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Case Report

Spontaneous Pulmonary Air Leak Syndrome in Severely Malnourished Child

Iman A Al Mukhtar[1]*, Fatimah Al Zahir[2], Israa Al Aythan[3], Zainab Al Daif[3] and Rehab Al Mohsin[4]

[1]Pediatric Critical Care Consultant, Qatif Central Hospital, Qatif, Saudi Arabia

[2]Pediatric Senior Registrar, Qatif Central Hospital, Qatif, Saudi Arabia

[3]Pediatric Resident, Qatif Central Hospital, Qatif, Saudi Arabia

[4]Intern Pediatrician, Qatif Central Hospital, Qatif, Saudi Arabia

***Corresponding Author:** Iman A Al Mukhtar, Pediatric Critical Care Consultant, Pediatric Department in Qatif Central Hospital, First Health Cluster Eastern Province, Ministry of Health Saudi Arabia, Saudi Arabia.

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Abstract

We describe an 8-year old Saudi boy who is a known case of neurodegenerative disease, severe failure to thrive and severe malnutrition. He was hospitalized with hypovolemic shock due to severe dehydration. He was found to have spontaneous cervical subcutaneous emphysema on initial presentation progressed to pulmonary and mediastinal air leak syndrome, in addition to spontaneous pneumoperitoneum. To the best of our knowledge, this is the first reported case of pulmonary air leak syndrome associated with spontaneous pneumoperitoneum in a pediatric aged child, due to the effect of severe malnutrition.

Keywords: *Pulmonary Air Leak; Spontaneous Pneumoperitoneum; Severely Malnourished Child*

Introduction

Pulmonary air leak syndrome is a disorder characterized by air-leakage from the lung into extra-alveolar spaces where it is not normally present [1]. Pneumomediastinum is a condition where air leaks into the center of chest it usually does not have an impact on breathing [2].

This condition is very rare and usually occurs only in neonates. It affects 1 - 2% of normal neonates due to the physiological changes when infants start to breath [3]. The disorders resulting from this condition vary, depending upon the location of the air leakage. The most common conditions are pneumothorax, pneumomediastinum, pulmonary interstitial emphysema, and pneumopericardium. Other rarer forms are subcutaneous emphysema and pneumoperitoneum, which can occurs when extra-pulmonary air decompresses into the peritoneal cavity [4].

In literature reviews, the reported cases of spontaneous air leakages were cited in adult age group suffering from severe anorexia nervosa [5]. The pulmonary systems are adversely affected by spontaneous pneumothorax and the theory behind that is severe malnutrition.

In this case study, we are focusing on an 8-year-old boy, who is a known case of neurodegenerative disease with long lasting quadriplegia, chronic aspiration and severe malnutrition. Our main aim for reporting this case is to raise awareness of a very rare presentation of a disorder that is known to be in a certain age group, neonate, but can present in other age groups due to chronic disease.

Case Presentation

An 8-year-old Saudi boy, who is a known case of Neurodegenerative disease, severe failure to thrive, and severe malnutrition. He was hospitalized to intensive care unit due to hypovolemic shock as a result of severe dehydration. The Patient weighted only 8 kg, which is way below third centile of ideal weight for his age -using a local standard growth chart of Saudi Arabia. Clinically he had dry, loose and wrinkled skin, brittle hair and muscle wasting particularly in the buttocks and thighs area. Micronutrient deficiency is another interpreting factor in his diagnosis with malnutrition. His blood values were as following: vitamin D deficiency (19.5 ng/dl), low minerals (calcium, phosphorus, magnesium level of 1.9 mmol/L, 0.84 mmol/L, 0.6 mmol/L, respectively) as well as macronutrient deficiency as he had very low albumin of 11 G/L with total protein of 54.3 G/L.

Regarding patient's family household living it was complicated and it had a big impact in his malnutrition and general condition. Patient developed normally tell the age of 3 years old when he started to have his 1st symptoms of his neurodegenerative disorder, losing mile stone and regression of his motor and cognitive function were the first symptoms that the family noticed. After that, he became totally disabled, bed bound and dependent for care.

Family was seeking medical advice at the beginning of his deterioration, when they were told that there was no curative treatment other than supportive therapy. The family started to lose the hospital follow up and started to look for other alternative medicine that did not benefit him in any way. Family has been advised multiple times by physician for nasogastric tube feeding and gastrostomy tube feeding and they refused. They kept trying with oral intake despite the fact that it wasn't adequate and patient was not gaining weight. Mother was the primary care giver; she had two other kids below the age of five years. She is a student in college, with a part time job trying to provide for her family. The father was working as well with poor socioeconomic status. The grandmother was helping in the care of this disabled child when his mom was at work or at college. The mother is kind, and cares about her child but yet was not convinced about physician's opinions of inserting feeding and there is no signs of child abuse or neglect, rather a low level of education about how to take care of such child and the importance of medical follow up.

