Intracerebral abscess: A complication of severe cystic fibrosis lung disease

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Intracerebral abscess is an uncommon complication of severe cystic fibrosis lung disease. The present report describes a case of fatal multiple intracerebral abscesses in a patient with a severely bronchiectatic, nonfunctioning right lung and chronic low-grade infection. The patient was previously turned down for pneumonectomy. Intracerebral abscess in cystic fibrosis and the potential role of pneumonectomy in the present patient are discussed.

Key Words: Cystic fibrosis; Intracerebral abscess; Pneumonectomy

Brain abscess as a complication of cystic fibrosis (CF) is reported infrequently in the literature (1-4). We present a case of multiple intracerebral abscesses in a patient with unilateral, severe CF-related lung disease.

CASE PRESENTATION
An 18-year-old male Hutterite, diagnosed with CF (genotype unknown) at three months of age, presented with bifrontal and occipital headache, neck stiffness and vomiting for 72 h. No recent dental work or surgical interventions were performed. He did not have an implanted intravenous access device. He had regular contact with animals because he lived on a farming colony. His right lung was severely damaged, with absence of both ventilation and perfusion demonstrated on ventilation-perfusion scanning. Severe unilateral cystic bronchiectasis was demonstrated on chest radiography (Figure 1) and plain computed tomography (CT) imaging (Figure 2). His condition developed postoperatively following a nasal polypectomy at 11 years of age. Subsequently, he had difficulty maintaining his body weight, had chronic high-volume purulent sputum production, and had chronic low-grade fever with occasional hospitalization. He received chronic chest physiotherapy and daily treatment with nebulized deoxyribonuclease. The lungs were colonized with Klebsiella pneumoniae (resistant to ampicillin and trimethoprim-sulfamethoxazole) and fully sensitive Pseudomonas aeruginosa (sputum culture), and he was treated with inhaled tobramycin 80 mg twice daily (continuously) and ciprofloxacin 500 mg twice daily (two weeks on and two weeks off). At several points over the course of follow-up, the notion of a right pneumonectomy was raised, but it was rejected by two thoracic surgeons because of concerns about perioperative complications; specifically, a 23% incidence of bronchopleural fistula in one report (5). His forced expiratory volume in 1 s, three months before admission, was 41% of predicted (1.7 L).

On examination, he was afibrile with stable vital signs. There were no papilledema or focal neurological findings. However, there was significant meningismus. There was tracheal deviation to the right, and bronchial breathing and diminished breath sounds in the right hemithorax. Digital clubbing was prominent. Investigations revealed a leukocytosis of $22 \times 10^9/L$ with a left shift, and a cerebrospinal fluid white blood cell count of $109/L$ with a ...

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and he developed respiratory failure. In the end, his family decided to withdraw active management and pursue compassionate terminal care. He died after 34 days in the hospital. A histological examination of cerebral tissue was not performed because an autopsy was declined by the family.

DISCUSSION
Brain abscess has long been recognized as a rare complication of CF, with only 12 cases previously reported in the literature (1-3,6-11). It is a complication almost never seen in children with CF, with only one previously reported case (11). Cooper et al (6) postulated that its predilection for young adults over children may reflect an increased susceptibility to disseminated infection with advancing disease. However, systemic pseudomonal infection is exceedingly rare in CF patients, suggesting a robust immune system. Previous reports have suggested that many cases may be due to sinusitis and/or immunosuppression. Abscess formation from sinusitis is usually ipsilateral and frontal in location, and contiguous with the affected sinus (12) – a scenario not demonstrated in our case. Others (7) have suggested a hematogenous spread of infection, most likely from an extrapulmonary source. Collis (4) postulated that intracerebral abscess associated with suppurative lung infection was due to embolic spread via retrograde flow through the vertebral veins. Wiersbisky et al (8) also suggested that a possible mechanism for hematogenous spread from infected lung tissue to brain is via local erosion of blood vessels in chronically inflamed bronchi, leading to occult bacteremia. We hypothesize that the development of intracerebral abscess may be an effect of time and chance. There may be repeated embolic exposures throughout the life of a chronically infected CF patient, and eventually one or more will not be removed by the immune system, establishing local infection. In the present case, the presence of multiple widely dispersed abscesses favours a hematogenous mechanism of spread, with the lung as the most likely source.

The role of pneumonectomy in CF patients is controversial. Pneumonectomy has been used to try to delay progression of the disease in the affected lung through resection of the affected segments. Some authors have advocated its use to reduce infection, improve quality of life and prolong survival in chronic infection (13). Others, however, have reserved this practice only for cases with respiratory failure as a bridge to transplant (14,15). Most reported cases of pneumonectomy in CF are in patients with respiratory failure (15). Our patient, who did not have respiratory failure, raises the question of the value of pneumonectomy in patients with a chronically infected/colonized nonfunctioning lung with a 'preserved' forced expiratory volume in 1 s. Pneumonectomy in CF patients has multiple considerations – chief among them are complications (eg, bronchopleural fistula, inoculation or colonization of the resected hemithorax, and...

Figure 1) Chest x-ray showing severe unilateral cystic bronchiectasis of the right lung

Figure 2) Computed tomography of the chest, showing severe unilateral cystic bronchiectasis of the right lung

Figure 3) Computed tomography of the head demonstrating ring-enhancing lesions. There is also a ventricular drain in the right frontal area
ventilator dependence) and potential adverse effects on lung transplantation. Current literature suggests that pneumonectomy is not a contraindication to future lung transplant (14-17). The issue of pneumonectomy was raised at several points before his final hospitalization, but he was dismissed as being too risky (ie, high incidence of bronchopleural fistula) (5). A lung transplant specialist indicated that an uncomplicated postoperative course in the present patient would not preclude a future unilateral or single lung transplant of the left lung. The literature around this issue suggests that most patients do well and have improved quality of life, and reduced numbers of infections or hospitalizations (13,14,16,17).

The absence of positive cerebrospinal fluid cultures in our patient, likely the result of chronic use of antibiotics, make firm associations regarding the primary source of the infection difficult. Only eight of 12 reported cases (67%) of CF-associated lung abscess have had positive culture results, the majority of which are pathogens associated with the oral cavity rather than the lung (10). Our patient did not grow any positive cultures from the infected cerebral tissue, cerebrospinal fluid or blood cultures. The sputum culture was unchanged from the known colonizing microbes (Pseudomonas species and Klebsiella species).

CONCLUSIONS
A high index of suspicion for intracerebral abscess is required in patients with severe CF-related lung disease and headache. Our patient was afebrile and had no focal neurological findings, despite the presence of multiple intracerebral abscesses. Perhaps an earlier right-sided pneumonectomy may reduce or even eliminate the risk of brain abscess in the present patient. He is likely to have tolerated the resection because his right lung was not contributing to the measured lung function. We suggest that in cases such as ours, with a nonfunctioning lung and chronic or repeated infectious exacerbations, consideration should be given to pneumonectomy to reduce the risk of complications and improve the quality of life.

REFERENCES