Tracheal Bronchus Associated With Lung Cancer*  
A Case Report

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Tracheal bronchus is a rarely found congenital bronchial anomaly. It usually originates from the right lateral wall of the trachea at the level < 2 cm above the tracheal bifurcation. The patients usually are asymptomatic, but some may experience recurrent pneumonia, chronic bronchitis, or bronchiectasis. It is very rare for a malignant tumor to grow from this aberrant bronchus. There are only four cases of lung cancer developing from the tracheal bronchus reported in the world literature, and we present a fifth case.

Key words: lung cancer; tracheal bronchus

The normal right upper lobe bronchus trifurcates into the apical, posterior, and anterior segments. The occasionally seen tracheal bronchus first described by Chiari in 1889 usually arises from the right lateral wall of the trachea less than 2 cm above the major carina. It could be displaced or supernumerary, depending on the numbers of segmental bronchi of the anatomical right upper lobe bronchus. If the anatomical right upper lobe bronchus bifurcates, the tracheal bronchus is defined as displaced; if it trifurcates, it is defined as supernumerary. Most cases of tracheal bronchi are asymptomatic, and detected only incidentally on bronchoscopy or radiologic examination. We found only four case reports of lung cancer that developed from the tracheal bronchus in the world literature. This report is the first case reported in Taiwan and the fifth in the world.

Case Report

A 72-year-old man was admitted to the chest ward with the chief problem of an incidental abnormal finding on a chest radiograph during a routine physical examination. He was a smoker who smoked nearly two thirds of a pack of cigarettes per day for about 50 years. The initial chest radiograph showed a patchy consolidation in the right upper lung field (Fig 1). CT disclosed a 4.5 × 3-cm noncalcified soft-tissue mass in the medial aspect of the right upper lung, surrounded by pulmonary infiltration (Fig 2). A right-sided tracheal bronchus was also seen (Fig 3). Bronchoscopy was performed and a right-sided tracheal bronchus was found; it originated from the lateral wall of the lower trachea about 2 cm above the tracheal bifurcation (Fig 4). The right upper lobar bronchus had two segmental bronchi. A tumor growth was found in the tracheal bronchus and was causing nearly total occlusion of it. Both biopsy and cytologic brushing of the tumor were performed, and squamous cell carcinoma was confirmed. Tumor staging was carried out and the clinical stage was T2N0M0.

Right upper lobe lobectomy and radical lymph node dissection were performed on January 9, 1998. The tumor was 4 × 4 × 3.5 cm in size and located in the apical region, with surrounding pneumonic change. The apical segment was supplied by the tracheal bronchus, and the posterior and anterior segments were supplied by the right upper lobar bronchus. The tracheal bronchus was cut and sutured using the open method, and the stump was covered with a pleural flap.

Figure 1. Chest radiograph shows a patchy consolidation in the right upper lobe area.
On pathologic examination, the tumor revealed moderately differentiated squamous cell carcinoma. The tumor invaded but did not penetrate through the visceral pleura, and the bronchial cut ends were free from tumor involvement. All lymph nodes surveyed showed reactive hyperplasia without metastatic tumor; therefore, the pathologic staging was pT2N0M0 (stage Ib).

The patient was well 12 months after the operation and has had no evidence of recurrence.

**DISCUSSION**

Tracheal bronchus is an aberrant bronchus usually originating from the right lateral wall of the trachea, with an incidence ranging from 0.1% to 2%.3,4 The term is used to designate any bronchus originating from the trachea above the level of the main carina. Tracheal bronchus is a normal finding in sheep, swine, cattle, camels, goats, and giraffes, but a rare and usually incidental finding in humans.5 It can develop from any point above the main carina, but is usually within the 2 cm range. Its diameter ranges from 0.5 to 1.0 cm; its length ranges from 0.6 to 2.0 cm.6

Tracheal bronchus is of no clinical significance, although some reports indicate that it is associated with recurrent pneumonia, chronic bronchitis, and bronchiectasis.5,7 This may be caused by relatively poor local drainage of the involved bronchi. In these cases, surgical excision of the involved segment may be indicated.8,9 Other congenital anomalies, such as laryngeal web, rib and vertebra anomalies, tracheal stenosis, and congenital heart disease, are occasionally associated with this condition. Awareness of this condition may be important because the presence of a tracheal bronchus may complicate endotracheal intubation.7

Tracheal bronchus can be identified during bronchoscopic examination by the presence an ectopic opening from the tracheal wall. CT is the newest modality for evaluating tracheal bronchus. It can be easily diagnosed on CT as a small, round translucency posterolateral to the trachea.5,10

It is rare for a tumor to grow in a tracheal bronchus, and only four such case reports can be found in the literature. Uchikov11 in Bulgaria reported the first case of lung cancer that developed in an anomalous tracheal bronchus in 1974. In 1985, Moriya and colleagues12 reported the second case, a small cell lung cancer in Japan. The third case was reported by Calvet et al13 in a French journal in 1997. The fourth case, reported by Kim et al10 in Korea, was the first case treated with surgical resection. Our case is the first case reported in Taiwan and the fifth in the world. Our patient also underwent surgical resection and was free of recurrence after a 12-month follow-up.

