Case report

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Fatal neck necrotizing fasciitis caused by hypermucoviscous Klebsiella pneumoniae

U Wen Yeap

Abstract

Klebsiella pneumoniae is a gram-negative rod enterobacteria that is notorious for its role as carrier of extended spectrum beta-lactamase (ESBL) and its carbapenem resistant Enterobacteriaceae (CRE) species. However, hypermucoviscous Klebsiella pneumoniae is distinctly different from ESBL and CRE. We report a case of neck necrotizing fasciitis caused by hypermucoviscous Klebsiella pneumoniae in a 56-year-old male who presented to our emergency department (ED) with a swollen neck. His condition deteriorated rapidly requiring emergency intubation to secure his airway. Despite aggressive resuscitation and treatment with broad spectrum antibiotics and cytokine adsorption therapy, the patient succumbed to his disease. This report describes the clinical characteristics of hypermucoviscous Klebsiella pneumoniae and emphasizes the importance of early detection and subsequent aggressive source control interventions in necrotizing fascitiis caused by this particular bacteria.

Key words: Hypermucoviscous, Klebsiella pneumoniae, necrotizing fasciitis.

Introduction

Necrotizing fasciitis (NF) is an acute, rapidly progressive and invasive infection that spreads along fascial planes causing extensive inflammation and necrosis of muscles and adjacent soft tissues. NF was first described by Joseph Jones, an army surgeon during the American Civil War, but was made popular after being introduced as an infection caused by 'flesh eating bacteria' in the 1990s. NF can affect various parts of the body, but NF of the head and neck is rare. Klebsiella pneumoniae (K. pneumoniae) is a gram-negative rod that commonly causes hospital-acquired pneumonia, urinary tract infections and bacteremia. The high mortality associated with extended spectrum beta-lactamase (ESBL) and carbapenem resistant enterobacteriaceae (CRE) species of K. pneumoniae is probably due to the lack of effective treatments and underlying morbidities of the affected patients, rather than virulence of the bacteria. We hereby report a case of neck fasciitis caused by a strain of K. pneumoniae called hypermucoviscous K. pneumoniae. Unlike CRE and ESBL, this strain is commonly found in the community and despite being sensitive to almost all antibiotics, is extremely lethal due to its high virulence.

From Department of Anesthesia and Critical Care, Japanese Red Cross Nagoya Daini Hospital, Nagoya City, Aichi Prefecture, Japan (U Wen Yeap).

Address for correspondence:
U Wen Yeap, MD EDIC
Department of Anesthesia and Critical Care
Japanese Red Cross Nagoya Daini Hospital
2-9, Myoken-cho, Showa-ku, 466-8650, Nagoya, Aichi Prefecture, Japan
Tel: +81-52-832-1121
Fax: +81-52-832-1130
Email: uwyeapmd@nagoya2.jrc.or.jp

Case presentation

A 56-year-old male presented to our emergency department (ED) with a three-day history of swollen neck. He had a history of hypertension, type 2 diabetes mellitus, and liver cirrhosis secondary to non-alcoholic steatohepatitis. He was a non-smoker and he never drank alcohol. His medication included sitagliptin, voglibose, amlodipine and aldactone. He denied any history of intravenous drug use and unsafe sexual practices. He had no recent travel history or sick contacts.

The swelling of his neck started three days prior and worsened over the next few days. He denied

any recent trauma or flu. He had difficulty swallowing solid food, but denied any breathing difficulty. On the day of presentation, he drove himself to the ED. His vital signs on arrival to the ED were a temperature of 36.5 ℃, blood pressure of 140/72 mmHg, sinus tachycardia of 108 beats per minute, respiratory rate of 24 breaths per minute with an oxygen saturation of 94% on room air. On examination the patient was in moderate distress and appeared ill. His skin and conjunctiva were icteric. His throat was not injected, but the anterior neck was grossly swollen, extending down to his left clavicle. The swelling was warm, tender and hard. His cardiovascular and respiratory examinations were unremarkable. His abdomen was distended, but not tender. A rhino-laryngoscopy was performed in the ED which revealed a normal epiglottis and patent upper airway.

