

275* Infection by *Nocardia farcinica* in CF

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Infection by *Nocardia* is an uncommon disease, affecting immunocompromised patient. This bacteria has rarely been isolated from CF patients, especially without any oral corticosteroids. We report a case of a patient with CF harbouring *Nocardia farcinica*.

Methods: the patient is a 18-year-old male with diagnosis of CF at 8 years old (F508del/G85E). An allergic bronchopulmonary aspergillosis was treated in 1998 with itraconazole and a primocolonization with *Pseudomonas aeruginosa* (Pa) was eradicated in 2003. From may 2006, he presented recurrent left and right pneumothorax. In June 2006, he presented with dyspnea, fever and nodular eruption on the ankles. Chest X-Ray and CT scan revealed a right pneumothorax, severe bronchiectasis and bilateral alveolar consolidation. Sputum specimen isolated *Nocardia farcinica*, without any other pathogens. A treatment with intravenous cotrimoxazole associated with imipenem and amikacin during 3 weeks was initiated followed by oral cotrimoxazole during 9 months. The symptoms and the alveolar consolidation CT scan improved.

During 2007, his respiratory condition worsened. His FEV1 declined from 50 to 26% predicted, pneumothorax recurred. He was presenting a chronic colonization with Pa and was expecting a lung transplantation. Unfortunately he died in 2008.

Results: *Nocardia*, a gram positive bacilli can cause mainly pulmonary infection, usually in the setting of immunodepression. The most frequent is *Nocardia asteroides*. In CF, very few cases have been reported, almost always *Nocardia asteroides*, but exceptionnaly *Nocardia farcinica*.

Conclusion: In case of worsening pulmonary condition in CF patients, physicians managing CF patients should consider such uncommon organisms in patients failing to respond to usual therapy or worsen their lung's function without any explanation.

276* First colonisation with *P. aeruginosa* – only detected in the upper airways – eradicated with sinonasal inhalation of Tobramycin

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Case report: The 11 year old CF patient suffered from chronic rhinosinusitis as predominant symptom. He was included in our longitudinal study on non-invasive assessment of upper and lower-airway colonisation with *P. aeruginosa* (P.a.).

In June 2006 P.a. was detected in his airways for the first time: exclusively in nasal lavage. Despite a two-weeks anti-pseudomonas i.v.-course the bacterium again was detected in upper airway sampling in August 2006, but again not in lower airway probes.

In off-label use the patient inhaled Tobramycin 80 mg (GernebcinTM) once daily for 4 weeks with the novel Pari SinusTM nebuliser, which in in-vitro analyses was shown to deposit aerosol into the paranasal sinuses. Since then, he did not reveal positive Pa. samples from upper and lower airways in continuous monitoring and Pa. antibodies in serum remained negative.

Discussion: The upper airways are regularly involved in CF as the epithelial CFTR-defect equally concerns sinonasal mucosa. Impaired clearance of this airway segment implies the risk of first colonisation with P.a. and its persistence. This would not have been detected in our patient applying the current best standards of CF-care.

Secondary, our case report includes the first record on antibiotic inhalation with the novel Pari SinusTM device. After sinonasal therapy with Tobramycin we did not detect Pa. in the patients airways in frequent controls.

Conclusion: Upper airway sampling with non-invasive methods like nasal lavage can allow earlier detection of P.a.-colonisation. Eradication of P.a. in this airway segment could have the potential to postpone chronic pulmonary P.a.-infection, the leading cause of morbidity and mortality in CF.

277* Lung transplantation in patients with cystic fibrosis and *Mycobacterium abscessus* infection

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Mycobacterium abscessus is a multiresistant rapid-growing non-tuberculous mycobacteria. *M. abscessus* lung infection is difficult to treat and has become an increasing problem in patients with cystic fibrosis (CF). Fatal infection has been described after lung transplantation and *M. abscessus* infection has thus been considered a relative contraindication for transplantation.

We have performed double lung transplantation in three CF patients with ongoing treatment for chronic *M. abscessus* lung infection and, most recently, one CF patient with intermittent growth. The surgical approach was sternotomy (2), clamshell (1) and bilateral single sequential lung transplantation through anterior thoracotomy (1). The patients were 10, 28, 26 and 20 years old, respectively, at the time for surgery and all are alive and have a stable lung function after 6.5, 4, 2.5 and 0.8 years. Despite prolonged post-operative treatment with a combination of antimycobacterial drugs, the first three patients developed severe mycobacterial skin infection with abscesses, requiring skin transplantation in two cases. They were also treated for infections with *Pseudomonas aeruginosa* (3) and *Aspergillus fumigatus* (1). All three have developed severe kidney dysfunction and hearing impairment due to prolonged aminoglycoside treatment.

Conclusion: Lung transplantation in patients with *M. abscessus* lung infection is feasible but associated with severe complications.

278 Sequential bronchoscopy in the management of patients with lobar atelectasis

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Early recognition and treatment of lobar and segmental atelectasis is essential in order to prevent progression of bronchiectasis and irreversible collapse. A common cause of atelectasis in patients with cystic fibrosis (CF) is mucus plugging and secretions secondary to allergic bronchopulmonary aspergillosis (ABPA) and aspergillus bronchitis. Despite early reservations it is now accepted that bronchial lavage and instillation of rhDNase is effective in re-expanding collapsed lobes. In a previous case series; however, a significant number of patients failed to re-expand lung despite this approach in conjunction with standard medical management.

We report a series of 5 CF patients (2 males, 3 females, ages 7–66 years) who presented with pulmonary exacerbations and evidence of lobar collapse radiologically between 2007 and 2008. All patients were diagnosed with either ABPA or aspergillus bronchitis and treated with steroids, anti-fungal medication, nebulised rhDNase, and physiotherapy. Patients underwent sequential flexible bronchoscopy for lavage and instillation of rhDNase; procedures were performed at least one week apart (see Table 1). All patients achieved complete resolution and lung function returned to baseline.

In conclusion, sequential bronchoscopy is capable of achieving complete radiological resolution in patients who have failed to re-expand after a single procedure.

Table 1. Outcome of sequential bronchoscopies

Patient	Gender/age (years)	Microbiology	Site of atelectasis	Instilled dose rhDNase	Number of bronchoscopies	Complete resolution
1	M/17	<i>P. aeruginosa</i> / <i>A. fumigatus</i>	Left upper lobe	5 mg	2	Yes
2	F/17	<i>A. fumigatus</i>	Right upper lobe	5 mg	2	Yes
3	M/66	<i>P. aeruginosa</i> / <i>A. fumigatus</i>	Right lower lobe	5 mg	4	Yes
4	F/7	<i>A. fumigatus</i>	Left upper lobe	2.5 mg	3	Yes ^a
5	F/8	<i>A. fumigatus</i>	Right lower lobe	2.5 mg	2	Yes ^b

^aAfter 2 weeks; ^bAfter 8 weeks.