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Continuous Intravenous Epoprostenol Therapy for Pulmonary Hypertension in Gaucher’s Disease*

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Gaucher’s disease is a rare disorder characterized by a deficiency of lysosomal $\beta$-glucosidase. Pulmonary hypertension, the etiology of which is unclear, has been reported to occur in association with Gaucher’s disease. We report the use of continuous intravenous epoprostenol (prostacyclin), which has been used to treat other forms of pulmonary hypertension, in a patient with pulmonary hypertension associated with Gaucher’s disease. Although its mechanism of action remains unknown, epoprostenol may be an effective form of therapy for chronic pulmonary hypertension due to a variety of conditions, one of which is Gaucher’s disease.

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Key words: epoprostenol; Gaucher’s disease; pulmonary hypertension

Abbreviations: PA = pulmonary artery; PPH = primary pulmonary hypertension; PVR = pulmonary vascular resistance

CASE REPORT

A 47-year-old white woman was diagnosed with type 1 Gaucher’s disease in 1958 at 8 years of age, presenting with hepatomegaly and bone disease. Pulmonary hypertension was diagnosed in 1990 when she developed progressive dyspnea. She was started on replacement therapy with aglucerase in 1991 with improvement of her symptoms and a marked reduction in liver size.

In 1993, she presented with progressive exertional dyspnea and chest pain, and right heart catheterization was performed (Table 1). Based on the absence of an acute response during a vasodilator trial, she was started on continuous IV infusion of epoprostenol and oral anticoagulants in August 1993. Right heart catheterization was repeated in March 1994 (Table 1).

In May 1995, a repeat right heart catheterization (Table 1) demonstrated that the mean pulmonary artery (PA) pressure had decreased from 34 to 25 mm Hg and the pulmonary vascular resistance (PVR) had decreased from 363 to 176 dynes.cm$^{-5}$.

During the catheterization, the epoprostenol infusion was discontinued, resulting in a prompt increase in PA pressure and a reduction in cardiac output. Hemodynamic measurements were also evaluated during exercise (Table 2).

Her most recent catheterization in May 1997 demonstrated a PA pressure of 45/12 mm Hg (mean, 23), a right atrial pressure of 2 mm Hg, and a cardiac output of approximately 9 L/min.


DISCUSSION

Lung involvement in Gaucher’s disease has been reported to occur in the following three distinct patterns:1 (1) interstitial infiltrates of Gaucher cells with associated fibrosis; (2) alveolar consolidation by Gaucher cells filling alveolar spaces; and (3) pulmonary hypertension. Several
possible mechanisms for the pulmonary hypertension have been suggested: (1) One possible mechanism is capillary plugging by Gaucher cells. Ross et al³ recovered Gaucher cells from a sample of pulmonary capillary blood aspirated from a PA catheter during balloon occlusion in the wedged position. (2) In another report, however, few Gaucher cells were found in the lungs, and the pathologic findings resembled those of primary pulmonary hypertension (PPH).³ (3) A clinical pattern similar to the pulmonary hypertension associated with liver disease has also been observed.⁴ Some authors have suggested that, in those patients without infiltration of the lung by Gaucher cells, the pulmonary hypertension may be related to either the possible presence of contaminants in the enzyme replacement therapy⁵ or to closure of intrapulmonary vascular dilatations (arterial-venous shunting) following the reduction of hepatomegaly by enzymatic treatment, resulting in increased blood flow through a pre-existing restricted pulmonary vascular bed.⁴ Using echocardiography to estimate pulmonary arterial systolic pressure, Elstein and colleagues⁶ found an unexpectedly high rate (7%) of plugging by Gaucher cells. Ross et al² recovered Gaucher cells from a sample of pulmonary capillary blood aspirated from a PA catheter during balloon occlusion in the wedged position. (2) In another report, however, few Gaucher cells were found in the lungs, and the pathologic findings resembled those of primary pulmonary hypertension (PPH).³ (3) A clinical pattern similar to the pulmonary hypertension associated with liver disease has also been observed.⁴ Some authors have suggested that, in those patients without infiltration of the lung by Gaucher cells, the pulmonary hypertension may be related to either the possible presence of contaminants in the enzyme replacement therapy⁵ or to closure of intrapulmonary vascular dilatations (arterial-venous shunting) following the reduction of hepatomegaly by enzymatic treatment, resulting in increased blood flow through a pre-existing restricted pulmonary vascular bed.⁴

In conclusion, the frequency of pulmonary hypertension complicating Gaucher’s disease, coupled with our observations that epoprostenol may be useful in treating pulmonary hypertension secondary to connective tissue diseases.⁸ However, there have been no reports on its use in patients with pulmonary hypertension secondary to Gaucher’s disease. The decision to use epoprostenol in our patient was based on her symptoms and the lack of a favorable acute response to vasodilators during catheterization.

