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Case Report

The First Case Report in the Middle East and Literature Review of a Cystic Fibrosis (CF) Infant with Recurrent *Staphylococcus hominis* and Methicillin Resistant *Staph aureus* (MRSA) Sepsis

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Abstract

Background: Cystic Fibrosis (CF) has been described before in Saudi Arabia and all Arab countries. The presentation is usually recurrent chest infection, failure to thrive and electrolyte imbalance. Sepsis is a rare presentation in such group of patients due to normal immunological status.

Objective: To describe the first case report of CF infant presenting as recurrent *Staphylococcus hominis* and Methicillin Resistant *Staph aureus* (MRSA) sepsis with recurrent chest infection despite normal immunological workup.

Method: A case report and literature review of an infant presenting with recurrent septicemia and negative immunological work up with positive CFTR gene.

Results: A 7 months old infant presented to the local hospital with recurrent fever, diarrhea, failure to thrive with Z score for weight/Age of (-8.2) and recurrent pneumonia monthly since the age of 2 months. He was referred to our center to rule-out immunodeficiency. He developed hypoxic respiratory failure requiring Intubation for 3 weeks with endovascular infection with polymicrobial central line associated blood stream infection (CLABSI) and persistent positive cultures of MRSA septicemia. He developed tension pneumothorax on top of parenchymal lung disease that required chest tube drainage for 10 days. His Broncho alveolar lavage grew: *Klebsiella pneumoniae*, *Stenotrophomonas maltophilia* and *Candida parapsilosis* in addition to Influenza B virus and RSV. His sepsis improved with the appropriate antimicrobial treatment and weight improving to Z score of -5.9 after one month of nutritional support and CF management. He was discharged in a good clinical condition. His immunological workup was normal but the whole genome study confirmed the diagnosis of CF with homozygous c.1375_1383del. His weight Z score at 2 years was (-1.02).

Several case reports in the literature describe CF presenting as sepsis in pediatric patients. One report describes three infants who initially presented with pneumonia and septic shock which led to their CF diagnosis. Such presentation in uncommon. 5 publications reported similar presentation with total of 8 infants.

Conclusion: CF could present with recurrent sepsis. Proper antimicrobial treatment and detailed history could prevent prolonged morbidity and mortality of this group of patients.

Keywords: *Cystic Fibrosis Septicemia; Staphylococcus hominis; CF MRSA Sepsis*

Introduction

Cystic fibrosis (CF) is an autosomal recessive disease characterized by pancreatic insufficiency and chronic endobronchial airway infection [1]. It affects approximately 89,000 people worldwide and is associated with a spectrum of systemic disease leading to reduced life expectancy [2].

CF has been reported before in Saudi Arabia with different cystic fibrosis transmembrane conductance regulator (CFTR) gene mutations compared to north America and Europe [3-7].

While respiratory infections are common in CF, systemic infections leading to septicemia are less frequently reported as the primary presenting symptom. Septicemia, a life-threatening condition resulting from the body's extreme response to infection, requires prompt diagnosis and treatment [8].

In 1980 [9], there was 2 reported cases of cystic fibrosis patients who presented with bacteremia in their first year of life at the ages of 7 and 2 months respectively. Their immunological work up was unremarkable. Both presented with respiratory symptoms and their blood culture was positive for *Staphylococcus aureus* for the 7 months old infant and *Serratia marcescens* for the second one.

Our case is the first case to be reported in the middle east with such presentation.

Objective of the Study

We describe the first case report and literature review in the middle east of CF patient presenting as recurrent *Staphylococcus hominis* and MRSA sepsis with recurrent chest infection.

Methods

A retrospective case report of an infant presenting with recurrent septicemia and negative immunological work up with positive CFTR gene in addition to a review of literature of similar presentation.

Case History

This is a case of 7 months old full term baby boy, who was born via normal spontaneous vaginal delivery, his birth weight was 1.9 kg (below 3rd centile, z score – 3.5) with no history of NICU admission.

At the age of 2 months, the mother noticed ecchymosis after a minor trauma, she went to local hospital but was reassured. After few days he was admitted to the hospital with acute gastroenteritis and fever, found to be pancytopenic and required multiple blood transfusions. Patient was also found to have elevated liver enzymes, disseminated intravascular coagulation (DIC) and septicemia with blood culture showing *Staphylococcus hominis* (*S. hominis*). He was admitted for 1 month for intravenous (IV) antibiotics. He was discharged in a good health with only mild persistent cough. He was admitted around 5 times due to recurrent chest infections, fever and recurrent positive blood culture for *S. hominis*.