Bloodwork on initial presentation to the ED revealed leukocytosis (13,800 cells/μl). C-reactive protein (14.6 mg/dl), serum creatinine (3.01 mg/dl), and lactate (32 mg/dl) were elevated. Urinalysis was negative for nitrates and leukocytes. The patient did not show signs of encephalopathy, but a point-of-care abdominal ultrasound showed mild ascites. Total bilirubin was 2.9 mg/dl, albumin was 1.5 g/dl, and prothrombin time-international normalized ratio (PT-INR) was 2.9, leading to a total Child-Pugh score of 10 (Grade C).

Two hours later, he became hypotensive requiring massive fluid resuscitation and high dose vasopressors. He also developed respiratory distress and was immediately brought to the operating theatre for semi-awake intubation as difficult airway was anticipated due to his grossly swollen neck.

A contrast enhanced pan-computed tomography (CT) after intubation revealed a hypodense area in the retropharyngeal space, but no other findings suggestive of any other focus of infection or abscess could be found.

The patient was admitted to the ICU with a diagnosis of septic shock associated with a possible retropharyngeal abscess. After ICU admission, he continued to require high doses of vasopressors. He was continued on meropenem and vancomycin, which were started in the ED. Continuous hemodiafiltration (CHDF) using a cytokine-adsorbing hemofilter (Sepxiris, Gambro Industries) was initiated for renal indications at a dose of 10 ml/kg/hr (800 ml/hr is the maximum dose allowed by the Japanese National Health Insurance Scheme). Despite fluid resuscitation and continuous renal support, the patient's metabolic acidosis failed to improve and his lactate reached 130 mg/dl 12 hours after ICU admission. Several hours later, the left side of the patient's swelling had further deteriorated, developing petechiae and blisters (**Figure 1**). Concern for necrotizing fasciitis was immediately raised and the spinal and Ear-Nose-Throat (ENT) services were consulted for surgical indications. Although a clinical diagnosis of necrotizing fasciitis was made, surgical debridement of the area involved was deemed too difficult and beyond the surgical skill capacity of both the spinal and ENT services in our hospital. Unfortunately, before a final decision could be made, the patient went into a cardiac arrest and died within 24 hours after ICU admission.

Discussion

Necrotizing fasciitis (NF) of the head and neck is rare, but is associated with a higher mortality rate than NF in the extremities, trunk, or abdomen. (1) Diagnosis of NF in the neck is challenging due to its rarity and the fact that in the early stages, it can be mistaken for a more common neck abscess, which has a more benign course. In this case, retropharyngeal abscess was initially suspected. However, needle aspiration efforts on the day of admission and the next day, targeting the hypodense area in the CT, were unsuccessful. A clinical diagnosis of NF was made after the skin manifestations became more apparent on the second day. Nevertheless, the CT findings which included the thickening and infiltration of the cutis and subcutis, diffuse thickening of the sternocleidomastoid fascia, and fluid collections in the surrounding compartments were consistent with NF (**Figure 2**).

NF can be categorized into three distinct types. Type 1 is a polymicrobial infection caused by mixed aerobic and anaerobic bacteria. Type 3 is a gas gangrene caused by Clostridium perfringens and Vibrio vulnificus. Type 2 is caused by a monomicrobial infection, predominantly by group A Streptococcus (GAS), but cases of type 2 NF caused by K. pneumoniae have been increasingly reported worldwide. (2) However, to our best knowledge, this is the first report of type 2 neck NF caused by hypermucoviscous K. pneumoniae, which was confirmed with genotyping.

Hypermucoviscous K. pneumoniae has several strains, which are categorized into capsular serotype K1 and non-K1 based on their clinical characteristics. K1 strains are more virulent in terms of concomitant distant abscess complications and higher in vitro resistance. The different strains are separated by several virulence related gene profiles such as the rmpA, magA, iutA and ybtS. (3) Multilocus Sequence Typing (MLST) analysis of

the isolate from our patient confirmed the presence of capsular serotype K2, rmpA, ybts, mrkD, entB, and iutA alleles indicating that the isolate belonged to the Sequence Typing (ST) 65 strain (non-K1) (**Figure 3**). Genotyping tests, while being cost and time consuming, can only be of epidemiological significance. On the contrary, a simple 'string test' can confirm the hypermucoviscosity phenotype of K. pneumoniae. A positive 'string test' is defined as the formation of viscous strings of more than 5 mm in length when a loop is used to stretch the colony on an agar plate. (4) The test has a turnover time short enough to confer clinical significance and thus, in our opinion, should be performed on all isolates especially those from high risk patients.

Although extensive debridement remains the priority of NF treatment, NF of the neck is extremely challenging due to its complex anatomy and proximity to the mediastinum. (5) In cases caused by hypermucoviscous K. pneumoniae, clinical deterioration can happen very quickly as demonstrated in this case. Hence, high clinical suspicion in high risk patients is vital for early diagnosis of this deadly disease, which should in turn trigger a more rapid and aggressive debridement, complemented with empiric broad spectrum antibiotics and adjunctive treatment of sepsis, without any delay.

Conclusion

Hypermucoviscous K. pneumoniae infection should be suspected in high risk patients, such as those with liver disease or diabetes mellitus, presenting with rapidly progressive sepsis. While genotyping is important for epidemiological purposes, we strongly advocate performing the 'string test' on all K. pneumoniae isolates as confirmation results of hypermucoviscous K. pneumoniae in the lab may be able to warn the clinicians of the impending rapid deterioration in time for more aggressive management of the patients.

Figure 1. Diffuse swelling of the neck extending down to the left shoulder and chest with petechiae and blisters