Upon admission, it was discovered that he had to have spontaneous cervical subcutaneous emphysema on physical examination and on his initial Chest X-ray without any respiratory symptoms (Figure 1).

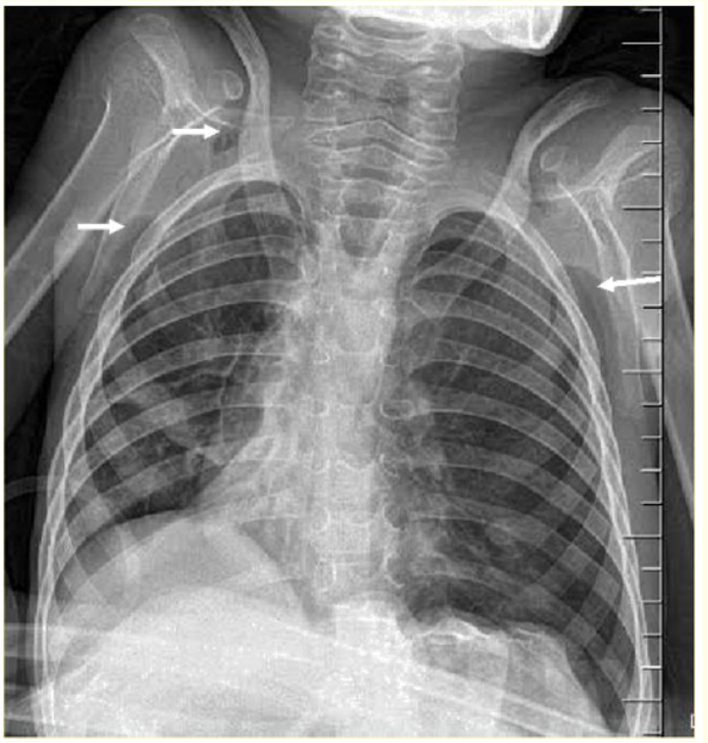

Figure 1: *Chest X-ray showing multiple subcutaneous emphysema.*

The Patient was treated for his hypervolemia by giving him fluids and electrolytes replacement. Initially, he did not have any respiratory symptoms and he did not require any respiratory support. However, a few days after admission, patient's temperature started to spike fever and he was showing signs of septicemia. He was started on IV antibiotics, soon after.

Subsequently, the patient showed signs of pulmonary air leak syndrome and spontaneous pneumothorax while he was in room air (Figure 2).

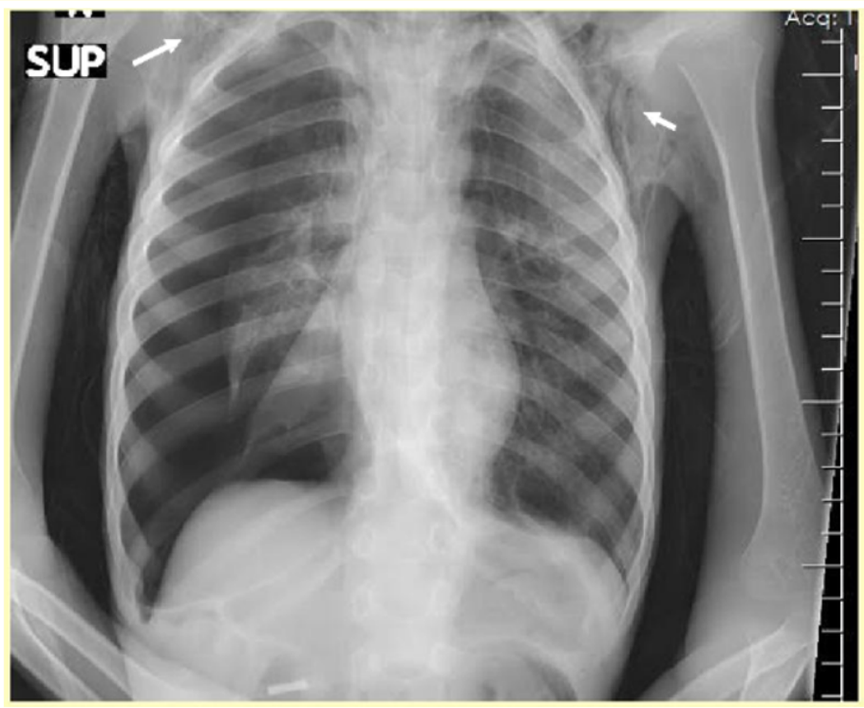

Figure 2: *Pulmonary air leak syndrome and Rt. Side spontaneous pneumothorax.*

His septicemia progressed to catecholamine refractory septic shock in which he required invasive ventilation for hemodynamic instability with gentle mode of ventilation.

Subsequently, his pulmonary air leakage was progressing to bilateral pneumothorax, in which bilateral chest tubes were inserted as the patient was on impending tension pneumothorax situation with increased respiratory effort and oxygen requirement. Chest x-ray (Figure 3A and 3B).