Since birth patient had history of on/off diarrhea around 4 - 5 times/day. There was no history of oral thrush, fungal skin infections or deep organ infection/abscess. He had no previous history of recurrent ear/sinus infection. There was no history of delayed umbilical cord separation. He had some mild developmental delay with no other neurological symptoms. Patient was fully vaccinated till the age of presentation with no history of previous surgeries other than circumcision.

Mother and father are first degree relatives, they have 3 healthy siblings. There was history of abortion once with no family history of immunodeficiency, metabolic or hematological diseases.

His investigations from the referring hospital showed normocytic normochromic anemia with reactive lymphocytes. He had normal metabolic screen. Computed Tomography (CT) chest was showing cylindrical bronchiectatic changes. Patient was referred from a local hospital for further investigation to rule out Primary immune deficiency (PID).

At the age of 7 months, patient was seen in allergy/immunology clinic for evaluation, his clinical assessment at that time showed severe cachectic child with weight (3.5 kg, z-score -8.2) and height 54 cm, z-score -7.8) both below 3rd centile (Figure 1). His saturation was maintained on room air, he had mild tachypnea, clear chest and crepitations bilateral with unremarkable other systemic examination. Patient was planned for admitted for nutritional support and for further investigation.

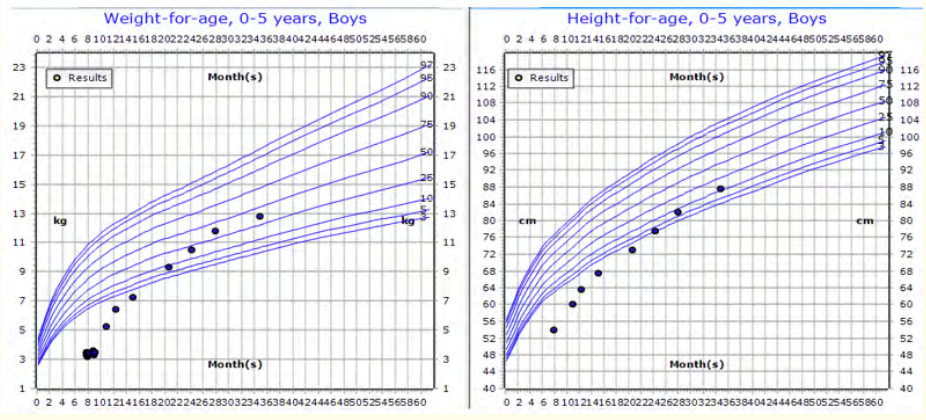

Figure 1: *Growth chart of a patient with CF septicemia showing improvement with proper CF treatment.*

He was assessed by swallowing team: patient had functional oral phase with normal modified barium swallow and no signs of silent aspirations. His immunological work up came to be unremarkable.

After 5 days of admission, he developed tachypnea and severe respiratory distress with grunting and mottled skin, he was shifted to PICU due to hypoxic respiratory failure secondary to new onset pneumonia and required intubation. His CXR showed perihilar bronchial wall thickening, with new infiltrates and consolidation in the right upper lobe (Figure 2).

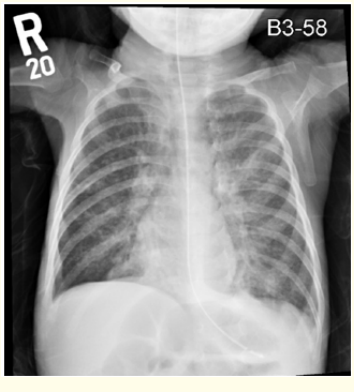

Figure 2: *Chest x-ray of a patient with CF septicemia at presentation showing: perihilar bronchial wall thickening, with new infiltrates and consolidation in the right upper lobe.*

Patient was managed as hospital acquired pneumonia with possible septicemia as patient was continuously spiking fever. Patient was started on tazocin and vancomycin initially, then bactrim was added to cover pneumocystis carinae pneumonia.

During his PICU stay, his initial venous blood gas (VBG) showed PH 7.71, PCO_2 was 21 mmHg (2.8 kPa) with bicarbonate of 26.6. CBC showed mild leukocytosis with neutrophil predominance, renal profile, hepatic profile and bone profile were within normal limits. His ADEK vitamin levels were low, Vitamin A 117 (N = 343 - 838), Vitamin E <2.2 (N = 5.5 - 15.5), Vitamin D was 15. He had prolonged PT 20.8 (N = 11.5 - 15.3), INR 1.6 (N = 0.8 - 10.2). His CRP was elevated and fluctuating (between 0.5 - 103.5). He had +ve nasal and skin MRSA screening.

