Stenotrophomonas Maltophilia Isolated from the Sputum of the Patient with Cystic Fibrosis Mutation I1234V: First Qatari Report

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Abstract:
A 17 year old Qatari female of Arab descent with cystic fibrosis (CF) carrying pathogenic mutation I1234V had severe respiratory disease associated with chronic Pseudomonas aeruginosa broncho-pulmonary infection with recurrent episodes of mild hemoptysis. Despite regular courses of intravenous anti-pseudomonal antibiotics, she continued to deteriorate over six months and died.

It is suggested that the presence of Stenotrophomonas maltophilia was an important factor in this case which illustrates the need for continuing vigilance in considering the acquisition of resistant organisms in such patients on long-term antibiotic therapy.

Key words: CF, Chronic Pseudomonas Aeruginosa, Stenotrophomonas Maltophilia

Introduction:
Chronic bacterial pulmonary infections are a major cause of morbidity and mortality in patients with cystic fibrosis (CF) (1,2). Pseudomonas aeruginosa is the chief Gram-negative organism recovered from the respiratory tract of patients with CF (3). Colonization of the respiratory airway with multi-resistant pathogens leads to ineffective conventional antimicrobial therapy (4). One of these multi-resistant organisms, Stenotrophomonas maltophilia, is generally considered to be a late colonizer of CF patients (5).

For the first time in Qatar, we report the isolation of S. maltophilia from the respiratory secretion of a female with cystic fibrosis mutation I1234V. The patient later died.

Case Report:
Following recurrent chest infections and two elevated sweat chloride concentrations of 68 mmol/L and 87 mmol/L, a Qatari female was first diagnosed at 7 years of age with cystic fibrosis (homozygous for I1234V mutation). Pseudomonas aeruginosa was isolated from her sputum at 11 years of age. She had one episode of major hemoptysis at 13 years of age and was started on three-monthly elective courses of intravenous antibiotic for chronic P. aeruginosa infection. On each occasion she received intravenous anti-pseudomonal antibiotics for at least two weeks. Subsequent recurrent episodes of minor hemoptysis required several admissions for acute and chronic respiratory infections. Both mucoid and non-mucoid strains of P. aeruginosa were isolated frequently from the sputum.

Maintenance treatment included inhaled fluticasone propionate and oral azithromycin for its presumed anti-inflammatory and immune modulating effect. Despite this intensive approach, she continued to deteriorate over six months with worsening respiratory function, increasing oxygen dependence and decreased exercise tolerance.

Her physical examination was notable for her weight of 28 kg and body mass index of 14 (below the 5th centile) and a respiratory rate of 30/min. She had significant digital clubbing and diffuse crackles on chest auscultation. There were no nasal polyps. The rest of examination was unremarkable. Tests of pulmonary function revealed mixed severe obstructive and restrictive lung disease and her spirometry trend showed a significant drop in forced expiratory volume in one second (FEV1) from 41% to 28% over the last six months. Repeated sputum cultures grew profuse growth of multi-resistant organisms; S. maltophilia resistant to aminoglycosides and cephalosporins but moderate resistant to trimethoprim. In addition her sputum cultures grew both mucoid and non-mucoid P. aeruginosa sensitive to cefepime and amikacin.

She received intravenous anti-pseudomonal antibiotics and trimethoprim-sulfamethoxazole but continued to deteriorate with worsening respiratory function and failure to maintain adequate oxygenation and ventilation despite intensive therapy and non-invasive ventilation. She died about two weeks later.
Discussion:

*Stenotrophomonas maltophilia*, previously known as *Pseudomonas maltophilia* and later as *Xanthomonas maltophilia*, is a gram-negative rod ubiquitous multi-resistant commensal that is isolated readily from water, soil, and sewage and is found increasingly in nosocomial infections. It has been reported in patients with CF for almost 20 years (6).

Although this organism has been primarily described as an opportunistic organism in hospital outbreaks and in debilitated patients (7,8), there have been recent publications reporting its incidence and prevalence among CF patients (9,10), as well as its possible pathogenic role in CF pulmonary disease (11). We speculate that *S. maltophilia* might have played a role in the course of CF lung disease in our patient. She had several factors making her susceptible to the emergence of *S. maltophilia*: significantly low growth parameters, low spirometric values and long-term antibiotic therapy as suggested in a recent investigation (12).

Therapy for *S. maltophilia* is problematic because of the broad antibiotic resistance that typifies this organism (13). In a recent survey of 130 isolates, the most active agents were minocycline, trimethoprim-sulfamethoxazole, and ticarcillin/clavulanic acid (14).

A recent study from our institution reported homozygous 11234V mutation in 29 subjects (17 families) belonging to the same Arab tribe, with ages ranging from 8 months to 21 years (15).

To the best of our knowledge this is the first case reported where *S. maltophilia* has been isolated from the sputum of a patient with CF mutation 11234V. At present it is not known whether this organism is an emerging pathogen in CF patients or is merely colonizing the lung. This organism, which has been reported increasingly from some CF centers during the last decade (2,3), displays an intrinsic resistance to several antimicrobial agents. As the life span of CF patients is increasing, it is important to identify the risk factors for, and the clinical significance of, *S. maltophilia* acquisition.

References: