Invasive Pneumococcal Disease Presenting as Purpura Fulminans in a Child with Mondini's Dysplasia

Purpura Fulminans ile Başvuran Mondini Displazi Tanılı Çocukta İnvaziv Pnömokokkal Hastalık

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Abstract

A case of Mondini's dysplasia who presented with purpura fulminans caused by *Streptococcus pneumoniae* is described in this case report. Invasive pneumococcal disease is usually related to immune deficiencies or anatomical defects of the upper respiratory tract, like Mondini's dysplasia in our case, which is a developmental anomaly of the middle ear. It is still controversial as to how to protect these people and prevent invasive pneumococcal disease. In this report we tried to discuss possible reasons of invasive infection despite chemoprophylaxis and vaccination. *(J Pediatr Inf 2011; 5: 74-6)*

Key words: Mondini's dysplasia, purpura fulminans, invasive pneumococcal disease

Özet

Bu makalede Mondini displazisi tanısı ile izlenen bir olguda *Streptococcus pneumoniae* etkeni ile gelişen purpura fulminans tablosu tanımlanmıştır. İnvaziv pnömokokkal hastalık, çoğunlukla altta yatan bir immun yetmezlik ya da olgumuzda varolon Mondini displazisi gibi ortakulak gelişim sorunu ve dolayısı ile solunum yolu ile ilişkili bir anatomik defekt varlığında gelişir. Bu olguların korunması ve invaziv pnömokokkal hastalık gelişimine karşı alınacak önlemler halen tartışmalıdır. Bu yazıda aşılama ve kemoprofilaksiye rağmen invaziv enfeksiyon gelişiminin olası nedenlerini tartışmaya çalıştık. *(J Pediatr Inf 2011; 5: 74-6)*

Anahtar kelimeler: Mondini displazisi, purpura fulminans, invaziv pnömokkal hastalık

Introduction

Purpura fulminans (PF) is a condition characterized by disseminated intravascular coagulation

(DIC) presenting with ecchymotic skin lesions, fever, bacteremia and hypotension. In the majority of cases, PF is usually associated with *Neisseria meningitidis* sepsis. However, even if it is a rare agent, *Streptococcus pneumoniae* can be responsible for purpura fulminans (1,2). Other encapsulated bacteria such as *Haemophilus influenzae* and group A and B streptococci have been implicated in infants and adults. *Staphylococcus aureus* as a cause of purpura fulminans is considered rare, probably because these cases are reported as toxic shock syndrome, but may be as common as other nonmeningococcal organisms (3).

Cases secondary to pneumococcal infections usually occur in those with underlying predisposing conditions such as congenital (primary) or acquired immunodeficiencies (e.g. HIV infection) or splenic dysfunction. In the pediatric age group, splenic dysfunction is the most frequent cause of invasive pneumococcal infections (4,5). However, other underlying factors like temporal bone defects, or congenital anomalies like Mondini's dysplasia are also possible risk factors for invasive disease. Here we present a patient who had Mondini's dysplasia with invasive pneumococcal infection related purpura fulminans.

Case

A five year old girl was admitted to the hospital because of fever. She had an implant in her

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left ear because of Mondini's dysplasia (Figure 1) and findings of inflammation were seen on her eardrum. Amoxicillin-clavunate therapy was started according to these findings but loss of consciousness in follow up led the physicians to consider the diagnosis of an intracranial infection. Parenteral antibiotherapy with ceftriaxone and vancomycin was started after a computerized tomography of the cranium was performed. Thereafter she was transferred to our hospital because of the presence of subarachnoid bleeding and sepsis. On admission, she was unconscious, dehydrated and was in septic shock. There were disseminated palpable purpuric lesions on her skin as a sign of purpura fulminans and disseminated intravascular coagulation. She was entubated and intraarterial blood pressure monitoring was started. Clinical and laboratory measurements showed signs of septic shock. She was treated with fluid replacement and antibiotics. Because of the septic state and subarachnoid bleeding; lumber puncture for diagnostic purposes could not be performed. Her blood pressure was high and antihypertensive drugs were needed to regulate blood pressure. Further tests to show possible agents of purpura fulminans and sepsis were first evaluated in urinary antigen test (BinaxNOW®) with *Streptococcus pneumoniae* antigen positivity. Also blood culture results revealed a penicillin resistance species of *Streptococcus pneumoniae* in the antibiogram. Other antibiotics such cephalosporins were effective for this agent. After adequate supportive treatment and antibiotherapy; the treatment was terminated within 2 weeks and she completely recovered in 3 months with continued physical therapy to prevent spasticity on both lower extremities, which was represented as a sequel after subarachnoid bleeding.

