IDCases 11 (2018) 36–38

Contents lists available at ScienceDirect

IDCases

journal homepage: www.elsevier.com/locate/idcases

Case report

# Case report: Guillain-Barre syndrome with pneumococcus – A new association in pediatrics

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ARTICLE INFO

*Keywords:*
Guillain-Barre syndrome
Pneumococcus
ARDS
Streptococcus
Molecular mimicry

ABSTRACT

Guillain-Barre Syndrome, an acute flaccid paralysis known to be caused by recent Gastro-intestinal infections mainly campylobacter, and Respiratory infections mainly mycoplasma pneumoniae and influenza. One reported case of severe invasive pneumococcal disease in a 68 year old female, that presented with Austrian's triad of meningitis, pneumonia and endocarditis, and progressed to develop Guillain Barre syndrome, an association never been documented before.

We present a case of 13 year old male, presented with hypoactivity and inability to bare his own weight, developed septic shock due to pneumococcus with Acute Respiratory Distress Syndrome, and was found to have neurological findings of Guillain-Barre Syndrome. A new association in pediatric age group, never been reported before.

## Introduction

Guillain-Barre syndrome (GBS) is an immune-mediated disorder of the peripheral nervous system which is triggered by either infectious or noninfectious factors [1]. It is the most common form acute flaccid paralysis at any age. Historically, Guillain-Barre syndrome (GBS) was considered a single disorder. It is now known to be a heterogeneous syndrome with several variant forms [2]. Controlled epidemiological studies have linked it to infection with *Campylobacter jejuni* in addition to viruses, including cytomegalovirus and Epstein Barr virus [3]. *Streptococcus pneumoniae*, a very important pathogen in the pediatric age group that causes many illnesses including pneumonia, acute otitis media and meningitis, has never been associated with GBS in the pediatric age group.

## Case presentation

A 13 year old male, previously known to have atopic dermatitis, not fully vaccinated, presented to our care for management of progressive fatigue, hypoactivity and slurred speech. 4 days prior to presentation when the patient started to complain of lower extremity weakness, that was progressive, associated with hypoactivity. Two days later, the patient started to have slurred speech, multiple episodes of choking, with both urinary and fecal incontinence. At the day of presentation, he started to have difficulty breathing, drooling of saliva and ascending weakness that led to inability to bear his weight so was presented for management.

Upon presentation to the Emergency Department he was conscious, alert, pale, and hypoactive. His vital signs: T: 36.4, P: 110, SBP: 70 with cold extremities and poor perfusion. He had respiratory distress with Silverman score 5/10, and Saturation: 89 on room air, bilateral decreased air entry with diffuse crackles. GCS was 15/15, decreased in motor power bilaterally in lower limbs with absent deep tendon reflexes, and a negative Babinski reflex. The rest of examination was within normal limits.

With a picture of septic shock, he received IV fluid boluses, his systolic blood pressure became 100, arterial blood gases showed hypoxemia with metabolic acidosis, and chest x-ray (Fig. 1) revealed right upper lobe collapse and pneumonia with diffuse infiltrates in both lung fields, picture of early ARDS. Non-invasive ventilation was applied, however he clinically deteriorated with a decreasing level of consciousness, so urgent CT scan of brain was done, which showed no bleeding or any mass, and the patient was then intubated and attached to mechanical ventilation. Urgent MRI Brain was done and was normal. Broad spectrum antibacterial stat doses were given and he was transferred to the pediatric intensive care unit.

Laboratory tests showed WBC 6000 (Bands 29, Neutrophils 47, Lymphocytes 13), with elevated inflammatory markers (CRP: 22, ESR: 110), with a picture of disseminated intravascular coagulopathy (Platelets: 57,000, INR: 1.4, Fibrinogen: 456, D-dimer: 0.36).

