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Case Report

Complicated Community Acquired Pneumonia and Air Leak Syndrome, Case Report

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Abstract

We are reporting a 7 months old girl, who had a febrile illness, signs of upper respiratory tract infection for couple of days, progressed rapidly to worsening respiratory manifestations, difficulty of breathing, unequal chest movement and hypoxia, physical examination reviled marked decrease air entry on RT side, chest xray showed Rt sided spontaneous pneumothorax and pulmonary air leak syndrome.

Patient required mechanical ventilator and had difficult ICU course with sever complicated community acquired pneumonia, spontaneous pneumothorax, progressed to necrotizing pneumonia and that was the first presentation of primary immunodeficiency disorder.

Keywords: *Community Acquired Pneumonia (CAP); Pulmonary Air Leak Syndrome; Necrotizing Pneumonia (NP); Primary Immunodeficiency Disorders (PID)*

Introduction

Overview of pneumonia

Pneumonia is a common disease in both adult and pediatric groups and can lead to significant comorbidity as well as mortality if not treated well. It's defined as acute infection of the lung parenchyma that is caused by numerous microorganisms mostly bacteria, viruses are important causative agents though. Pneumonia is further sub-divided into Hospital (HAP) and community acquired pneumonia (CAP). As the names imply, the former being caused by pathogens that are commonly found in the society such as *Streptococcus pneumoniae, Mycoplasma pneumoniae, Hemophilus influenzae* and respiratory viruses, including, influenza A and B, adenovirus, respiratory syncytial virus (RSV) and recently corona viruses. *Staphylococcus aureus* is mostly found in cases of severe CAP where affected patients are mostly hospitalized in the ICU.

Manifestations of CAP are variable and can range from very mild symptoms to severe symptoms, comorbidity and eventually death. Patients present with CAP will usually exhibit aspecific symptoms of fever, chills, tachycardia, myalgia, arthralgia and even gastrointestinal manifestations like nausea and vomiting. Cough either productive or non-productive, chest pain due to pleuritic irritation, dyspnea, tachypnea and sometimes apnea are important presentations in pneumonia. Physical examination findings are found in table 1.

Type of physical examination	Findings
Inspection	Cyanosis, flushing, accessory muscle of respiration utilization
Palpation	Tactile fremitus
Percussion	Dull or flat percussion
Auscultation	Rales, rhonchi, bronchial breath sounds, pleural friction rub

Table 1: *Physical examination findings in pneumonia.*

One of the rare but serious complications of CAP is necrotizing pneumonia and pulmonary air leak syndrome. The latter to be discussed hereunder, while necrotizing pneumonia is defined as rapid progression of consolidation to develop pulmonary tissue necrosis and cavitation and eventually lead to pulmonary gangrene. Treatment is targeted based on the causative microorganisms and the possible complications or comorbidities and it's beyond the topic of this paper.

Overview of pulmonary air leak syndrome

Pulmonary air leak syndrome (ALS) is a known significant complication of pneumonia in ventilated and even non-ventilated patients. It occurred when air escapes from the alveoli to the extra-alveolar spaces such as the interpleural space, pulmonary interstitium, mediastinum or pericardium and being associated with respiratory compromise and signs of pulmonary distress or failure. It can complicate infection such as CAP and necrotizing pneumonia, empyema, chest trauma, pulmonary surgeries, ARDS and mechanical ventilation use. This is especially profound in patients with pre-existing lung disease, COPD, smokers, diabetics and chronic steroid users. In a published study, prevalence of ALS is calculated to be 40% of infants on Mechanical ventilation (MV).

The pathophysiology behind this phenomenon is the air trapping in the alveoli and the decreased pulmonary compliance of the diseased lung. Patients with ALS experience sudden deterioration of respiratory condition, the need for higher settings to reach target oxygen saturation and decreased air entry or even silent chest on examination, associated with hyper-resonance percussion confirmed by chest x-ray which can show pneumothorax, pneumo-mediastinum or even (pulmonary) interstitial emphysema.

As air escape from the alveoli and reside in the pleural cavity which supposed to have negative pressure, this implicit remarkable negative effect on the pleural dynamics and gas exchange. In addition, it creates noteworthy challenges in the ventilation of sick patients especially those with persistent air leak syndrome beyond 2 - 7 days despite the insertion of proper chest tube. The inspiratory volume increases and expiratory volume decreases leading to ventilation/perfusion mismatch (V/Q mismatch). Oxygenation is disturbed if the escaped air is not quickly removed from the extra-alveolar space and respiratory alkalosis arise due to the continuous removal of CO_2 via the chest tube, subsequently multi-organ involvement become an issue with high mortality rate, estimated in one study to reach 67%.