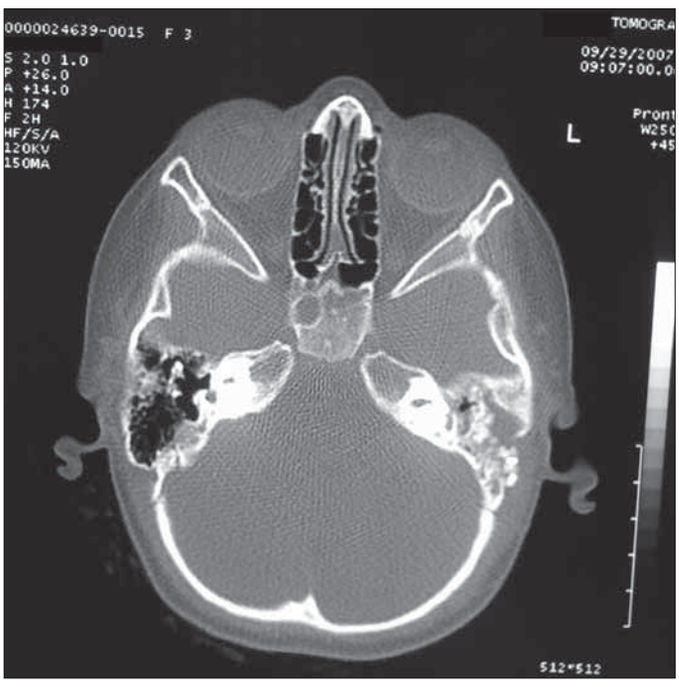

Figure 1. Left sided Mondini's dysplasia in the patient

Discussion

Our patient was diagnosed as invasive pneumococcal disease and purpura fulminans. Even if they are unusual agents in purpura fulminans, pneumococci are responsible for this disease if there is an underlying condition, and it is rarely described in healthy infants (6). In the presented patient the main risk factor was the Mondini's dysplasia, which is a developmental anomaly of the middle ear characterized by cochlear malformation with dilation of the vestibular aquaduct, vestibule and ampullar ends of the semicircular canals (7). She was operated on and an implant was placed in her left ear three years previously. the blood brain barrier was intact and there was no proof of cerebrospinal fluid (CSF) leakage even on examination after recovery. Also there was no proof of immunodeficiency; her immunoglobulin levels and antibody responses were consistent with her age group.

Like other temporal bone defects or other congenital anatomic causes of recurrent meningitis; in Mondini dysplasia the CSF leaks into the middle ear through a deficient foramen ovale, which is a risk factor for recurrent pneumococcal infections (8). Therefore prophylaxis plays an important role for preventng sequelae through central nervous system infections. However, the efficacy of prophylactic use of antibiotics in preventing meningitis in children could not be proved and might possibly do harm by increasing the risk of infection due to antibiotic-resistant organisms. Such use of antibiotics has been more often been recommended for patients with complement or immunoglobulin subclass deficiency.

Another preventive method is to vaccinate the patients. Meningococcal vaccine is recommended for patients who have terminal complement component deficiency and for those who have asplenia, but the vaccine's clinical efficacy has not been documented in such patients. Although the heptavalent pneumococcal conjugate vaccine is not sufficient for older children and adults in high risk groups, immunization with 23-valent polysaccaride vaccine is recommended for older children with recurrent infections (7). In our case, prophylactic penicillin was administered monthly before she was admitted to our hospital and she was vaccinated once with 23-valent polysaccaride vaccine. The question of whether it is possible to protect the patients with penicillin prophylaxis is controversial, because the responsible species of *Streptococcus pneumoniae* in our case is also resistant

to penicillin. Even there is no certain evidence, it can be hypothesized that previously used penicillin prophylaxis could be the reason for infection with a resistant strain and this strain is already the cause of invasive disease.

In conclusion we suggest that patients presenting with purpura fulminans should be examined for pneumococcal disease, especially if there is an underlying condition like Mondini's dysplasia.

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