individuals undergo annual magnetic resonance imaging examinations [7]. Surveillance may begin as early as 20 years of age and should continue for the lifetime of the individual. Finally, family members of the patient should be counseled and offered genetic screening.

In summary, thoracic surgeons should have a high index of suspicion for BHD syndrome. The initial presentation is often a pneumothorax, and unless further imaging is pursued, the condition may be misdiagnosed as a spontaneous primary pneumothorax. Sequence analysis of the FLCN gene is diagnostic, and those diagnosed with the syndrome should be referred for counseling and radiographic surveillance of the kidneys.

References

Reoperative Lung Transplantation for Donor-Derived Pulmonary Mucormycosis
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A 64-year-old male with end-stage lung disease underwent right orthotopic lung transplantation. After doing well initially, he developed acute hypoxic respiratory failure with allograft pneumonia. Donor operative cultures demonstrated mold of the *Mucor* species, which were corroborated by donor endobronchial cultures obtained near the right mainstem bronchial anastomosis. The patient was treated with reoperative bilateral orthotopic lung transplantation in combination with antifungal agents. The operation was performed successfully, using lungs donated after cardiac death and treated with ex vivo lung perfusion. The patient has recovered well, remaining on room air with good allograft function, without evidence of fungal disease.

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Pulmonary mucormycosis has an extraordinarily high mortality. The extant literature suggests that a combined strategy of extirpation and antifungal therapies yields the best possible outcomes. However, little is known about mucormycosis in lung allografts and whether pneumonectomy for extirpation with retransplantation is efficacious. Donor organ management can also be challenging in the setting of reoperative lung transplantation. Strategies to expand the donor organ pool may be required for these higher-risk patients to ensure timely retransplantation. Such strategies include the use of organs obtained in the setting of donation after cardiac death and screening for prospective allograft function using ex vivo lung perfusion.

A 64-year-old man with end-stage restrictive lung disease secondary to idiopathic pulmonary fibrosis and no other noteworthy medical or surgical history was referred to the University of Maryland Medical Center for lung transplantation evaluation. At referral, the patient required noninvasive support via a high-flow supplemental $O_2$ system with a fraction of inspired $O_2$ of 60%, with declining activity tolerance. The remainder of his pre-lung transplant evaluation was unremarkable, and he was deemed a suitable prospective lung transplant recipient.

Given the patient’s disease process, age, and rapidly declining functional capacity, he was listed for single lung transplantation. A suitable donor lung with excellent gas exchange and mechanics was obtained from a healthy male donor; the history was noteworthy for the donor being found with his face in soil. However, donor bronchoscopy was satisfactory, with minimal secretions present. The patient underwent right orthotopic lung transplantation. Cardiopulmonary bypass, via cannulation of the ascending thoracic aorta and right atrium, was required because of exacerbated hypoxemia upon institution of single lung ventilation. The operation was technically satisfactory, with a total organ ischemic time less than 3 hours, and excellent completion arterial blood gases, pulmonary circulatory hemodynamics, transesophageal echocardiography, and flexible fiberoptic bronchoscopy. The patient was extubated within 24 hours following the operation, with 3 L of supplemental $O_2$ via nasal cannula within 48 hours and without any inotropic...
or vasoactive infusions within 48 hours. He continued to improve over the next few days. On postoperative day 7, he developed acute onset dyspnea and hypoxemia. Flexible fiberoptic bronchoscopy revealed acute mucous plugging of the right mainstem bronchus, which was suctioned successfully. The anastomosis was widely patent and without distal plugs. The patient’s condition was subsequently improved and stable on 40% fraction of inspired O2 via high-flow nasal cannula. At this time, donor cultures taken at organ procurement were returned and demonstrated mold of the *Mucor* species, consistent with possible donor respiratory tract soil exposure. Intravenous and inhaled amphotericin B were initiated. The patient’s condition deteriorated over the course of several days, with worsening hypoxemic respiratory failure necessitating invasive mechanical ventilation. Diagnostic testing failed to identify allograft rejection, pulmonary embolism, left ventricular or left-sized cardiac valvular dysfunction, or allograft pulmonary edema. However, endobronchial biopsy identified allograft right mainstem bronchial wall-invasive mucormycosis (Figs 1A, 1B), just distal to the bronchial anastomosis. A computed tomographic chest scan demonstrated infiltrates within the right lower lobe, and to a lesser extent in the right middle lobe, with total atelectasis of the right lower lobe and partial atelectasis of the right middle lobe (Fig 1C). Pulmonary mucormycosis was thought to be the principal etiology of the patient’s allograft and respiratory failure.