Patient had +ve cultures with poly microbial sepsis, he had MRSA, *Acinetobacter pittii* and *Staphylococcus aureus* isolated from blood. His respiratory culture/tracheal aspirate isolated MRSA, *Staphylococcus aureus, Stenotrophomonas maltophilia* and *Klebsiella pneumoniae*. His urine culture remained negative and there was no evidence of vegetation on echocardiography.

Bronchoalveolar Lavage (BAL) was done and showed *Stenotrophomonas maltophilia, Klebsiella aerogenes*-Extended spectrum Beta lactamase (ESBL). Body fluid showed RBC 3, WBC 1,870 (neutrophilia) with positive RSV and Influenza B Virus. Fungal culture isolated many *Candida parapsilosis*.

Over his PICU stay, patient had a stormy course starting with pneumonia that was complicated with ARDS. Child had hyperinflated lung with emphysematous changes in addition to non compliant lung requiring higher ventilatory setting. This critical course was complicated with air leak and pulmonary hemorrhage. In addition, he had overwhelming sepsis with disseminated MRSA infection (respiratory and blood). Patient had persistent polymicrobial bacteremia that was complicated with central line associated bloodstream infection (CLABSI) despite tailored antibiotic therapy based on sensitivity. Patient received meropenem, vancomycin, gentamicin, bactrim and caspofungin. Patient was extubated successfully after 15 days of mechanical ventilation and weaned gradually to room air.

His whole exome sequence (WES) result came positive for a confirmed variant in CFTR with homozygous c.1375_1383delp.(Ser459_Gly461del) and heterozygous (a carrier for RAG2:NM_001243786.1:exon3:c686G>C:p.Arg229Pro) mutation for immunodeficiency.

Patient started on airways clearance techniques along with ADEK vitamin supplement as patient was having prolonged coagulation which is caused by vit K deficiency related to his primary disease (i.e. CF). Patient was also started on pancreatic enzymes. He was discharged after 40 days in a good and stable condition. He was seen in our clinic at the age of 10 months, patient was doing well, gaining weight with no concerns (Figure 1).

Reference Number	Study Author	Year	Population Age	Clinical Presentation	Organism isolated from blood culture
[9]	Michael M., *et al.*	1980, USA	1. Seven months 2. Two months	1. Dyspnea, wheezes and RCI 2. Meconium ileus, admitted with pneumonia	1. Coagulase- positive *S. aureus* 2. *Serratia marcescens*
[10]	Eman S. Al-Khadra, MD., *et al.*	2012, USA	1. Five months 2. Four month 3.32-days	1. Lethargy, decrease oral intake, moderate RD, leukocytosis and RUL infiltrate. 2. Fever, poor feeding, SOB and desating. CXR: RUL infiltrate. 3. Cough, poor oral intake, pallor, apnea, cyanosis and low O_2 sat. CXR: complete opacification of right lung. -All 3 cases required intubation and mechanical ventilation. -Had normal immunological work up, low albumin, low fat-soluble vitamin levels. -FTT with improvement in their weight after starting proper management.	1. *Pseudomonas aeruginosa* 2. *S. aureus* 3. *S. aureus*

[11]	Fernando Gatti Menezes	2013, Brazil	Preterm 30 weeks	Admitted to NICU due to RD, midgut volvulus, laparotomy twice with bowel resection.	*Ochrobactrum anthropic*
[12]	Amit Jain, Olufunke Afolabi-Brown	2016, Philadelphia	2.5 Y/O	Fever, decrease oral intake, low urine output, RCI. CXR: bilateral basal irregular airspace opacities.	*S. aureus*
[13]	Jogender Kumar., *et al.*	2020, India	2 months old	Progressive pallor, generalized body swelling and pale stool since neonatal period. Died due to severe sepsis, shock, and pulmonary hemorrhage. An autopsy confirmed the diagnosis of CF.	*S. hominis*

Table 1: *Literature review of CF septicemia at presentation.*

RCI: Recurrent Chest Infection; S. aureus: Staphylococcus aureus; RD: Respiratory Distress; SOB: Shortness of Breath; RUL: Right Upper Lobe; FTT: Failure to Thrive; CF: Cystic Fibrosis.

