CHEST

A 24-Year-Old Woman With Precipitous Respiratory Failure After Lung Transplantation

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CASE PRESENTATION: A 24-year-old woman with ΔF508/Y1092X cystic fibrosis (CF) complicated by severe obstructive lung disease (FEV_1 of 30% predicted) was admitted for IV antibiotics for planned sinus surgery resulting from severe chronic sinusitis causing frequent exacerbations and declining lung function. She had persistent airway infection with multidrug-resistant *Pseudomonas aeruginosa,* methicillin-resistant *Staphylococcus aureus*, and growth of a fungus presumed to be an airway colonizer, identified as *Stephanoascus ciferrii* 1 year before presentation. Two days after surgery, she developed acute respiratory failure requiring mechanical ventilation. On day 4 of mechanical ventilation, venovenous-extracorporeal membrane oxygenation (VV-ECMO) was initiated for refractory respiratory failure. The following day, she was listed for bilateral lung transplant and was transplanted 4 days later. Following transplantation, she was decannulated from ECMO; however, over the next 12 hours, oxygenation deteriorated requiring reinstitution of VV-ECMO for presumed severe primary graft dysfunction. Despite treatment with broad spectrum antimicrobial coverage with piperacillin/tazobactam, ciprofloxacin, linezolid, micafungin, voriconazole, and ganciclovir, she failed to improve and developed complex bilateral pleural effusions.

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Physical Examination

Twelve hours posttransplant the patient was afebrile with blood pressure of 105/44 mm Hg supported by norepinephrine. Her respiratory rate was 36 and oxygen saturation was 88% on volume-controlled mechanical ventilation with tidal volume of 300 mL, positive end-expiratory pressure of 12, and F_{IO_2} of 1.0 before resumption of VV-ECMO. On examination, she was sedated and had diminished breath sounds with diffuse rales.

Diagnostic Studies

The patient's WBC count was 38,500/μL with neutrophil predominance 12 h after transplant. Her arterial blood gas showed a pH of 7.41, P_{CO_2} of 51.4 mm Hg, and P_{O_2} of 66.8 mm Hg. Renal function was normal. Chest radiographs were done with progressive bilateral opacifications compared with prior imaging (Fig 1). Bronchoscopy preformed on the day of transplantation demonstrated growth of her

ABBREVIATIONS: CF = cystic fibrosis; VV-ECMO = venovenous-extracorporeal membrane oxygenation

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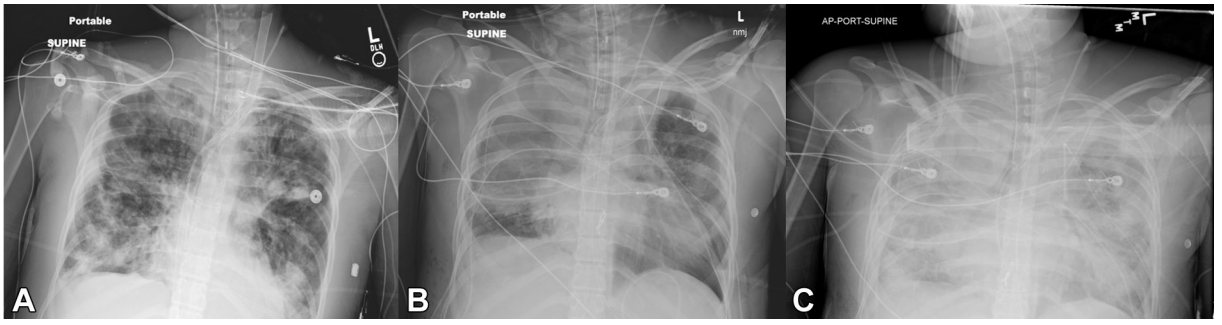

Figure 1 – *A, Chest radiograph at initiation of extracorporeal membrane oxygenation before transplant. B, Chest radiograph immediately after transplant. C, Chest radiograph on postoperative day 1. Used with written permission from the patient.*

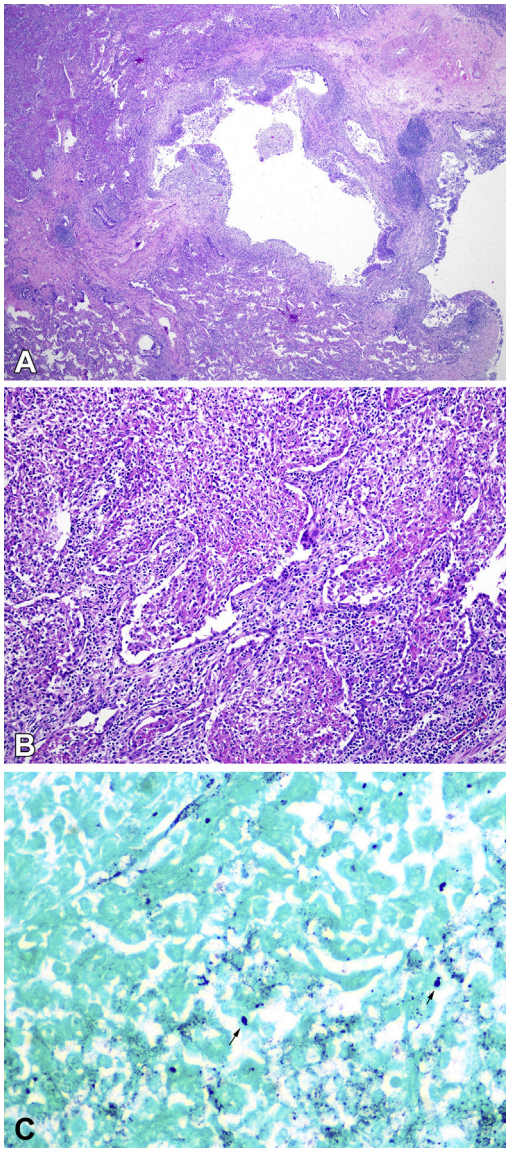

Figure 2 – *A, Explanted lungs showed bronchiectasis and airway inflammation consistent with cystic fibrosis (hematoxylin and eosin, ×20). B, In addition, there was widespread lobar pneumonic exudate (hematoxylin and eosin, ×100). C, Gomori-Grocott methenamine silver stain identified yeast fungal organisms (×600), some that showed budding (arrows). Used with written permission from the patient.*

pretransplant airway colonizer identified as *Stephanoascus ciferrii,* but the remainder of her cultures were negative. One week posttransplant, blood cultures also grew the fungus previously identified as *S ciferrii.* Two weeks posttransplant, video-assisted thorascopic surgery with decortication was done for complex bilateral pleural fluid collections. Pleural debris was cultured with heavy growth of fungi similar to prior cultures from blood, bronchoalveolar lavage, and explanted lungs (Fig 2).

What is the diagnosis?