Given the relatively poorer prognosis of patients with pulmonary mucormycosis who do not undergo surgical extirpation (see Comment), a decision was made to pursue retransplantation, because this was the only feasible strategy to eliminate infection and provide adequate pulmonary function. Moreover, the patient’s indication for retransplantation was now infectious lung disease, with potential infection of the contralateral native lung. Consequently, the patient was relisted for bilateral lung transplantation. Suitable donor organs were obtained in the setting of donation after cardiac death (DCD; see Comment) [1, 2]. Although the lungs were satisfactory from both anatomic and physiologic standpoints, we subjected them to ex vivo lung perfusion (EVLP; see Comment) [3]. The lungs were perfused for 4 h, with excellent gas exchange (outflow $P_{O2}$ 430–490 mm Hg) and mechanics (static compliance 65–90 mL/cm H2O). Subsequently, bilateral orthotopic lung transplantation was performed using these donor lungs. A bilateral thoracosternotomy was performed, and cardiopulmonary bypass via ascending thoracic aortic and right atrial cannulation was used, given the anticipated technical complexity of the case. Right allograft pneumonectomy was performed, with an additional resection of two recipient bronchial rings to ensure adequate extirpation of infection. The new donor right lung was then implanted, followed by native left pneumonectomy and donor left lung implantation. The procedure was satisfactory, with optimal completion arterial blood gases, pulmonary circulatory hemodynamics, transesophageal echocardiography, and flexible fiberoptic bronchoscopy. The patient’s postoperative course was noteworthy for prolonged mechanical ventilatory requirements, without the need for tracheostomy, because of the development of a bleeding duodenal ulcer requiring surgical treatment. A course of both intravenous and inhaled amphotericin B was continued postoperatively. The patient weaned from mechanical ventilation and has done well since, having been discharged recently from the hospital. Pathology from the explanted right lung allograft demonstrated endobronchially invasive mucormycosis, with coexistent parenchymal evidence of acute cellular rejection (ISHLT Grade A3B3).
Comment

Fungal infections in the lung transplant population are increasingly recognized, with major functional morbid complications, notably an increased risk of bronchiolitis obliterans syndrome [4]. Of these, pulmonary mucormycosis is uncommon, but has an extraordinarily poor prognosis across all organ transplant recipient populations. The extant literature on pulmonary mucormycosis clearly demonstrates that a combined strategy of surgical resection and amphotericin B yields the best survival outcomes. The largest series, which also reviewed previous reports within the literature, reported a mortality of 11% in patients undergoing combined surgical therapy, in contrast to 68% in patients treated with antifungal agents alone, although high-risk patients may have been denied surgical treatment [5]. Consequently, with clearly donor-derived bronchial invasive infection in the absence of disseminated disease, we chose an aggressive strategy of bilateral retransplantation. Our review of the literature suggests that this is the first reported case of donor-derived pulmonary mucormycosis successfully managed with reoperative lung transplantation. Two previous cases of allograft mucormycosis manifesting in the peritransplant period have been reported [6, 7]; these patients were managed nonoperatively with successful outcomes.

Obtaining donor organs in the DCD setting is common in abdominal organ transplantation, but comparatively rare in thoracic organ transplantation. However, recent results regarding the use of DCD lungs from centers in the United States [1], Canada [8], and Australia [2] are promising. We obtained a well-functioning donor organ block, but given possible ischemic injury and pulmonary edema in the DCD setting, we chose to evaluate the lungs using EVLP. In EVLP, donor lungs are ventilated mechanically and perfused with crystalloid-based Steen solution in a closed circuit [3]. EVLP can serve as a diagnostic tool for allograft functional assessment and to rehabilitate marginal donor lungs [8]. Thus, salient features of this case include reoperative bilateral lung transplantation as the critical element of the treatment strategy, the use of a DCD lung donor subjected to EVLP, and continued adjunctive systemic and local antifungal therapies.

References


Pulmonary Metastatic Gastric Cancer Mimicking A Giant Mediastinal Cyst

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Cysts and cavities are common radiologic abnormalities. Pulmonary metastasis comprises a rare entity of thoracic cystic diseases. We reported a case of giant cyst at the left anterior mediastinum that was pathologically confirmed as a lung metastasis from previously resected gastric cancer. The cyst was completely removed with wedge resection of the surrounding lung through a left anterior thoracotomy. One should always keep in mind the possibility of an intrathoracic cyst near or at the mediastinal region that may originate from metastatic lesions to the lungs when patients have previous cancer history.

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Cysts and cavities are common abnormalities on chest radiographs and chest computed tomography (CT). Confirmation of the nature of thoracic cystic or cavitary lesions may be challenging. Differential diagnoses of such lesions range from congenital to the acquired cystic diseases but very rarely cases of metastatic cancer [1, 2]. We report a case of lung metastasis from gastric cancer presenting as a giant cyst at the left anterior mediastinum that mimics a cyst of thymic origin.

A 73-year-old man was hospitalized on account of episodes of hemoptysis with a large mass abutting from the left anterior mediastinum on chest radiograph (Fig 1A). He had been treated for a stage II gastric cancer by subtotal gastrectomy half of year prior to this admission. The chest radiograph taken before gastrectomy appeared to be normal. Chest computed tomography (CT) revealed a well-defined thin-walled cyst, 10 cm in diameter, at the...