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CASE REPORT

Moraxella catarrhalis sepsis in a patient with juvenile spinal muscle atrophy

I.C.D. Westendorp[1*], M.A. Tiemessen[1,2], M. de Jong[1], A. Soomers[1], I.M.M.J. Wakelkamp[1], W.G. Boersma[2]

Departments of [1]Intensive Care and [2]Pulmonary Diseases, Alkmaar Medical Centre, Alkmaar, the Netherlands, [*]corresponding author: Department of Cardiology, Onze Lieve Vrouwe Gasthuis, Amsterdam, the Netherlands, tel.: +31 (0)20-599 30 32, fax: +31 (0)20-599 39 97, e-mail: iwestendorp@cs.com

ABSTRACT

***Moraxella catarrhalis* rarely causes severe infections or bacteraemia in healthy subjects. In the literature only four cases of clinical sepsis with *M. catarrhalis* have been described, mostly in immunocompromised patients. We describe a case of a 34-year-old patient with Kugelberg-Welander disease and low body weight (28 kg) who developed clinical sepsis due to *M. catarrhalis* bacteraemia. A review of the literature is given.**

KEYWORDS

Juvenile spinal muscle atrophy, Kugelberg-Welander disease, malnutrition, *Moraxella catarrhalis*, pneumonia, sepsis

INTRODUCTION

After *Streptococcus pneumoniae* and *Haemophilus influenzae, Moraxella catarrhalis* is the third commonest pathogen of the respiratory tract among patients with chronic obstructive pulmonary disease (COPD).[1] Although *M. catarrhalis* is often present in the respiratory tract as a commensal micro-organism, it can clearly be pathogenic under certain circumstances.[2,3] Its importance as a pathogen, both in children and in adults, has been increasingly recognised in the past decades.[4]

Invasive infections with *M. catarrhalis* have seldom been described: in the literature only four cases of clinical sepsis (i.e. septic shock) with *M. catarrhalis* have been described in adults, although more cases (at least seven) have been reported in children. Bacteraemia (at least 61 reported cases) and severe infections with *M. catarrhalis* are also infrequent but have been described in immunocompromised patients, mainly those with haematological malignancies, in patients with chronic obstructive pulmonary diseases and in the elderly, as well as in healthy subjects. Our patient, with severe muscle atrophy, very low body weight and restrictive lung disease and therefore extremely limited (respiratory) reserves, demonstrates the favourable outcome with aggressive treatment of this rare sepsis.

CASE DESCRIPTION

A 34-year-old male, known to have juvenile spinal muscle atrophy type III (JSMA-III or Kugelberg-Welander disease) and moderately severe restrictive lung disease, was admitted with pneumococcal pneumonia, diagnosed by sputum cultures. He was treated with bronchodilators, prednisolone (50 mg/24 h) and amoxicillin, and later penicillin, intravenously. The patient had been treated with prednisolone twice before, but not recently. His condition improved at first, but later deteriorated and on day 5 of admission the patient became respiratory insufficient with recurrence of fever (39.2 °C). Laboratory investigations showed low creatinine (18 μmol/l), urea (2.0 mmol/l), albumin (31.1 mmol/l), and a severe hypophosphataemia (0.26 mmol/l), with an increased white blood cell count (24.2 x 10^9/mm^3) and C-reactive protein (126 μm/l). During his admission his body weight decreased from 28 to 22 kg, probably as a result of decreased dietary and

fluid intake. Differential diagnostic considerations consisted of nosocomial superinfection, aspiration – as the patient had weakened and had difficulty in swallowing – development of resistance to penicillin, ARDS or empyema.

The patient was transferred to the ICU for mechanical ventilation. Blood pressure was 85/45 mmHg and heart rate 135 beats/min. Measurements with a pulmonary artery catheter showed a septic profile with low systemic vascular resistance (450 dynes.sec/cm^5), a central venous pressure of 8 mmHg, a systolic pulmonary artery pressure (SPAP) of 18 mmHg and diastolic pulmonary artery pressure (DPAP) of 10 mmHg. Cardiac output and index were increased (CO 7.8 l/m, CI 5.9 l/min/m^2). Possible causes for the severe hypoposphataemia that were considered were, in the first place, respiratory alkalosis, which often occurs in sepsis. Secondly, the use of glucocorticoids can cause hypophosphataemia. Furthermore saline infusion, which can cause hypocalcaemia and parathyroid hormone release, or a decreased dietary intake were considered. Treatment consisted of intravenous substitution, restoration of acid/base balance, and rapid commencement of parenteral feeding. Additional treatment consisted of fluid replacement, dobutamine, and noradrenalin. On day 5 in the ICU (day 10 after admission), the sputum and two blood cultures that had been taken on admission to the ICU yielded *M. catarrhalis* and the penicillin was changed to amoxicillin/clavulanic acid, which improved the patient's condition markedly. After a total stay of 48 days the patient could be discharged from hospital, on ventilation via a tracheal cannula for 20 hours/day. Six months after discharge from our ICU, the patient is on night time ventilation only.