According to literature and guidelines, treatment of adult ALS emphasizes the requirement of early surgical intervention to facilitate quick recovery and rehabilitation and minimizes PALS duration. The guidelines in pediatric groups however are still controversial and not conclusive. Surgical interventions as well as chemical agents were also recommended in the past by the American and British thoracic societies, but these references don't include patients with severe presentations or ICU patients who should have minimal surgical interventions. Most recent publication in PALS in pediatric ICU patients targeted ventilation and oxygenation methodology.

Conventional ventilation is one of the early methods to be used, with synchronized intermittent mandatory ventilation (SIMV) being the most chosen mode with low set inspiratory time and high respiratory rate. Tidal volume should not exceed 6 - 8 ml/kg, allowing permissive hypoxemia and permissive hypercapnia with target PH 7.2, this is to avoid high peak pressure and thus limiting alveolar perforation and ALS. If conventional ventilation fails to improve PALS, trial of high frequency oscillatory ventilation (HFOV) is attempted.

HFOV aims to deliver very low TV (1 - 3 ml/kg) at extremely hyper-physiologic rates (220 - 900 breaths Per minute). This to minimize the risk of volutrauma and barotrauma and their consequent effect on ALS development or progression. Despite the proposed hypothesis, clinical trials and meta-analysis on neonates suffering ALS have not confirmed this theory, but it has shown non-significant increases of ALS in those ventilated with HFOV. On the other hand, one old randomized clinical trial in 1993, stated significant reduction of ALS in neonates treated with HFOV. In a case report published in 2014 by Soni Kd., *et al*. a couple of retrospective studies were used which reported positive effect of HFOV on the reduction and treatment of severe cases of ALS and it concluded that HFOV is a reasonable alternative treatment in patients with ALS caused by fistulas.

Overview of severe combined immuno-deficiency (SCID)

"Primary immunodeficiency" denotes diseases resulting from inherited defects of the immune system. Severe Combined immunodeficiency syndromes are a heterogeneous group of primary immunodeficiency disorders arising from a disturbance in the development and function of both T and B cells (cellular and humoral immunity) and may also involve natural killer (NK) cells, and that lead to early death from overwhelming infection, typically in the first year of life. Patient with SCID may present with recurrent, severe, or long-lasting infections, with suboptimal response to extended appropriate treatment, and might result in devastating complications and mortality early in the first year of life. Other clinical manifestations could be opportunistic infections with unusual pathogens, persistent oral thrush, chronic diarrhea and hyper-metabolism leading to failure to thrive (FTT).

Physical examination may reveal absent peripheral lymphoid tissues, while chest X-ray may show absent thymic shadowing. However, some rare cases of SCID may have well developed thymus and its absence doesn't exclude the diagnosis of SCID.

Laboratory studies that are suggestive of SCID including: peripheral lymphopenia, abnormal flow cytometry with low or absent naïve T-lymphocytes (CD3+), B-lymphocytes (CD19+) or Natural Killer (CD16,56). Other laboratory findings include hypogammaglobulinemia and lack of response of T-cells or B-cells to mitogens stimulation. Treatment of SCID patients is initiated from prenatal periods starting with maternal booster administration to facilitate antibodies transfer to the fetus. The affected patients should be kept in reverse-isolation and should avoid administration of live vaccines. If needed, only irradiated, WBC-depleted, and CMV-negative blood product should be used. Moreover, SCID patients should receive antibody replacement therapy in the form of IVIG, in addition to prophylaxis against pneumocystis jirovecii, RSV and herpes simplex virus. The definitive treatment of SCID patient is transplant of hematopoietic stem cells early in life prior to 3 months of age when the maternal immunoglobulins levels are still high and infections frequency is still low [1-6].

Case Presentation

This is a 7-month-old Saudi girl, a product of late preterm of 34 weeks, who presented to the emergency department with a chief complain of fever and URTI symptoms (runny nose and cough) for 2 days duration and difficulty breathing for a few hours prior to presentation.

The child was previously well tell 2 days prior to presentation when the mother noticed her to have a subjective fever responding to antipyretic, followed by a progressive productive cough throughout the day, associated with increased work of breathing, decreased activity, blocked nose, snoring at sleep, and interrupted feeding.

No history of apnea, choking or facial congestion, no cyanosis, no recurrent URTI, or chronic cough.

Few hours prior to the ED visit, the child has episodes of cough followed by noticeable unequal chest movements and difficulty of breathing.