The pulmonary vascular bed can be injured by various different stimuli that, in the susceptible host, result in the following characteristic pathologic findings that combine to produce increased PVR: smooth muscle cell hypertrophy, intimal proliferation, and in situ thrombosis. Therefore, irrespective of the cause of the pulmonary hypertension, be it PPH from anorexigens, portopulmonary hypertension, or pulmonary hypertension associated with systemic sclerosis, the pathologic injury pattern is indistinguishable and may be amenable to the beneficial effects of epoprostenol. In addition to its vasodilatory and antiplatelet properties, epoprostenol may have effects on vascular growth and remodeling, which could facilitate the restoration of endothelial-dependent functions that normally serve to maintain the low-resistance state of the pulmonary vascular bed. The response of our patient to epoprostenol supports the impression that epoprostenol can decrease PVR in patients unresponsive to acute vasodilatory challenges, regardless of the nature of the stimulus or the vascular injury pattern.

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this disorder, suggest that all patients (treated and untreated) may benefit from echocardiographic screening.

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Serial Scintigraphic Assessment of Iodine-123 Metaiodobenzylguanidine Lung Uptake in a Patient With High-Altitude Pulmonary Edema*

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Iodine-123 metaiodobenzylguanidine (123I-MIBG) can be considered an indicator of pulmonary endothelial cell function. Serial 123I-MIBG images of the chest were acquired in a patient with high-altitude pulmonary edema (HAPE). The initial evaluation was performed 7 days after admission. The lung to upper mediastinum ratios (LMRs) of 123I-MIBG uptake were 1.33 (for the right lung) and 1.12 (for the left lung). The second examination of 123I-MIBG lung uptake, which was performed 2 months later, showed LMRs of 1.39 (right lung) and 1.33 (left lung). We speculated that the decreased lung uptake of 123I-MIBG at the early recovery stage could reflect an impairment in pulmonary endothelial cell metabolic function in the development of HAPE.

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Key words: high-altitude pulmonary edema; hypoxia; iodine-123 metaiodobenzylguanidine; pulmonary endothelial cell function

Abbreviations: HAPE = high-altitude pulmonary edema; 123I-MIBG = iodine-123 metaiodobenzylguanidine; LL = left lung; LMR = lung to upper mediastinum ratio; MIBG = metaiodobenzylguanidine; RL = right lung

High-altitude pulmonary edema (HAPE) is a condition that occurs in individuals who fail to acclimatize to altitudes > 2,500 m. The mechanism of HAPE remains unknown. However, the pathophysiology of HAPE clearly involves increases in pulmonary vascular pressure and/or microvascular permeability.1–4 Radioiodinated metaiodobenzylguanidine (iodine-123 metaiodobenzylguanidine [123I-MIBG]), an analog of guanidine that shares many neuronal transport and storage mechanisms with norepinephrine, has been used to assess the sympathetic activity and innervation of the heart.5–8 Several clinical studies of the lung uptake of metaiodobenzylguanidine (MIBG) have been performed, and MIBG can be considered an indicator of pulmonary endothelial function.9–11 We demonstrated a case of HAPE that showed extremely reduced 123I-MIBG uptake in the lung at the early recovery stage and almost normalized lung uptake at complete recovery.

CASE REPORT
A 50-year-old Japanese male subject, who was healthy before climbing, arrived at the foot of the Japan Alps (Kamikouchi) at 1,500 m above sea level on July 21, 1998. The next morning, he started climbing Mt. Yari (3,180 m) and reached about 2,800 m above sea level. On the third day, while climbing Mt. Tsubakuro (2,690 m), he developed general fatigue, followed by headache, cough, and fever (38.0°C) in the afternoon. His condition progressively deteriorated during the night, and he was lethargic and comatose. The subject was rescued and admitted to our Shinshu University Hospital (660 m). On admission, he exhibited cyanosis and was unconscious. Coarse crackles were audible over both sides of the lung. Chest radiograph revealed patchy infiltrates in both lung fields. Arterial blood gas analysis (room air) showed PaO2 of 31.5 mm Hg, PaCO2 of 36.2 mm Hg and pH of 7.46. The patient was treated with adequate oxygen and fluid infusion. No other therapy was administered. His condition gradually improved, and the infiltration on chest radiograph disappeared within 2 weeks. Examinations using 123I-MIBG scintigraphy were performed 7 days and 2 months after admission.

123I-MIBG Scintigraphy and the Results
A dose of 111 mEq of commercially available 123I-MIBG (Daichi Radioisotopes Labs; Tokyo, Japan) was administered IV after the patient lay in bed undisturbed for 15 min. Anterior planar images were acquired 15 min after the injection of

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123I-MIBG and were stored in a 64 × 64 matrix using a scintillation camera (model ZLC 7500; Siemens; Solana, Sweden) equipped with a low-energy, general purpose collimator interfaced to a minicomputer (SCINTIPAC 2400; Shimazu; Kyoto, Japan). The energy window was set at the 159 keV photopeak of 123I. Regions of interest were placed over the upper mediastinum and the right lung (RL) and left lung (LL) in planar images. Referring to the isocount line, the regions of interest corresponding to the contours of the RL and LL were manually assigned. The total counts of each lung were measured, and a geometric mean was calculated as counts per pixel. To quantitate the degree of lung uptake of 123I-MIBG, the lung to upper mediastinum mean was calculated as counts per pixel. To quantitate the degree of lung uptake of 123I-MIBG, the lung to upper mediastinum mean was calculated as counts per pixel. To quantitate the degree of lung uptake of 123I-MIBG, the lung to upper mediastinum mean was calculated as counts per pixel. To quantitate the degree of lung uptake of 123I-MIBG, the lung to upper mediastinum mean was calculated as counts per pixel. To quantitate the degree of lung uptake of 123I-MIBG, the lung to upper mediastinum mean was calculated as counts per pixel.