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https://doi.org/10.1016/j.idcr.2017.11.009
Received 26 July 2017; Received in revised form 29 November 2017; Accepted 29 November 2017

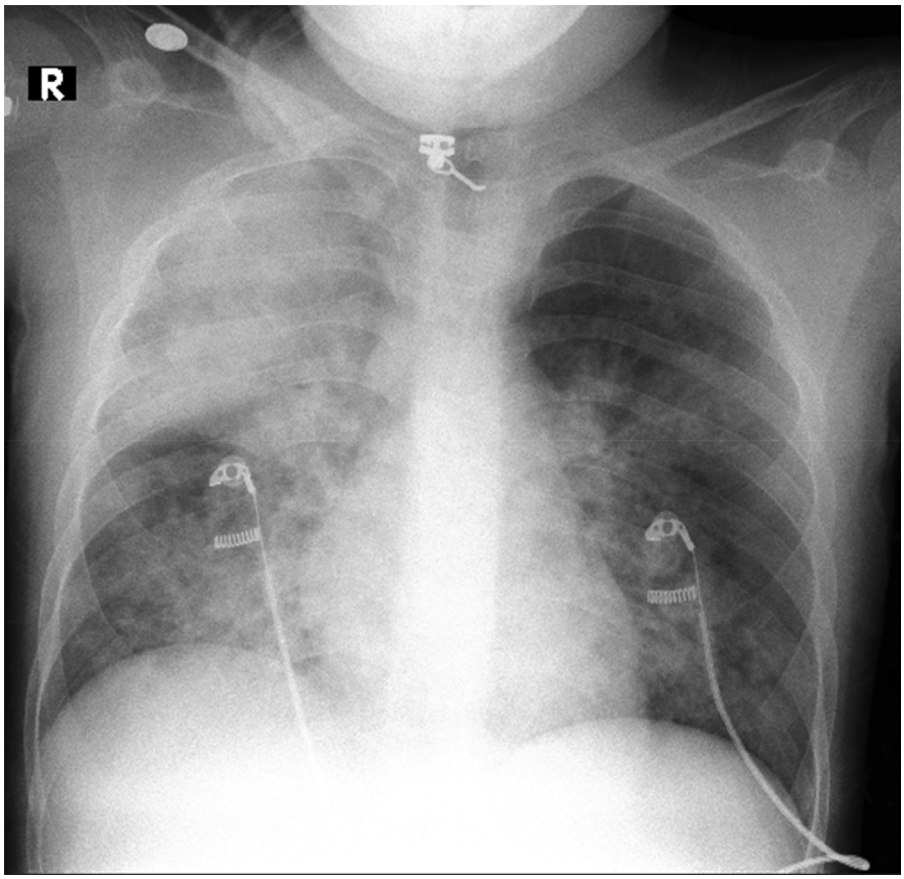


**Fig. 1.** Right upper lobe collapse and pneumonia with diffuse infiltrates in both lung fields, picture of early ARDS.

His history of inability to bear weight, that was followed by choking with his clinical deterioration suggested for the diagnosis of Guillain-Barre syndrome in particular especially because of absent deep tendon reflexes, so treatment with IVIG was begun. Lumber puncture couldn't be done due to his unstable cardiorespiratory status.

Blood cultures were positive for *Streptococcus pneumonia* after 48 h. Mycoplasma pneumonia serology and nasal wash for influenza were negative.

On day 4, he developed severe ARDS, hypoxia with respiratory acidosis, and his condition continued to deteriorate until he died.

## Discussion

*Streptococcus pneumoniae* (pneumococcus) is a very important pathogen that kills more than 1 million children each year [4]. *S. pneumoniae* is a Gram-positive, lancet-shaped, polysaccharide encapsulated diplococcus, occurring occasionally as individual cocci or in chains. More than 90 serotypes have been identified by type specific capsular polysaccharides. Encapsulated strains tend to cause most serious disease in humans. Capsular polysaccharides impede phagocytosis. Virulence is related in part to capsular size, but pneumococcal types with capsules of the same size can vary widely in virulence [4]. Infection is predominantly of the respiratory tract (Acute Otitis Media, sinusitis, and pneumonia); however, invasive disease may occur in the form of bacteremia, sepsis, meningitis, or other organ systems involvement [5].