Evaluation upon presentation to pediatric emergency, clinically was conscious, sick looking, and in respiratory distress in form of tachypnea, and on/off grunting.

Her initial vital signs Temp: 37.2C, HR: 150-170 bpm, BP: 92/65 mmHg, RR: 55-60 b/min, SpO_2 saturation: 91 - 92% at room air. Chest examination revealed unequal chest expansion by inspection, and there was significant decreased air entry on the right side by auscultation.

Her initial chest x-ray (Figure 1) showed: Radiolucency of peripheral space and loss of lung marking of the upper and middle zone of the right lung representing pneumothorax, for which a chest tube was inserted.

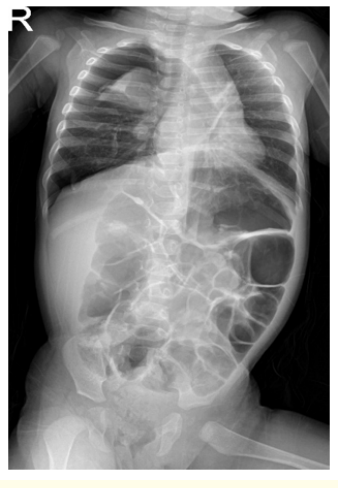

Figure 1

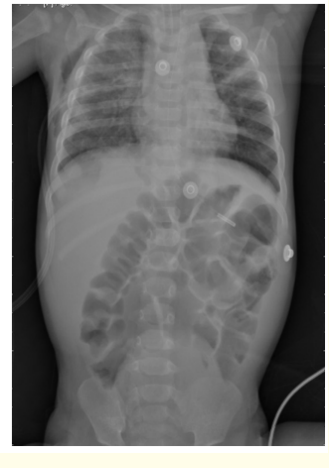

Figure 2: *Post-chest tube insertion.*

The patient was admitted to PICU with an impression of Acute hypoxic respiratory failure and right-sided spontaneous pneumothorax secondary to severe chest infection (community acquired Pneumonia), to rule out congenital pulmonary airway malformation CPAM, and congenital lobar emphysema.

On 1st day of admission, the patient was intubated due to worsening respiratory status requiring high settings of conventional MV to maintain her oxygenation, while her blood gas was maintained. The patient was sedated, hemodynamically supported by inotropes, and covered with broad-spectrum antibiotics.

By 3rd day, the patient became hypoxic requiring higher FiO_2, chest X-ray was showing worsening aeration with right upper zone consolidation vs collapse reflecting severe acute respiratory distress syndrome ARDS with oxygen saturation index, OSI 14, for which the patient was shifted to high frequency oscillatory ventilation, HFOV.

Medical management and the goal of therapy provided were to keep the patient deeply sedated and muscle relaxed. To optimize her hemodynamics and ventilate her with permissive hypoxia and permissive hypercarbia strategy, in addition to sepsis control and close organ function monitoring, then proceed with a diagnostic workup (chest CT).

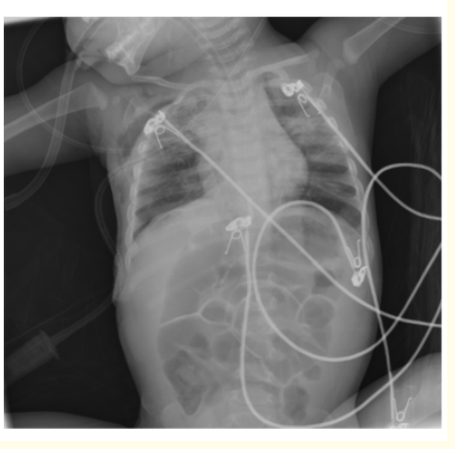

Figure 3: *Right upper lung consolidation, Severe ARDS OSI 14.*

After 3 days on HFOV, the patient shifted again to conventional mechanical ventilator MV as she was showing clinical and radiological improvement.

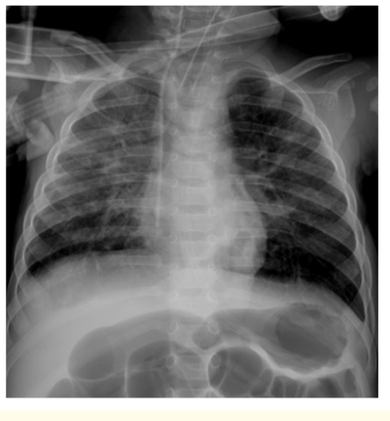

Figure 4: *Improving ARDS, mild OSI 7.*

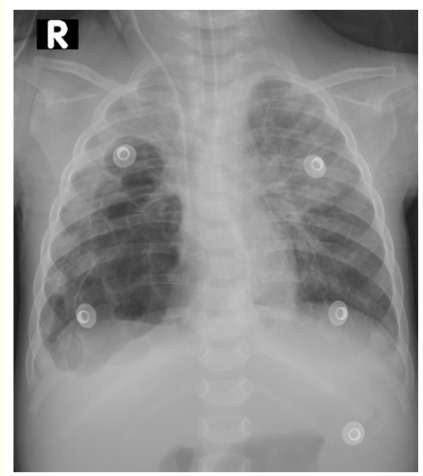

Figure 5: *Upon shifting the patient to conventional MV on the 5th day of admission.*