The LMR values in the initial 123I-MIBG scintigraphy (Fig 1) were 1.33 (RL) and 1.12 (LL). The second examination of 123I-MIBG uptake showed LMR values of 1.39 (RL) and 1.33 (LL; Fig 2). Thus, decreased lung 123I-MIBG uptake was detected at the early recovery stage. In our laboratory, the mean ± SD LMR values in patients who had no pulmonary diseases and/or edema were 1.56 ± 0.16 (RL) and 1.28 ± 0.16 (LL).12

**Discussion**

Serial 123I-MIBG images of the lung were acquired in a patient with HAPE. We found that 123I-MIBG lung uptake was extremely reduced at the early recovery stage and was normalized at complete recovery. This finding suggested that impairment in pulmonary endothelial cell metabolic functions occurred during the development of HAPE.

Several studies on lung uptake of MIBG have been reported.7–14 MIBG has been demonstrated to represent active, sodium-dependent, saturable transport similar to that of norepinephrine.9 Lung uptake of MIBG is primarily in endothelial cells. Slosman et al initially evaluated lung uptake of MIBG using animal models, and demonstrated decreased MIBG lung extraction in bleomycin-induced9 and endotoxin-induced10 endothelial injury. Thus, MIBG lung extraction appears to be an indicator of pulmonary endothelial dysfunction.

There is now convincing evidence that the pulmonary hypertension plays an important role in the development of HAPE.1–4 Stress studies have demonstrated that the pulmonary endothelium exposed to high transmural pressures undergoes ultrastructural changes similar to those found in models of HAPE.15 In addition, an inflammatory process including cytokine activation may be involved, as shown by the findings of bronchoalveolar fluid.3,4 The findings suggested an alteration in pulmonary circulation involving endothelial cell function. The decreased lung MIBG uptake in the present case indicates pulmonary endothelial cell dysfunction in the development of HAPE.

Richalet et al14 examined lung MIBG uptake in normal volunteers after exposure to hypoxic conditions at high altitude (4,350 m) over an 8-day period. They found significant decreases in lung MIBG uptake compared to normal subjects exposed to low altitude. Furthermore, the results were independent of circulating catecholamine concentrations. This finding may also be related to regulations of pulmonary microcirculation during acclimatization to hypoxia.

Although MIBG lung uptake appears to be a marker of endothelial cell function, it is likely dependent on the pulmonary vascular surface that is available to the blood pool. In rats with an experimentally decreased pulmonary vascular surface, a linear relationship was found between reduced MIBG uptake and loss of the pulmonary vascular bed.9 The condition of unperfused or collapsed areas due to pulmonary edema has been elucidated in patients with HAPE. However, Glowniak et al8 and Dae et al8 showed that the lung uptake of 123I-MIBG was slightly increased in patients with congestive heart failure. Thus, although the mechanisms of pulmonary edema are different between HAPE and congestive heart failure, it is unknown how congestion or edema influences MIBG lung uptake. As shown in Fig 1, when the first examination using 123I-MIBG scintigraphy was performed, infiltration on plain chest radiograph was slight and PaO2 was 80.5 mm Hg (room air) in the present case, suggesting a small amount of remaining edema. Although massive pulmonary
edema may interfere with the evaluation of MIBG uptake, decreased lung uptake of $^{123}$I-MIBG in the present case could not be explained by reduced vascular surface area alone.

In conclusion, several factors may affect MIBG uptake in the lung. However, the present case suggests an impairment of endothelial cell metabolic function in the development of HAPE, as detectable by the noninvasive MIBG scintigraphic method described.

References


Contralateral Tension Pneumothorax Following Unilateral Chest Tube Drainage of Bilateral Pneumothoraces in a Heart-Lung Transplant Patient*

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Bilateral pneumothoraces can result from unilateral air leak after heart-lung transplantation. The recommended initial management of such patients is insertion of a unilateral chest tube. We report a patient who developed bilateral pneumothoraces after undergoing transbronchial biopsies 2 years after a heart-lung transplant. The unilateral chest tube failed to drain the contralateral pneumothorax and a tension pneumothorax developed. The advocated approach should be used with caution.

