Neonatal sepsis and multiple skin abscess in a newborn with Down’s syndrome: A case report

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CASE REPORT

Please cite this paper as: Kali A, Umadevi S, Srirangaraj S, Stephen S. Neonatal sepsis and multiple skin abscess in newborn with Down’s syndrome: A case report. AMJ 2013, 6, 2, 91-93. http://dx.doi.org/10.4066/AMJ.2013.1574

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Abstract

Neonatal sepsis is a leading cause of neonatal mortality. Congenital heart disease accounts for additional risk of sepsis in neonates. Here we report a case of Down’s syndrome with late onset neonatal sepsis associated with multiple superficial skin abscesses simulating staphylococcal infection. The baby was empirically treated with vancomycin. Subsequently, multidrug resistant Klebsiella pneumoniae was detected from both pus and blood culture. Change to appropriate antibiotic resulted in clinical recovery. Although sepsis is one of the major ailments in neonates, atypical presentations of neonatal sepsis in Down’s syndrome patients are underreported. Here we highlight the atypical presentation of Klebsiella sepsis and the importance of early antibiogram in such cases.

Key Words
Down’s syndrome; Klebsiella pneumoniae; Extended spectrum beta-lactamase; Neonatal sepsis

Implications for Practice

1. Down’s syndrome affects one out of 800 newborns worldwide. Neonatal sepsis is a leading cause of neonatal mortality accounting for 12% of total neonatal deaths in India. The presence of sepsis increases mortality during the first year of life in Down’s syndrome patients. Infants with CHD (Congenital heart disease) have a higher incidence of culture proven sepsis and mortality. Early proper antibiotic treatment can significantly alter the outcome.
2. In this case report, a newborn with Down’s syndrome developed late onset neonatal sepsis with multiple skin abscesses due to extended spectrum beta-lactamase (ESBL) produced by K. pneumoniae. Although, pus culture, blood culture and elevated CRP were conclusive, absence of typical features of sepsis delayed the diagnosis and culture based antibiotic therapy.
3. Laboratory tests are critical to diagnose neonatal sepsis. Congenital heart disease in neonates with Down’s syndrome should be considered an additional risk factor of neonatal sepsis, which, if not detected early and treated judiciously, may worsen the prognosis.

Background

Neonatal sepsis is a leading cause of neonatal mortality accounting for 12% of total neonatal deaths in India. Early initiation of proper antibiotic can significantly alter the outcome. However, early diagnosis is often challenging due to nonspecific or atypical presentations. There are very few reports of atypical presentations of neonatal sepsis in Down’s syndrome. Skin and soft tissue involvement is a unique presentation in neonatal sepsis caused by K. pneumoniae and has not yet been reported in patients with Down’s syndrome.

Case details

A 30-year-old lady delivered a 2.8kg male baby by Caesarean section and was referred to our institute on the eighth postnatal day for diagnosis and management of congenital defects. On admission, the newborn presented with mongoloid facies, short neck with loose skin, depressed bridge of nose and decreased muscle tone. A soft ejection systolic murmur was audible over apical and pulmonary areas. Subsequently it was found to be due to mild atrial septal defect by echocardiography. It was clinically diagnosed as Down’s syndrome with a cyanotic congenital heart disease. No other congenital defects were detected in clinical examination and laboratory
involvement simulating staphylococcal infection. Localised impetigo and umbilical sepsis is most commonly caused by infection in neonate in the form of pustule, abscess, staphylococcal infection. Furthermore, superficial skin pus collection with signs of inflammation is characteristic of manifestations also have been observed in neonatal sepsis caused by Klebsiella pneumoniae. All isolates displayed same antimicrobial susceptibility pattern. They were resistant to ampicillin, ceftriaxone, piperacillin, gentamicin and co-trimoxazole but sensitive to ciprofloxacin, piperacillin-tazobactam, amikacin and imipenem. Combination of amikacin and imipenem starting from 15th day of birth resulted in cure of lesions along with negative blood and pus culture and a drop in CRP level.

**Discussion**

Late onset neonatal sepsis (LONS) is defined as sepsis developing after the third day of birth and is mostly due to acquisition of infection from caregiving environment. The pathogens usually colonise in the skin, umbilical stump, respiratory and gastrointestinal tract facilitated by factors like long hospital stay, vascular or urinary catheters, indwelling lines, H2 blockers or proton pump inhibitors and gastrointestinal pathology. LONS can present without characteristic signs, therefore laboratory parameters are critical. Elevated CRP and thrombocytopenia were reported to be associated with LONS. However, positive blood culture in presence of clinical parameters is more confirmatory. Gram negative *K. pneumoniae* has been documented frequently in LONS. Atypical clinical manifestations also have been observed in neonatal sepsis caused by *K. pneumoniae*. Viswanathan et al reported *K. pneumoniae* sepsis associated with maculopapular rash. In this case we found skin and subcutaneous tissue involvement simulating staphylococcal infection. Localised pus collection with signs of inflammation is characteristic of staphylococcal infection. Furthermore, superficial skin infection in neonate in the form of pustule, abscess, impetigo and umbilical sepsis is most commonly caused by *Staphylococcus aureus*. Hence, it was initially suspected to be staphylococcal infection and empirical vancomycin therapy was also started. Later, the pus culture and sensitivity report confirmed that the causative agent was *K. pneumoniae*. The same strain of *K. pneumoniae* was isolated from blood culture with same resistance pattern. Furthermore, *K. pneumoniae* is well recognised for multidrug resistance and outbreak potential in nurseries. Our finding in this patient corroborates with other studies. ESBL production was detected in 32-92% isolates of *K. pneumoniae* from neonatal sepsis where only treatment option was carbopenem or higher aminoglycoside. Down’s syndrome affects one per 800 newborns. The high mortality of 25-30% reported among Down’s patients during the first year of life is mostly associated with severe conditions like congenital heart disease, bronchopneumonia and sepsis. CHD is reported in 40-50% of Down’s patients. Endocardial cushion defect, atrial and ventricular septal defect, Fallot’s tetralogy and patent ductus arteriosus are most commonly encountered heart malformation. In addition, infants with CHD have a higher incidence of culture proven sepsis and mortality. Intrinsic defect of the immune system may be related to increased risk of neonatal sepsis in Down’s patients who suffer from T and B cell lymphopenia, impaired T and B cell response and defects of neutrophil chemotaxis from the very beginning of their life. Although, *K. pneumoniae* have been implicated as a major agents of LONS in Down’s syndrome, no correlation has been established between *K. pneumoniae* sepsis and Down’s syndrome. Therefore, our finding may be coincidental and needs further studies. However, here we addressed the atypical presentation of neonatal sepsis and the diagnostic difficulty arising from it.

In conclusion, we have reported a rare case of neonatal sepsis in absence of typical symptoms in a newborn with Down’s syndrome. Laboratory test was critical to diagnose the condition in this case. Congenital heart disease in neonates with Down syndrome should also be considered as an additional risk factor for neonatal sepsis.

**References**


PEER REVIEW
Not commissioned.

CONFLICTS OF INTEREST
The authors declare that they have no competing interests.