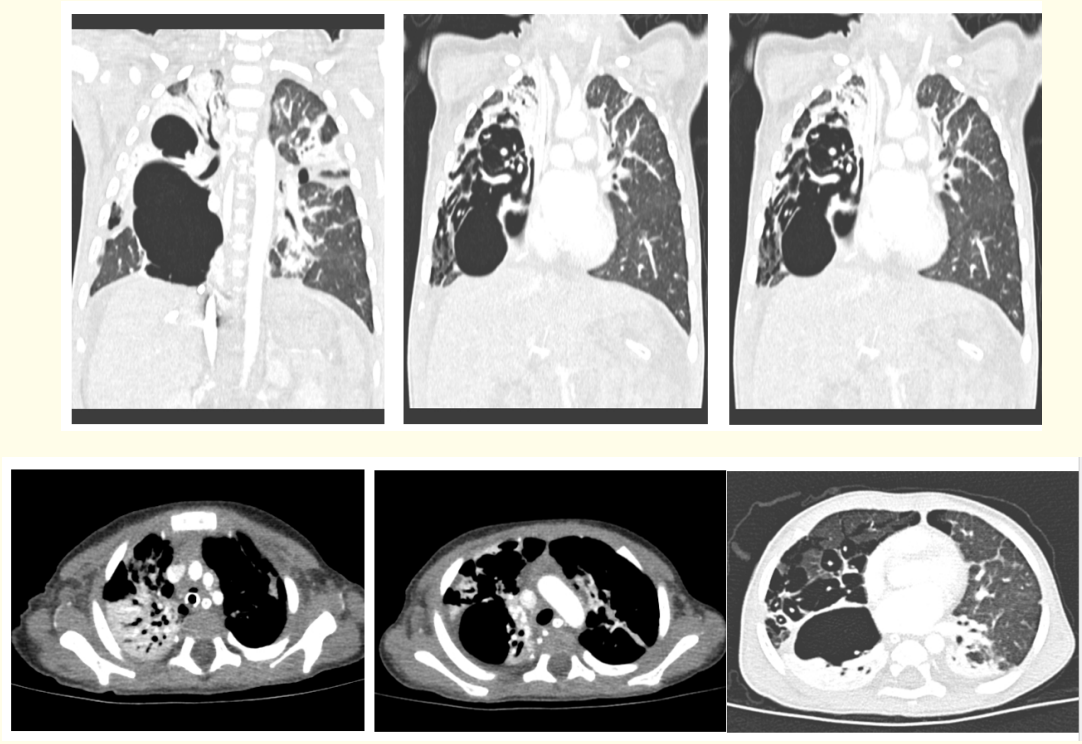

Figure 6: *Chest CT.*

Reported chest CT showed: Multiple cystic lesions with walled unlined cysts mainly in the right middle and lower lobes with the left lung showing multiple translucencies looking like air leak around lung fissure with an impression of severe necrotizing pneumonia with the possibility of underlying congenital cystic lung lesion vs CPAM type 4.

As the patient presented with severe necrotizing pneumonia, Flow cytometry was performed to her, and it showed marked depletion of T-cells (CD3) and NK-cells (CD16,56), preserved B-cells (CD19), and normal expression of TCR and HLA molecule; with impression severe combined immunodeficiency.

Lymphocyte subset		
CD3	610	)5900-1900 (
CD4	483	)4300-1400 (
CD19	701	)2600 -610 (
NK	14	)950-160 (
Immunoglobulin		
IGA	0.36	)2.46 - 0.39(
IGG	625	)1261 - 696 (
IGM	115	)153 - 54 (
IGE TOTAL	6.99	200=<IU

Figure

Further immunological and genetic workup was requested, and the patient started on IVIG replacement, Bactrim prophylaxis, and no live vaccines were to be given to her.