DISCUSSION

We describe a patient with benign JSMA-III and moderately severe restrictive lung disease who developed a nosocomial sepsis with *M. catarrhalis* after pneumococcal pneumonia. The diagnosis of JSMA-III, a disease of slowly progressive limb-girdle weakness, had been made in childhood in our patient. The prediction of progression and degree of disability is difficult and extremely variable, but most patients are bound to a wheelchair by their mid-thirties. The cause of death in these patients is often pneumonia. Our patient suffered severe wasting and muscle weakness and contractions, rendering him wheelchair bound.
The low body weight of our patient may well have played a role in the course of the disease. Defence mechanisms to infection have been shown to be strongly affected by nutritional status.[4] Both deficiency of protein energy and individual nutrients (trace minerals and vitamins, particularly zinc, iron, selenium, vitamins A, B_6, C and E) are associated with impairment of cell-mediated immunology, complement activation and secretory immunoglobulin antibody response. Complement plays an important role in host defenses against *M. catarrhalis*, as does the IgG3 response to *M. catarrhalis* outer membrane proteins.[2]
It has been estimated that approximately 1 to 5% of healthy adults is colonised with *M. catarrhalis*, and even higher percentages are found among children and among patients with COPD.[5] In the last decades *M. catarrhalis* has been recognised and a possible virulent pathogen, both in the respiratory tract as in other clinical situations.[2] While colonisation with *M. catarrhalis* is highly prevalent, over the last two decades only 61 cases of bacteraemia with *M. catarrhalis* have been described, whereas only four cases of clinical sepsis have been reported in adults, and at least seven cases in children. Although we must assume that the reported cases present only the 'tip of the iceberg' the *M. catarrhalis* sepsis remains relatively rare. Among adults 60 to 70% of the patients with bacteraemia were immunocompromised or had underlying pulmonary disease (COPD), but also other immunocompetent hosts have been described.[6,7] Bacteraemia was found to be associated with sinusitis, pharyngitis, pneumonia, meningitis, and endocarditis and without a focal source.[2,7] Clinical manifestations vary from mild febrile illness to severe and fatal disease.[2,7] Mortality from *M. catarrhalis* bacteraemia has been estimated in a recent review to be close to 20%,[7] with the highest mortality among patients with endocarditis (four out of five reported patients).[6,7]

Most cases described in the literature (at least seven) occurred in children.[8-10] Two cases were described as early as 1925 and 1928, but details of these cases are difficult to assess.[11,12] More recently only four cases of sepsis due to *M. catarrhalis* have been described in adults (*table 1*). In the three out of four adult cases comorbidity was present: lung cancer,[2] acute myelogenous leukaemia[13] and systemic lupus erythematosis.[14] The fourth patient was a previously healthy 68-year-old man who developed shock and disseminated intravascular coagulation after otitis media with *M. catarrhalis* in all blood and middle-ear cultures.[15] In our case the patient had severe muscle atrophy, restrictive lung disease, and severely reduced body weight, which probably played a role in the development of pneumonia. Only one of the four cases of sepsis in adults had a fatal outcome.
On admission to the ICU several treating physicians were hesitant to start intensive treatment and mechanical ventilation, as the prognosis of the patient appeared to be extremely poor. This case demonstrates that this sepsis can have a positive outcome with reasonable subsequent quality of life when treated, although the prognosis of sepsis with *M. catarrhalis* is strongly dependent on comorbidity.

Table 1 *Overview of adult cases of sepsis with Moraxella catarrhalis, as reported in the literature*

Authors	Year of publication	Sex	Age	Comorbidity	Focus of infection	Initiated therapy	Survival
Wallace *et al.*	1990	♂	71	Lung cancer	Pneumonia	1st erythromycine + piperacilline + amikacine	No
Zenklusen *et al.*	1985	♂	63	Aute myelogenous leukaemia	No focal source	Exact therapy unknown	Yes
Guthrie *et al.*	1988	♀	40	Lupus erythematosis (dormant)	Respiratory tract infection	1st erythromycin 2nd ceftazidime	Yes
Alaeus *et al.*	1991	♂	68	None	Otitis media	1st penicillin 2nd cefuroxim/gentamycin 3rd erythromycin	Yes
Westendorp *et al.*	2005	♂	34	Juvenile spinal muscle atrophy type III (Kugelberg-Welander syndrome)	Pneumonia	1st penicillin 2nd amoxicillin + clavulanic acid	Yes

1st = the first initiated therapy; 2nd = the second initiated therapy, after discontinuation of the first; 3rd = the third initiated therapy, after discontinuation of the first and second.

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