared the same way, but without the addition of the first layer antibody, served as negative controls. The anti-lgM antibody (Heintel) was used as directly conjugated with FITC. Analysis of 10,000 lymphoid cells was done using a flow cytometer (FACStar, Becton-Dickinson).

**DNA Extraction and Southern Blotting**

DNA was extracted from the peripheral blood mononuclear cells as described previously. The configuration of the T-cell receptor chain genes was investigated by probing EcoRI and Hind III DNA digests with a C B-probe M131B1O8B1. The configuration of the immunoglobulin (Ig) heavy chain locus was analyzed by the Ig heavy chain probe M12C76B51A after digestion of DNA with EcoRI and Hind III restriction endonucleases. All DNA probes originated from the same source (T.H. Rabbits, Cambridge, United Kingdom).

**RESULTS**

The patient was found to suffer from T-cell CLL (CD4+, 98 percent) both by cell surface marker as well as DNA rearrangement studies. Analysis of genomic DNA of peripheral blood mononuclear cells showed the presence of a monoclonal population of T cells, the T-cell receptor beta-chain gene being rearranged on both alleles while the Ig heavy chain genes were in germline configuration.

Cell surface marker and DNA studies on the mononuclear cells isolated from the pleural fluid showed this population of lymphoid cells to carry the same cell surface marker and DNA rearrangement characteristics as the cells of the peripheral blood. They were mature T cells (CD2+), 69 percent; CD3+, 78 percent; CD4+, 79 percent; CD8+, 16.9 percent; CD5, 34.9 percent), lacked the immature T-cell marker CD1, while B cells represented just a minority of the cell population (CD19+, 0.5 percent; sIgM+, 8.4 percent). Results of DNA studies showing a monoclonal population of T cells both in the pleural fluid and in the peripheral blood are shown in Figure 1.

**DISCUSSION**

Patients with malignant lymphoma not infrequently develop benign lymphoid-rich effusions. To help the differential diagnosis of malignant from benign pleural effusions, a computerized interactive morphometry system has been developed. While the predictive value of this expert system exceeded 90 percent in the diagnosis of effusions accompanying malignant lymphoma or benign pleural lymphocytosis, nevertheless the system was unsuitable for the diagnosis of malignancy in effusions from patients with CLL. Lymphoproliferative disease was ascertained by molecular analysis of the mononuclear cells of the pleural effusion in a case of non-Hodgkin’s lymphoma without palpable lymph nodes. The results of our DNA studies performed simultaneously on the mononuclear cells from the peripheral blood and from the pleural fluid led us to the conclusion that the neoplastic cells in the pleural fluid had the same clonal origin as the leukemic process itself.

In conclusion, cell surface marker and DNA studies of the pleural fluid mononuclear cells appear to be appropriate for the definition of the cellular origin of the effusion in patients with CLL.

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**Atrial-Esophageal Fistula Shown by Transthoracic Echocardiogram**

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Nontraumatic atrial-esophageal fistula is a catastrophic problem usually diagnosed postmortem and almost invariably fatal. We report the first case of a patient in whom the diagnosis of atrial-esophageal fistula was made from transthoracic echocardiography antemortem. Echocardiography showed multiple microbubbles in the left atrium and ventricle emanating from the posterior aspect of the left atrium adjacent to the pulmonary veins. The literature is reviewed and the significance of the case and the echocardiogram is discussed.

(Ches 1994; 106:1255-58)

**Key words:** esophageal ulcer, gastroesophageal-atrial fistula

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GI=gastrointestinal
Nontraumatic atrial-esophageal fistula is a catastrophic problem usually diagnosed postmortem and almost invariably fatal. Only 19 cases have been reported. In most cases, it is seen in the setting of chronic esophagitis or esophageal cancer, and the fistula has resulted from an ulcer penetrating the left atrium. Patients usually present with hematemesis and neurologic symptoms. We report the first case in which the antemortem diagnosis was made by a transthoracic echocardiogram and confirmed at autopsy.

**Case Report**

A 64-year-old white woman was admitted with fever and altered mental status after two syncopal episodes. Her cardiac history was significant for multiple myocardial infarctions. She had quadruple coronary artery bypass graft surgery 12 years ago and was admitted to the hospital for exacerbation of congestive heart failure. Two months before admission, she was seen by a gastroenterologist for symptoms of dysphagia and odynophagia. Upper gastrointestinal (GI) endoscopy performed at that time showed a large, circumferential, necrotic rock-hard ulcer in the middle portion of the esophagus, grossly suggestive of carcinoma, although biopsy showed dysplastic squamous epithelial cells with no definitive malignant cells. There was no recent history of GI bleeding. Further work up was not pursued because of poor surgical risks from her severe heart disease. She was treated with anticoagulants, digoxin, furosemide, and enalapril.