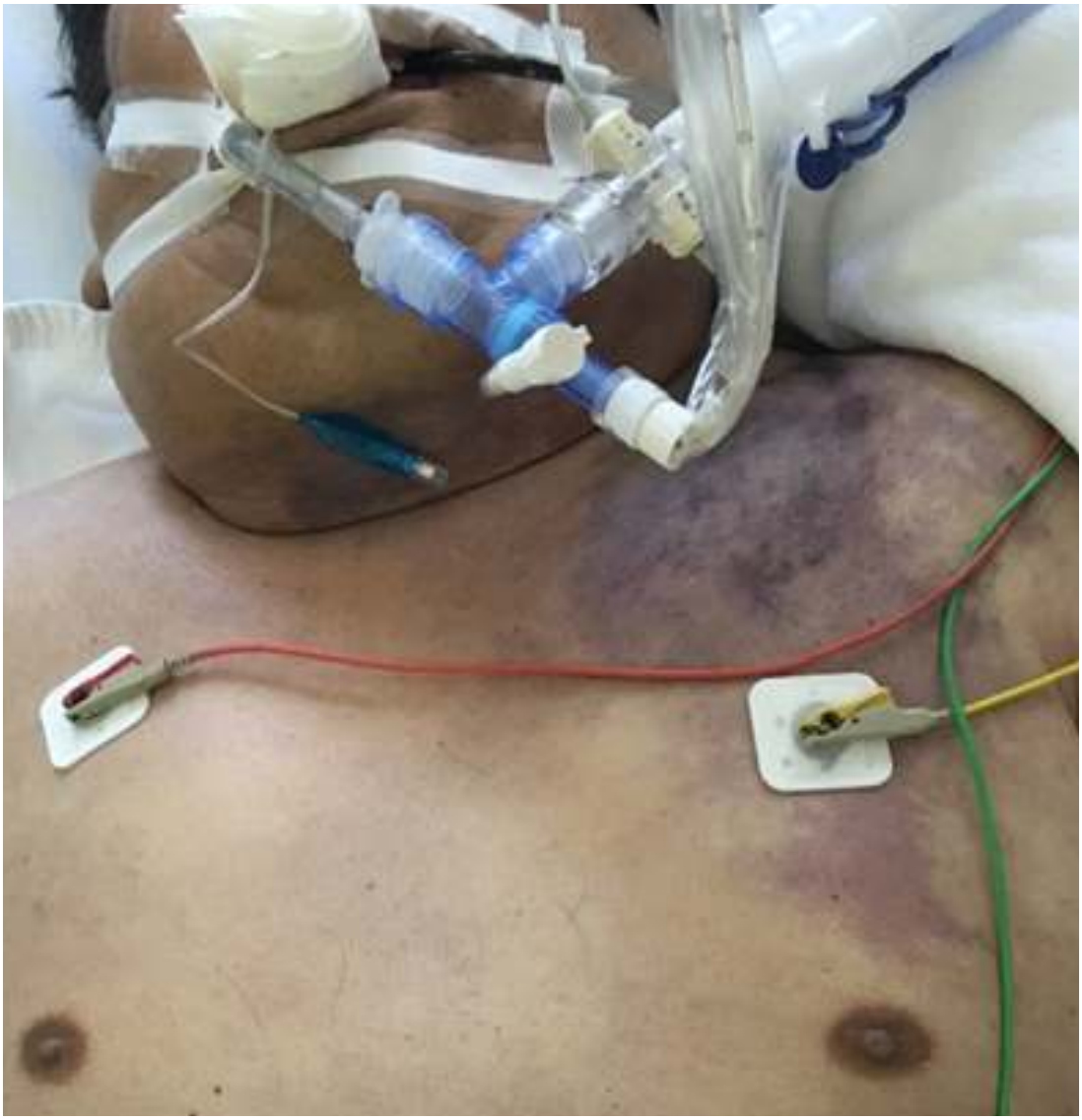


Figure 2. CT of the neck performed on the day 1. (a) Thickening and infiltration of the cutis and subcutis (⇦⋯), diffuse thickening of the sternocleidomastoid fascia (⇦) and fluid collections(*). (b) Hypodense area in the retropharyngeal space (⇦)