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Key words: heart-lung transplant; pneumothorax; transbronchial biopsy

After a heart-lung transplantation, the normal anterior barriers between the pleural cavities are interrupted.1 Simultaneous bilateral pneumothoraces from a unilateral air leak and rapid shifting of the accumulated air between the two hemithoraces, although rare, are potential complications of a persisting interpleural communication.2 It has been recommended that a single chest tube is sufficient for the drainage of bilateral pneumothoraces in this group of patients.3

We report one patient who developed bilateral pneumothoraces 2 years after her heart-lung transplant. The unilateral chest tube insertion failed to drain the contralateral pneumothorax and instead resulted in the radiologic picture of a tension pneumothorax.

Case Report

A 20-year-old white woman received a heart-lung transplant in October 1996 for congenital heart disease and resultant pulmonary hypertension. Postoperatively, she suffered an episode of acute rejection (grade A3), which responded to augmented steroid therapy. In the following 2 years, she had several episodes of respiratory tract infections from various organisms including cytomegalovirus, Staphylococcus aureus, and Cryptococcus. These were successfully treated with appropriate antimicrobial therapies. She had two episodes of grade A2 acute lung graft

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rejections during that period; both were treated successfully with augmented steroid therapy with restoration of baseline lung functions.

In September 1998, she developed dyspnea and her FEV1 dropped from her usual level of 2.2 L to 1.7 L. Bronchoscopy demonstrated bronchitic changes, and transbronchial biopsies showed no evidence of rejection. She developed a small right-sided pneumothorax following the procedure, which resolved spontaneously. Her FEV1 improved to 2.0 L after treatment with IV antibiotics.

Two months later, she developed a flu-like illness and presented again with dyspnea and a productive cough. Her FEV1 was again reduced to 1.6 L. Bronchoscopy performed on this occasion demonstrated inflamed mucosae and thick purulent secretions. A culture of bronchial washings grew a zygomycetes-like fungi with branching filaments as well as *Pseudomonas aeruginosa*. Multiple transbronchial biopsies taken from the right middle and lower lobes showed no evidence of acute rejection.

A chest radiograph performed 4 h after the bronchoscopy demonstrated small bilateral pneumothoraces (Fig 1). She was asymptomatic. A second radiograph 3 h later showed an increase in size of the bilateral pneumothoraces, and the patient developed tachypnea and desaturation on room air (from 95 to 89%). Since the transbronchial biopsies from the right lung were the most likely cause of the air leak, a chest tube was inserted into the right hemithorax.

This provided minimal relief of symptoms despite continual bubbling through the right chest tube, and suction at 30 cm H2O was initiated. A repeat chest radiograph 2 h afterwards showed complete resolution of the right pneumothorax (Fig 2). However, the left pneumothorax had increased in size with tension effect and significant mediastinal shift to the right. The patient remained tachypneic and hypoxic. Her BP was stable at 120/80, and her heart rate was 100/min. A second chest tube was inserted into the left hemithorax. A significant amount of air was released on insertion of the tube, and this was associated with dramatic relief of dyspnea.

A further chest radiograph the following morning revealed resolution of the left pneumothorax but reappearance of the right one. Both chest tubes remained patent. Over the next few days, there was radiologic evidence of a “shifting pneumothorax” between the two hemithoraces, which eventually resolved without further intervention. She received a 6-week course of amphotericin B for her fungal infection.

**Discussion**

The anterior pleural reflections are severed during heart-lung transplantation and occasionally after other cardiac surgery performed via median sternotomy. The resultant interpleural communication allows air or fluid to move between the pleural cavities. The development of bilateral pneumothoraces can be potentially life-threatening if it is under tension. Successful treatment with single chest tube drainage in some cases has led some to advocate this as the standard management of bilateral pneumothoraces in heart-lung transplant recipients.

In the case of this patient, the development of bilateral pneumothoraces following right-sided transbronchial biopsies and the subsequent “shifting pneumothoraces” confirmed that the communication between the two hemithoraces had persisted.

Single chest tube drainage failed to evacuate the accumulated air in the contralateral hemithorax and resulted in a potentially life-threatening tension pneumothorax. It is possible that the persistent anterior mediastinal communication was small and that the rapid drainage of the unilateral pneumothorax drew the adjacent scar tissue and adhesions together, thereby producing a valve-like effect and preventing drainage of the contralateral hemithorax.

Two small retrospective series reported that bilateral or shifting pneumothoraces occurred in 33 to 40% of heart-lung transplant recipients. In a retrospective study of heart-lung transplant patients after the immediate postoperative period, 6 out of 72 patients developed bilateral pneumothoraces. Two of the six patients were managed conservatively, two were unstable requiring urgent bilateral chest tube placement, and the remaining two were treated successfully with unilateral chest tube drainage.

Engeler et al reported that eight of 25 postoperative heart-lung transplant patients had either shifting pneumothoraces or decompression of pneumothoraces by contralateral chest tube, which is indicative of open communication of both hemithoraces. However, it is uncertain whether the anterior mediastinal defect would persist or

**Figure 1.** Bilateral pneumothoraces following transbronchial biopsies to right lower lobe (lung edges indicated by arrows).