Physical examination revealed a middle-aged woman who was in no acute distress. Her vital signs showed blood pressure of 100/60, heart rate of 90, respiratory rate of 16, and a temperature of 39.6°C. She had signs of mild congestive heart failure. Her abdomen was flat and nontender. There was no organomegaly. Neurologic evaluation was remarkable for partial orientation (to her name and city only), her right eye drifted to the right, and she had generalized weakness. The initial hemoglobin was 13.3 g/dl, leukocyte count of 27.2X10⁹, with 16 percent bands and toxic granulation noted. Digoxin level was 16 ng/ml. Prothrombin time was 17.3 s, chest x-ray film showed marked cardiomegaly with mild vascular redistribution. Her ECG showed sinus rhythm with frequent ectopies and evidence of old lateral infarction.

**Hospital Course**

She was administered broad spectrum antibiotics after blood cultures were obtained. These later grew mixed organisms including *Streptococcus viridans*, Lactobacillus, and yeast. Twelve hours after admission, she developed atrial fibrillation, became deeply comatose, and was intubated. Computed tomography of the head revealed small amounts of air in the brain parenchyma.

Echocardiography was performed on an ultrasound machine (Sonos 1000, Hewlett-Packard) using a 2.5 mHz transducer. Echocardiogram revealed evidence of a cardiomyopathy with multiple wall motion abnormality and ejection fraction of 25 percent. The orientation of cardiac chambers and great vessels were normal. The left atrium appeared enlarged. Microbubbles could be seen intermittently in the left atrium and left ventricle but not in the right-sided chambers. There was no evidence of right-to-left shunting shown by color flow Doppler echocardiography. There was no pericardial effusion or pericardial thickening noted. Figure 1 (top, middle, bottom) shows the echocardiographic findings. There were multiple microbubbles in the left atrium and left ventricle appearing to emanate periodically from the posterior aspect of the left atrium adjacent to the pulmonary veins. The figures which are still frames recorded over short-time sequences, from the apical window show the episodic appearance of masses of bubbles. The transducer position is held constant. There was no spontaneous contrast seen in the left atrium or left ventricle. The right-sided chambers were free of bubbles. The appearance of the bubbles were similar to those seen when agitated saline solution or albumin is injected during contrast echocardiography. Microbubbles, similar in appearance, can also be seen in patients at the termination of cardiopulmonary bypass imaged with transesophageal echocardiography.

The diagnosis of atrial-esophageal fistula was based on the history of the esophageal ulcer and the microbubbles entering the left atrium. Given the patient’s condition and family’s wishes, no further life support measures were performed and she rapidly deteriorated and died.

At autopsy, the findings were those of a large ulcer (3 cm) arising on the mid- to low-anterior wall of the esophagus, associated with a patent fistula tract extending into the left atrium. No malignancy was identified. There was severe triple vessel coronary artery disease with total or near total occlusion of all three vessels. The bypass grafts were severely diseased as well with recent and old mural thrombi and multiple stenotic lesions. There was also evidence of several areas of myocardial infarction involving the posterior, apical, and septal regions. About 500 cc of fresh blood was found in the stomach and duodenum. There was also evidence of acute and recent cerebral infarction involving the right frontal, parietal, and occipital lobes.

**Discussion**

Acquired nontraumatic atrial-esophageal fistula is a rare condition. The first case of such was reported in 1874.¹ Snyder et al² reviewed the literature in 1988 including 16 previously reported cases and 2 cases they identified. They found that the formation of the atrial-esophageal fistula usually involves chronic inflammation with obliteration of the pericardial space with fibrosis. The adherence of the esophagus and the myocardium, therefore, provides the potential for fistula development. The majority of cases were the result of chronic reflux (56 percent) and were related to benign esophageal ulcers. Sixty percent had chronic strictures. Seventeen percent of the cases were the result of esophageal cancer. Thirty-three percent had previous surgery and 11 percent had previous radiation. Significant left atrial enlargement with compression on the esophagus is probably contributory in two patients without esophageal symptoms. Both of these patients were taking sustained release potassium chloride tablets, with tablet retention and erosion in the esophagus proposed as a possible cause.³⁻⁴ Lehman et al⁵ found unexplained atrial fibrillation to be very common (up to 43 percent). Chest and abdominal pain are also common accompanying symptoms.

Only two cases in the literature were diagnosed before death. Flodmark⁴ reported a case where a gas-fluid level was seen in the left atrium on a chest x-ray film. Lehmann
esophagus, therefore, would permit transfer of contents between the two cavities. The diagnosis is suggested by the clinical triad of hematemesis, acute neurologic signs, and chronic dysphagia.\(^2\) Neurologic abnormalities, representing cerebral emboli, are the most frequent presenting symptoms.