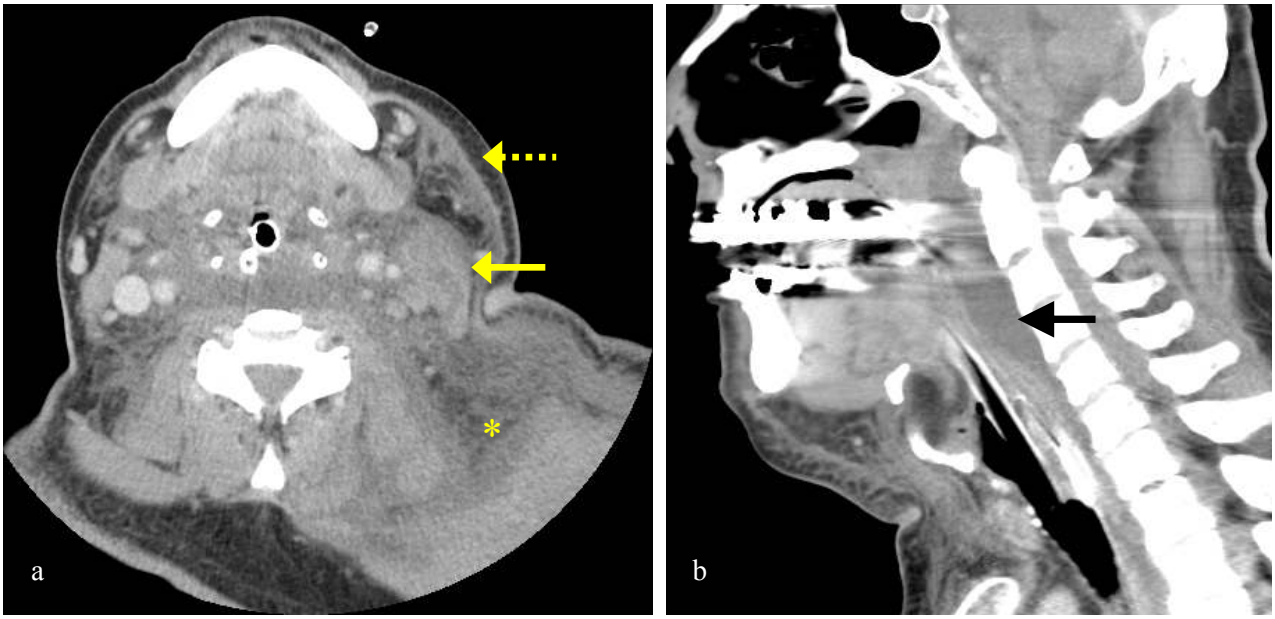
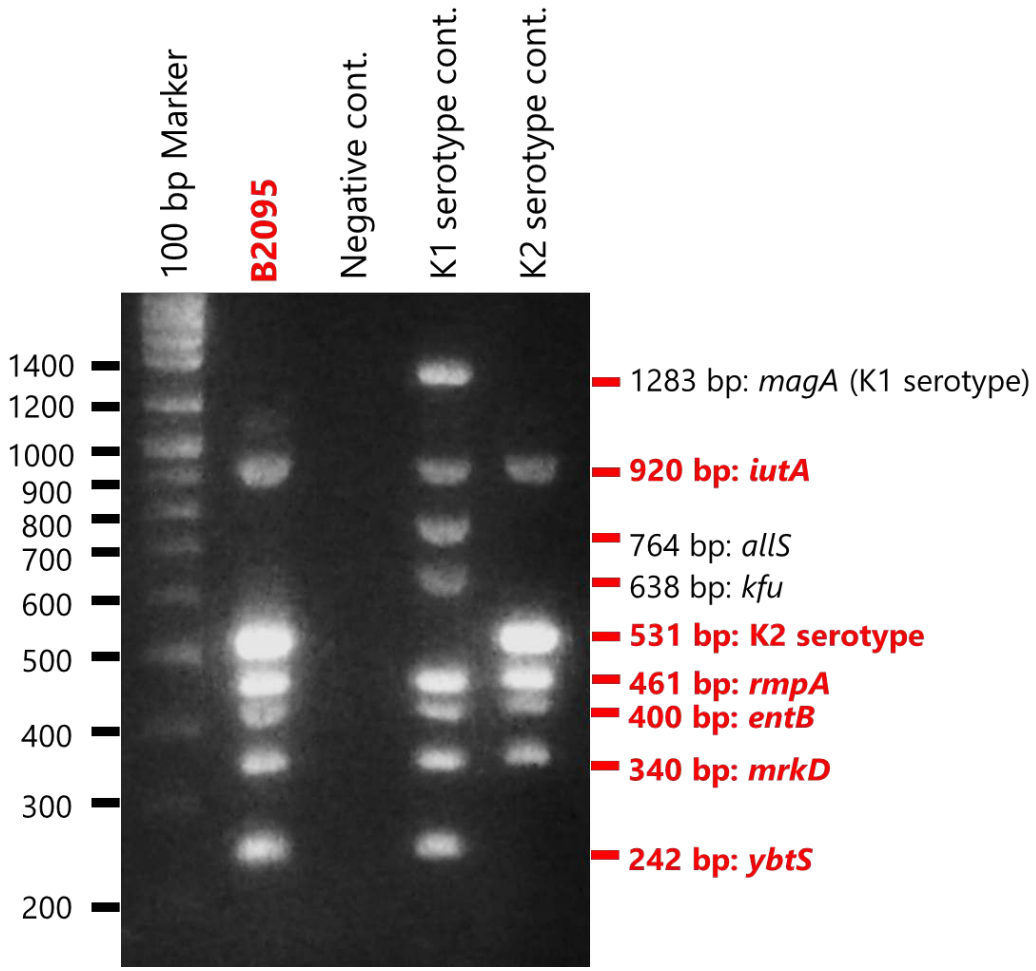


Figure 3. Multilocus Sequence Typing (MLST) analysis of the isolate from our patient. Capsular serotype K2, rmpA, ybts, mrkD, entB, iutA alleles were confirmed, indicating that the isolate belonged to the Sequence Typing (ST) 65



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