**Figure 2.** A chest radiograph taken after the insertion of a right-sided chest tube shows an increase in size of the left pneumothorax with a mediastinal shift toward the right.
whether it would eventually be bridged by postoperative scar tissues. Our patient demonstrated that an interpleural defect may remain patent for > 2 years after transplantation and therefore, is likely to be permanent.

Physicians should be aware that in heart-lung transplant patients, communication between both hemithoraces may be permanent. Bilateral pneumothoraces often develop from a unilateral air leak. Although a single chest tube may be adequate, this approach should be taken with care because the contralateral pneumothorax may not always be adequately drained, and potentially life-threatening tension pneumothorax may result.

**REFERENCES**


**Right Pneumothorax Resulting From an Endocardial Screw-In Atrial Lead***

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Right pneumothorax complicated by an endocardial atrial lead has never been reported. Herein, we report on a small-build 79-year-old Taiwanese woman who suffered from complete AV block and underwent dual-chamber permanent pacemaker implantation. An active fixation screw-in atrial lead was chosen. The procedure was complicated by right pneumothorax associated with atrial perforation. Since simple measurements of the implantation parameters could not be used to predict the occurrence of perforation, great caution should be taken in to avoid overscrewing the atrial lead, and in scrutinizing the penetration depth of the helix of the lead under fluoroscopy.

*(CHEST 1999; 116:1133–1134)*

**Key words:** atrial perforation; permanent pacemaker; pneumothorax; screw-in atrial lead

**Abbreviations:** AV = atrioventricular; ECG = electrocardiogram

An endocardial atrial screw-in lead was introduced to the market in 1980. A few complications have been reported, such as lead dislodgment, lead fracture, acute pericarditis, and cardiac perforation. However, right pneumothorax caused by an endocardial atrial lead has never been reported. We present a case of right pneumothorax induced by an active screw-in atrial lead in a small-build Taiwanese woman.

**Figure 1.** Both of the ventricular and screw-in leads were inserted via the left subclavian vein.

**Case Report**

A 79-year-old woman with a body weight of 51 kg and body length of 140 cm was admitted to our hospital for dizziness and recurrent syncope on December 12, 1996, without any previous systemic disease. The ECG on admission showed a complete AV block with a junctional escape rhythm of 42 beats/min. The chest radiograph was normal. Electrophysiologic study showed intra-Hisian block. A permanent pacemaker implantation via left subclavian approach was performed subsequently. The ventricular lead (Selute 4285-59cm; Guidant; St. Paul, MN) was introduced first, followed by insertion of a screw-in atrial lead (CapSule atrial J 4568-53cm; Medtronic; Minneapolis, MN) smoothly without any difficulty (Fig 1). Atrial pacing threshold was 1.3 V, pacing impedance was 650 ohms, and P-wave amplitude was 3.3 mV. Ventricular pacing threshold was 0.4 V, pacing impedance was 960 ohms, and R-wave amplitude was 11.8 mV. A generator (Vigor DR model 1230; Guidant) was then connected, and the ECG showed normal dual-chamber pacing. Unfortunately, the patient started to complain of difficulty in breathing 4 h later. Auscultation revealed coarse crackles over the left chest with diminished breath sounds on the right. ECG revealed an intermittent loss of atrial sensing. Therefore, the pacemaker was immediately programmed to the VVIR pacing mode. Tube thoracotomy drainage was instituted immediately after the confirmation of right pneumothorax by chest radiograph (Fig 2). The symptoms improved substantially after the procedure. Transthoracic two-dimensional echocardiogram showed no evidence of significant pericardial effusion, and the atrial lead could not be well demonstrated. Transesophageal two-dimensional echocardiogram also revealed no evidence of extravasation. The cinefluoroscopy in 30° left anterior oblique view clearly showed the extrusion of the screw-in atrial lead from the right atrium to the right lung (Fig 3). An atrial lead replacement was done 2 days later. Under local anesthesia, the screw-in atrial lead was unscrewed first and withdrawn care-
fully. ECG and BP monitoring were continued. The pericardiosentesis kit and echocardiography equipment were all on standby. Another steroid eluting, tined, hook-on bipolar endocardial lead (CapSule 5524M-53cm; Medtronic) was implanted from the same venous access uneventfully. The atrial pacing threshold was 0.8 V, pacing impedance was 840 ohms, and P-wave amplitude was 3.3 mV. The pacing and sensing parameters of the previously inserted ventricular lead remained unchanged as before. The chest tube was removed on the ninth day of hospitalization. The patient was discharged the following day with ECG showing normal dual-chamber pacing.

**DISCUSSION**

Atrial lead implantation is needed for more physiologic pacing, but the complications are still more frequent as compared to a ventricular lead. Implanting an atrial lead is sometimes difficult, and dislodgment is another common problem. To overcome these problems, active fixation mechanisms have been developed. Greene et al described that acute pericarditis developed in 4.9% of the patients, especially when the lead was placed in the lateral and anterolateral wall of the right atrium. A few cases involving atrial wall perforation by an endocardial pacing lead have been described previously. A recent publication by the Mayo Clinic showed a 2.4% rate of acute lead-related complications, ie, perforation, microdislodgment, and pericarditis.