Affected children may appear relatively well and have a mild to moderate illness course. However, they could also present acutely with abrupt severe illness, sepsis, purpura, disseminated coagulopathy, and progression to shock and multiorgan failure, especially in the setting of asplenia or sickle cell disease [5].

The routine use of a 7-valent (4, 6B, 9V, 14, 18C, 19F, and 23F) PCV (PCV7) in infants and children was followed by a significant reduction in overall invasive and respiratory pneumococcal infections [5]. Currently, the addition of other serotypes (1, 3, 5, 6A, 7F, and 19A) produced the 13-valent PCV. Our patient was not fully vaccinated and had not received any dose of pneumococcal vaccine, because of concerns about his atopic dermatitis.

In our case there is a new association between Invasive pneumococcal disease and Guillain-Barre Syndrome (GBS) in pediatrics. GBS is an important cause of acute flaccid paralysis, due to a triggering factor infectious (like mainly campylobacter, cytomegalovirus, Epstein-Barr virus, Mycoplasma pneumoniae, influenza-like illnesses), and non-infectious (like immunization, trauma, surgery and bone marrow transplant). This triggering factor evokes immune response that cross reacts with peripheral nerve components because of the sharing of cross-reactive epitopes (molecular mimicry), this immune response is directed toward the myelin or axon of the peripheral nerve.

It is possible therefore that pneumococcus has antigens which triggered an immune response and cross-reacted with peripheral nervous system surface components due to molecular mimicry. This is due to natural transformation of Streptococcus pneumoniae's capsular polysaccharide. This highlights the role of host immunity in determining clinical manifestations of disease; it is not solely due to bacterial virulence.

The diagnostic criteria for Guillain-Barre syndrome have not yet been defined and the condition is diagnosed primarily through a clinical history assessment and findings upon presentation [6]. The predominant complaint of the pediatric GBS includes weakness of the limbs, paresthesia, and pain [7], where our patient complaint of inability to bear weight. In 1994, Sarada and colleagues found that childhood GBS was associated with a higher incidence of cranial nerve palsy and had a more acute form of onset than adults [8], as seen in our patient had choking (absence of gag reflex, due to affection of either the glossopharyngeal nerve or the vagus nerve). These physical findings

could be supported by CSF elevated protein more than 45 mg/dl which is albumino-cytologic dissociation and electrodiagnostic studies, however, neither lumbar puncture or electrodiagnostic studies could be done in our patient due to the development of coagulopathy and his deteriorating cardio-respiratory condition.

A case was reported of a severe invasive pneumococcal disease in a 68 year old female, presented with uncommon but well documented complication of streptococcus pneumoniae bacteremia, Austrian's triad of meningitis, pneumonia and endocarditis. She then progressed to develop an atypical variant of Guillain-Barre syndrome, never previously documented in association with pneumococcal disease [9].

Another case was reported of a 78 year old male, presented with left basal lung infiltrates and a pleural effusion, developed 4 days later an acute symmetrical flaccid tetraparesis. Left sided pyogenic effusion culture was positive for *Streptococcus pneumoniae*. EMG showed a mixed type GBS, acute motor axonal and acute inflammatory demyelinating, and anti-GM1 and anti-GD1 were positive that induced a predominant motor form of GBS [10].

## Conclusion

Pneumococcus still one of the most challenging infections to clinicians, with variable range of presentation and clinical course severity. This case presents a new association between pneumococcus and a serious complication, Guillain-Barre syndrome, with high risk of morbidity and mortality.

Pneumococcus is a vaccine preventable disease, with documented efficacy of present vaccines in decreasing incidence and severity of invasive and respiratory pneumococcal disease. This sheds the light to implement effective vaccination programs.

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