The patient was kept ventilated for a total of 18 days, weaned from inotropic support successfully, shifted to Non-invasive ventilation NIV of High Flow Nasal cannula HFNC, started on Orogastric tube OGT feeding, and completed the course of antibiotics.

The patient was transferred to a tertiary hospital for further management of her suspected primary immunodeficiency disorder and to consider stem cells transplant.

Discussion

SCID is a syndrome caused by mutations in any of several genes whose products are crucial for the development and function of both T and B cells and may also affect natural killer (NK) cells. It is usually clinically apparent in the first few months postnatally and can lead to severe comorbidity and mortality mostly prior to the first year of life if affected patient is left untreated. It's manifested by failure to thrive, long-lasting infections, infections that are therapy-resistant, recurrent infections; namely: pneumonia, bronchitis, otitis media, meningitis or deep-seated infection like sinusitis, cellulitis or sepsis.

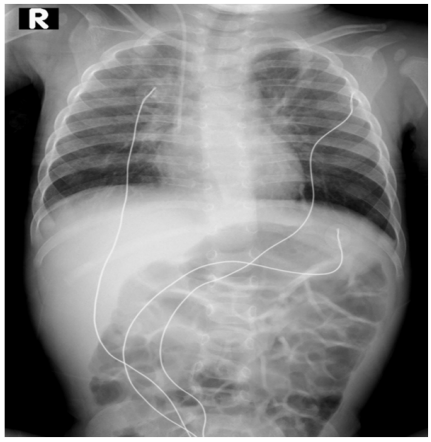

Figure 7: *Chest X-ray post extubation.*

Our reported 7 months old patient was healthy prior to this presentation, didn't suffer from recurrent or long-lasting infections and required no hospitalization other than the post-natal uncomplicated admission into the NICU due to respiratory distress syndrome which was related to her prematurity, requiring only two day of invasive mechanical ventilation. She didn't r experience any GI symptoms such as chronic diarrhea, and has received her vaccinations until the age of 4 months.

The only manifestations that could fit SCID presentation in our patient is the failure to thrive, probably she was experiencing minor infections that didn't require hospitalization, in which it was not counted by the family as significant during the history interview.

Her sickness developed two days preceding the hospitalization; with upper respiratory tract infection symptoms that rapidly progressed to acute hypoxic hypercapnic respiratory failure at the time of seeking medical advice. This was secondary to CAP, and was further complicated by pulmonary air leak syndrome in the form of spontaneous pneumothorax requiring chest tube insertion. Patient was then admitted to the PICU, sedated and ventilated requiring HFOV.

Due to the severe presentation, patient was covered with broad spectrum antibiotics, third generation cephalosporine (i.e. cefotaxime), anti-staph (vancomycin) and azithromycin as well as antiviral (oseltamivir).

Despite the good coverage, patient condition was not improving and she had developed new quick deterioration with severe Acute respiratory Distress Syndrome ARDS picture necessitating HFOV, the ventilation strategy was permissive hypoxemia and permissive hypercapnia as lung protective strategy. For this severe unusual course, and the requirement of HFOV, pulmonary structural congenital diseases were suspected as well as primary immune deficiency, in which patient was investigated for both.

Chest CT was ordered and showed multiple changes suggestive of complicated CAP and no evidence of congenital lung diseases. Flow cytometry was sent and confirmed the diagnosis of severe combined immunodeficiency SCID, a primary immune deficiency, hence the differential diagnosis of CPAM was deferred as the patient course can be explained by the primary immune deficiency.

As this child presentation seems to be late than expected and had good outcome at the end by surviving the severe form of chest infection and the air leak syndrome, this was considered to be unusual, and hence thought of reporting it.

Conclusion

- Severe combined immunodeficiency, SCID, is a rare primary immunodeficiency disorders PID caused by a severe deficiency in T-cell and B-cell function.
- SCID is generally considered to be the most serious type of PID and a true pediatric emergency.
- Community-acquired pneumonia can be a very early presentation PID.
- SCID should be considered in patients with community-acquired pneumonia CAP and severe necrotizing pneumonia.
- Prophylaxis for SCID includes IVIG, prophylactic antifungal, and no live vaccines.
- Definitive treatment is hematopoietic stem cells transplant.

Consent

Written consent was taken from the parents for approval of case reporting and publishing.

Acknowledgment

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Conflicts of Interest

None.

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None.

Contribution of Authors

Preparation of first draft: I.M, M.M, M.O.

Literature review: M.B, K.S, S.H, H.H.

Conceptualization: I.M, M.O, M.M, F.A.

Intellectual inputs for improvement of M.M, H.S, H.H, F.A.

Approval of final draft: I.M, M.M.

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