Discussion

This case report highlights an unusual presentation of cystic fibrosis (CF) with recurrent septicemia, which aligns with the findings of the study by Cargill and Etherington (2012) [14]. Their research identified 57 clinically significant episodes of blood stream infections (BSIs) in 48 CF patients over an 8-year period. The most common causative agents were: Coagulase-negative staphylococci, *Candida* species and *Stenotrophomonas maltophilia*.

These findings underscore the importance of considering a wide range of pathogens when evaluating septicemia in CF patients. Notably, 82% of significant blood stream infections (BSIs) were attributed to totally implantable vascular access devices (TIVADs), while only 9% originated from the lower respiratory tract. This emphasizes the need for vigilant care and monitoring of vascular access devices in CF patients. The study reported favorable outcomes for totally implantable vascular access devices (TIVAD) associated BSIs when managed with targeted antimicrobial therapy and appropriate device removal. In contrast, blood stream infections (BSIs) originating from the lower respiratory tract were associated with poor clinical outcomes, with a 60% mortality rate [14].

This case report and the supporting study highlight several key points such as: 1. The importance of considering septicemia as a potential complication in CF patients, even if it's not the typical presentation. 2. The critical role of prompt, targeted antimicrobial therapy and device removal when indicated. 3. The potential prognosis of BSIs originating from the lower respiratory tract in CF patients.

Our case report of CF presenting with recurrent septicemia aligns with the findings of Cargill and Etherington, emphasizing the need for heightened awareness of this potential complication in CF patients. It underscores the importance of thorough evaluation and aggressive treatment of BSIs to improve outcomes in this vulnerable patient population.

Conclusion

Recurrent septicemia is an infrequent yet serious complication in cystic fibrosis patients, typically arising from the chronic lung infections that characterize the disease. Findings underscore the importance of newborn screening programs, early diagnosis, and

comprehensive care for infants with CF. Clinicians should remain vigilant for atypical presentations of CF, including sepsis, particularly in regions where screening is not routinely performed or in cases missed by screening.

Bibliography

1. Goetz D and Ren CL. "Review of cystic fibrosis". *Pediatric Annals* 48.4 (2019): e154-e161.
2. Ong T and Ramsey BW. "Cystic fibrosis: A review". *Journal of the American Medical Association* 329.21 (2023): 1859-1871.
3. HH Banjar., *et al*. "Genotype patterns for mutations of the cystic fibrosis transmembrane conductance regulator gene: a retrospective descriptive study from Saudi Arabia". *Annals of Saudi Medicine* 40.1 (2020): 15-24.
4. H Banjar., *et al*. "Double homozygous Cystic Fibrosis Transmembrane Regulator gene (CFTR) mutation: A case series and review of the literature". *Global Journal of Rare Diseases* 5.1 (2020).
5. H Banjar., *et al*. "The first report of Methicillin-resistant *Staphylococcus aureus* (MRSA) in cystic fibrosis (CF) patients in Saudi Arabia". *International Journal of Pediatrics and Adolescent Medicine* 7.4 (2020): 186-190.
6. H Banjar and G Angyalosi. "The road for survival improvement of cystic fibrosis patients in Arab countries". *International Journal of Pediatrics and Adolescent Medicine* 2.2 (2015): 47-58.
7. Banjar Hanaaa., *et al*. "Geographic distribution of cystic fibrosis transmembrane conductance regulator (CFTR) gene mutations in Saudi Arabia". *International Journal of Pediatrics and Adolescent Medicine* 8.1 (2021): 25-28.
8. Sepsis Alliance. "Sepsis and Cystic Fibrosis" (2023).
9. McCarthy MM., *et al*. "Bacteremia in patients with cystic fibrosis". *Clinical Pediatrics* 19.11 (1980): 746-748.
10. Heather Hoch., *et al*. "Clinical outcomes in U.S. infants with cystic fibrosis from 2001 to 2012". *Pediatric Pulmonology* 53.11 (2018): 1492-1497.
11. FG Menezes., *et al*. "Ochrobactrum anthropi bacteremia in a preterm infant with cystic fibrosis". *Brazilian Journal of Microbiology* 45.2 (2014): 559-561.
12. A Jain and O Afolabi-Brown. D108. Pediatric cases: a room full of zebras (2016).
13. Kumar J., *et al*. "An infant with severe anemia and hypoalbuminemia". *Indian Pediatrics* 57.4 (2020): 349-355.
14. J Cargill., *et al*. "Bloodstream infections in cystic fibrosis: Nine years of experience in both adults and children". *Journal of Cystic Fibrosis* 11.4 (2012): 337-339.

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