There have been two other articles reviewing pathologic esophageal to cardiovascular connections.\(^ {10,11} \) Miller et al\(^10 \) reported a case of pyopneumopericardium secondary to an esophageal pericardial fistula. Echocardiography showed microbubbles within the pericardial space and a large pericardial effusion. Rapid medical and surgical intervention resulted in survival. Aortoesophageal fistulas can also accompany esophageal carcinoma or ulceration, although the most common clinical presentation is gastrointestinal hemorrhage because of the high pressure within the aorta.

**Unique Features**

This is the first case that the diagnosis was shown by a transthoracic echocardiogram and also one of the three cases so far which an antemortem diagnosis was made. The finding of microbubbles in the left heart chambers without involvement of the right heart chambers made the diagnosis of right-to-left intracardiac shunt unlikely as a cause. A diagnosis of atrial-esophageal fistula was, therefore, suggested, although the lungs and pulmonary veins could not be excluded as a source before autopsy. The finding of air in the brain parenchyma in a patient without history of head trauma or surgery was very unusual and could be explained by air in the left heart chambers. The patient’s history of esophageal ulcer, change in mental status, and evidence of sepsis are common clinical presentations. The use of sustained release potassium chloride tablets and the previous cardiac surgery could have made the formation of the fistula more likely.

This case of fatal atrial-esophageal fistula shows the use of echocardiogram in making the diagnosis. The echocardiogram showed the fistulous connection between the GI tract and the heart. The microbubbles emanating from the posterior portion of the left atrium (LA). The “m” identifies the microbubbles in the left ventricle (LV). The arrows in the center and bottom figures locate the initial appearance site for the bubbles.

et al\(^5\) reported a case of right atrial-esophageal fistula which was confirmed by a chest computed tomogram. His was the only case that had a nonfatal outcome.

The principal pathophysiologic process causing illness is interchange of esophageal and atrial contents, leading to both gastrointestinal hemorrhage and seeding of the circulation with food particles as reported by Bell et al.\(^6\) Khan et al\(^7\) found that resting esophageal pressure in asymptomatic women >60 years old was 9.4 ± 1.8 cm of water. The close pressure range between the atrium and

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Liposarcoma of the Posterior Mediastinum in a Child*


Liposarcoma is rare in children and rarely occurs in the posterior mediastinum in any age group. A massive intrathoracic tumor in a 17-year-old young man was a diagnostic dilemma; preoperative radiographic evaluation and biopsy led us to believe it was a teratoma. At operation, a poorly differentiated myxoid liposarcoma originating from the posterior mediastinum was found and excised. To our knowledge, this is the first liposarcoma of the posterior mediastinum reported in a patient less than 18 years old. (Chest 1994; 106:1258-59)

The following case demonstrates a rare occurrence of liposarcoma in a 17-year-old man.

CASE REPORT

A 17-year-old man presented to the emergency room with complaints of right-sided chest pain and shortness of breath for 1 month. He had no history of tobacco, alcohol, or drug use, and had no known contact with an active tuberculosis patient. On physical examination, the right side of the thorax was enlarged; markedly decreased breath sounds and dullness to percussion were noted on the right side of the chest. A chest x-ray film showed complete opacification of the right hemithorax with mediastinal shift to the left.

A computed tomography (CT) scan showed a huge mass involving the entire right hemithorax and the mediastinum, with elements of varying soft tissue densities and dense central calcification (Fig 2). A CT scan of the abdomen and pelvis did not disclose any abnormalities. The tumor was hypovascular as evidenced by arteriography, and displaced, but did not involve the esophagus as shown by barium swallow. Bronchoscopy showed extrinsic compression of the right main stem bronchus, but no endobronchial lesions. Transbronchial biopsy and bronchial washing did not show malignant cells, granulomas or acid-fast bacilli, or fungi. A CT-guided fine needle biopsy and large core needle biopsy showed only striated muscle and mature adipose tissue on histologic examination. Serum α-fetoprotein and chorionic gonadotropin levels were not elevated.

The preoperative diagnosis of teratoma of the posterior mediastinum was made, and the patient underwent exploration via a right thoracoabdominal incision. A myxoid liposarcoma (90X14X14 cm) arising from the inferior aspect of the posterior mediastinum was found. Despite complete excision of the tumor and exenteration of all nonvital tissue from the posterior mediastinum, a microscopically clear margin could not be obtained. Postoperatively, the patient received three cycles of chemotherapy (doxorubicin, 20 mg/day for 3 days; dacarbazine (DTIC) 450 mg/day for 3 days; ifosfamide and mesna 2,400 mg/day for 5 days). Despite these efforts, the patient died of recurrent tumor 9 months afterward.

DISCUSSION

While liposarcoma is a relatively common soft tissue neoplasm in adults, it rarely occurs in young people. Of 2,500 cases coded as liposarcoma at the Armed Forces Institute of Pathology, only 17 (0.7 percent) occurred in those...