Pneumothorax is common during subclavian vein puncture. However, we present this case in which pneumothorax was induced by the endocardial atrial lead. Several risk factors can be attributed to the implantation technique and lead design, including overscrewing, distal stiffness, and penetration depth of the helix of the lead. Since the lead screw itself has no electrical conductance property, its main function is mechanical support and stability. It does not serve as a discriminator in identifying cases when perforation occurs. Therefore, the implantation parameters could not be used to predict the occurrence of perforation.

Great caution must be taken when implanting an atrial screw-in lead, particularly into the anatomically thin right atrial free wall. We should avoid overscrewing, especially when positioning the lead in the lateral or anterolateral wall, without compromising the lead stability. The patient reported herein was a Taiwanese woman having a relatively small build. Whether the right atrial wall is thinner in Orientals than in whites remains to be determined.

In conclusion, atrial screw-in lead placement can still cause right pneumothorax under the current design in an Oriental patient. Thus, for placement of an atrial active fixation screw-in lead, we strongly recommend using great caution to avoid overscrewing the lead, and limiting the penetration depth of the helix of the lead under fluoroscopy. However, most centers now prefer the hook-on atrial leads used in the right atrial appendages instead of the atrial screw-in leads.

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**FIGURE 2.** The posteroanterior view of the chest radiograph clearly shows right pneumothorax.

**FIGURE 3.** The fluoroscopy shows the extrusion of the helix of the atrial screw-in lead through the high lateral right atrial wall into the right lung (arrowhead).
Advanced Heart Block as a Manifestation of a Paraneoplastic Syndrome From Malignant Thymoma*

William H. Pentz, MD

Malignant thymoma is a rare tumor that is associated with paraneoplastic syndrome. Myocarditis as a paraneoplastic syndrome has been rarely described. Reported herein is a young male patient with malignant thymoma and myocarditis as part of a paraneoplastic syndrome. This resulted in high-degree heart block and an asystolic cardiac arrest despite placement of a permanent pacemaker.

(CHEST 1999; 116:1135–1136)

Key words: advanced heart block; paraneoplastic syndrome; malignant thymoma

Abbreviation: CPK = creatinine phosphokinase

The thymus gland is very important to the development of the immune system, but its role in adults is not well understood. Malignant thymoma is an unusual disease process that typically affects middle-age adults. It is associated with a variety of paraneoplastic syndromes. This case describes a young male patient with malignant thymoma who developed myocarditis as part of a paraneoplastic syndrome. This subsequently lead to high-degree heart block, which has not been reported to have occurred as part of paraneoplastic syndrome.

Case Report

The patient is a 17-year-old male who had been in previously good health until he presented to his local emergency department with a cough and upper respiratory tract symptoms. A chest radiograph revealed an anterior mediastinal mass. A CT scan revealed a large mass in the mediastinum. A thoracotomy was performed that revealed invasion of the mass into the right pleural space and evidence of pulmonary metastases. A biopsy was diagnostic for an invasive thymoma, mixed epithelial and lymphocytic type. The patient is a 17-year-old male who had been in previously good health until he presented to his local emergency department with a cough and upper respiratory tract symptoms. A chest radiograph revealed an anterior mediastinal mass. A CT scan revealed a large mass in the mediastinum. A thoracotomy was performed that revealed invasion of the mass into the right pleural space and evidence of pulmonary metastases. A biopsy was diagnostic for an invasive thymoma, mixed epithelial and lymphocytic type. The tumor was believed to be too extensive and invasive for resection. He was treated with six courses of VP-16, ifosfamide, and cisplatin. A follow-up CT scan revealed no change in the size of the mass.

Six months after completing chemotherapy, the patient was readmitted with cough, night sweats, and productive sputum, and he subsequently developed progressive respiratory distress that required a transfer to the ICU and intubation. He also developed transient heart block that resolved quickly and did not require temporary pacing. He had creatinine phosphokinase (CPK) elevations > 10,000 U/L. A muscle biopsy was performed and was consistent with an inflammatory myositis. The CPK elevations were believed to be due to myocarditis. The myositis and myocarditis were believed to be a paraneoplastic manifestation of the thymoma, and treatment with IV immunoglobulins and prednisone was initiated. Eventually, tracheostomy placement and transfer to a chronic ventilatory facility were required. He was successfully weaned from the ventilator at this facility; subsequently, he was maintained on prednisone and monthly chemotherapy with cyclophosphamide, adriamycin, and cisplatin.

Several months later the patient presented to the emergency department complaining of chest pain and dyspnea. A 12-lead ECG revealed anterolateral ST-segment elevations (see Fig 1) that were changed compared to prior ECGs. Emergency coronary angiography was performed and revealed normal coronary arteries. A subsequent echocardiogram revealed normal systolic performance of the left ventricle. Peak CPK was 10,870 U/L, with 9.7% CPK-MB. He was presumed to have a recurrence of his myocarditis and was started on high-dose steroid therapy. He then developed 2:1 heart block with a heart rate around 50 beats/min and a systolic BP of 100 to 120 mm Hg. As a precaution, the patient was transferred to the ICU for further monitoring. Several hours later, he progressed to 1:1 heart block (Fig 2), and a temporary pacemaker was placed with capture of the ventricle achieved at 0.5 MV. Subsequently, he was taken to the electrophysiology laboratory for permanent pacemaker placement. Because the procedure was believed to be risky, it was performed under general anesthesia. Initially, it was difficult to obtain adequate pacing thresholds, but eventually these were achieved and the patient was sent to recovery, where he developed ventricular tachycardia and was given a bolus of lidocaine. He then suddenly developed asystole with failure of the pacemaker to capture. A temporary pacemaker was reinserted but failed to capture, and the patient died.

Discussion

The thymus gland is very important to the development of the immune system. Immature lymphocytes migrate to the thymus and undergo maturation there to become capable of recognizing antigen and generating an immune response. The absence or abnormal development of the thymus plays a role in the DiGeorge syndrome and in severe combined immunodeficiency. The gland undergoes atrophy in the adult, and its functional significance in the adult is unknown. Because of the central role of the thymus in the immune system, thymomas commonly result in immunologic manifestations, as will be discussed later.

About half of all tumors occurring in the anterior mediastinum originate from the thymus gland. Thymomas are classified according to their histologic appearance, and they are staged on the basis of the disease extent and invasiveness. Most thymomas (from 70 to 80%) have a benign appearance histologically, and when they do not invade surrounding structures, they are classified as benign. The malignant thymomas are classified into two different groups: (1) those with a histologically benign appearance, but with invasion into surrounding structures; and (2) those with a histologically malignant appearance. The latter are termed thymic carcinomas. Most tumors occur in adults; the mean age of patients at diagnosis is 45 to 50 years old.1

Most patients with thymoma are asymptomatic, and the tumors are discovered incidentally, often as a mediastinal mass on a chest radiograph. The most common symptomatic presentation is due to a paraneoplastic syndrome. Few carcinomas are associated with such a high rate of para-
neoplastic syndrome. The most common is myasthenia gravis, which occurs in as many as half of all patients with thymoma. In many patients, the myasthenia improves with thymectomy. Other common manifestations are hypogammaglobulinemia, anemia, and a variety of autoimmune disorders. The patient in this case had myocarditis in association with thymoma, which is extremely unusual. In a series of 148 patients from the Mayo Clinic with thymoma, only 1 patient had myocarditis.3

The 10-year survival for patients with malignant thymoma ranges from 86 to 100% with stage I disease to 26 to 47% with stage IV disease. Stage I is a completely encapsulated mass; stage II refers to invasion into the capsule or surrounding fatty tissue. Stage III is the extension of the tumor into an adjacent organ such as the pericardium or great vessels, and stage IV refers to distant metastasis. Surgical resection, if possible, is the preferred method of treatment. Even with advanced stage of disease, surgical resection resulted in a 77% and 59% survival at 5 and 10 years, respectively, in one study.1 Patients with complete resection did better than those with only a partial resection. Radiation therapy as an adjunct to resection is recommended for more advanced disease. It has not been until more recently that chemotherapy had been studied to treat unresectable disease. One study found a complete response rate of 68% and a partial response rate of 100% using a combination of cisplatin, vincristine, doxorubicin, and cyclophosphamide.1 A larger trial studied a cisplatin, doxorubicin, and cyclophosphamide combination and had an overall response rate of 50% and a median survival of just > 3 years. The most common cause of death is intrathoracic progression of the thymoma (a myasthenic crisis). In one series, almost one fourth of patients died of unknown causes.3

This case is unique in a number of respects. First, the young age of the patient is unusual, as most cases present in middle age. As stated above, there is only one other case report of a patient having myocarditis in association with malignant thymoma. This patient also suffered from rapidly progressive heart block that resulted in an asystolic arrest and death. Heart block has been described before in myocarditis, but never before as the result of a paraneoplastic syndrome. It is very possible that unrecognized myocarditis and heart block is a cause of death in patients with malignant thymoma who die of unknown causes. Heart block in cases of myocarditis seems to be directly related to myocardial necrosis and destruction of the conducting system, and those who develop heart block have evidence of more extensive necrosis than those who do not. Patients with myocarditis who develop conduction disturbances such as left bundle branch block have a worse prognosis than patients who do not.5

**CONCLUSION**

Heart block as a manifestation of a paraneoplastic syndrome is described for the first time. Clinicians who treat patients with these disorders should be more aware of myocarditis as paraneoplastic syndrome, and they should be alert to the possibility of the development of heart block. However, even in this case with prompt recognition and institution of pacing, a fatal outcome could not be avoided.

**REFERENCES**


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**Figure 1.** A 12-lead ECG at time of presentation showing sinus tachycardia with ST-segment elevation in leads V1-V5, as well as in leads III and aVF.

**Figure 2.** A 12-lead ECG showing 3:1 Mobitz II heart block with a ventricular rate of 37 beats/min.