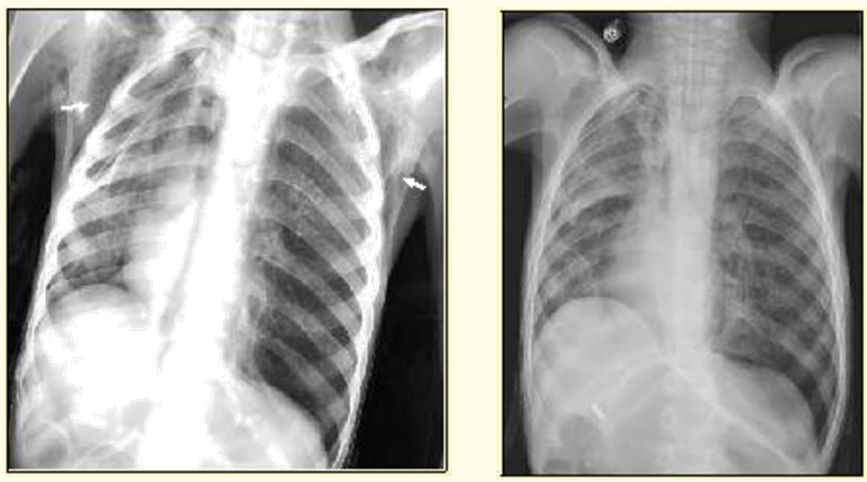

Figure 3A and 3B: *Showing bilateral pneumothorax resolving after 2 chest tubes insertion.*

Furthermore, the patient developed spontaneous pneumoperitoneum evident in examination and abdominal x-ray (Figure 4). CT with contrast rolled out bowel perforation. Spontaneous resolution of that pneumoperitoneum occurred after few days with no active intervention other than conservative support.

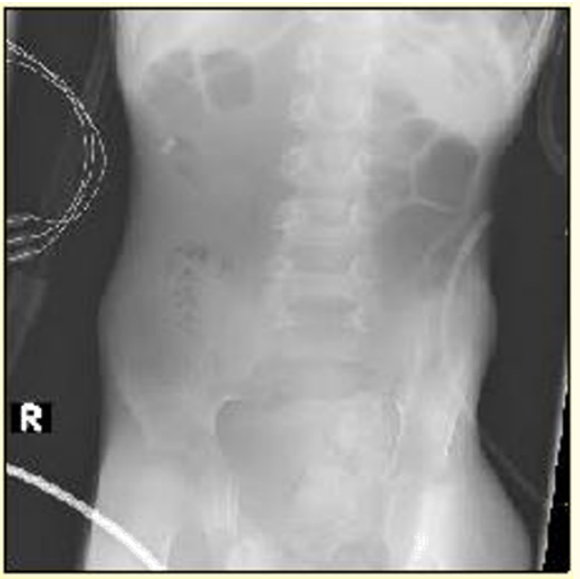

Figure 4: *Pneumoperitoneum.*

Main ventilation strategy was gentle mode of ventilation with low pressure to avoid further injuries and barotraumas with a goal of optimization for the patient's condition, well planned weaning, and relatively fast tract roadmap extubation to remove the effect of positive pressure ventilation, and to prevent further air leakage. That goal was nearly reached with minimal pressure support of spontaneous mode of ventilation PS 8/5. The patient was clinically tolerating well and maintains gas exchange and complete resolution of pneumothorax even after clamping the chest tubes (Figure 5).

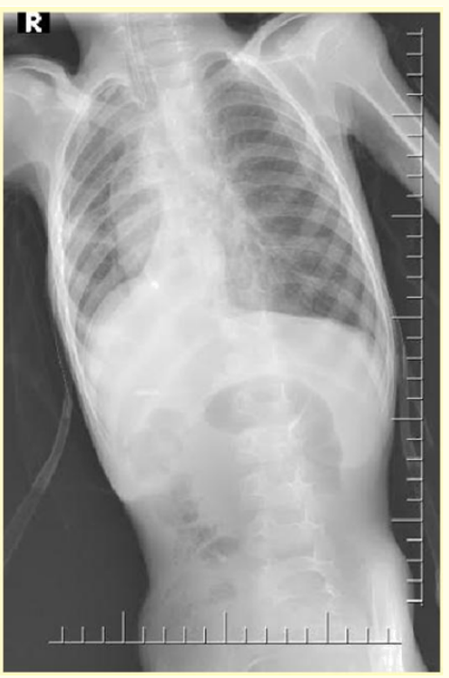

Figure 5: *Complete resolution of pneumothorax.*

The extubation was challenging. The patient's condition was worsening from a different aspect as he developed new septicemia, in which he had multi-organ failure and critical condition and he was not candidate of extubation.

Discussion

Spontaneous pulmonary air leak syndrome is well known condition in neonates with fragile immature lungs. This syndrome can occur early in adult where there are reported cases about patient with anorexia nervosa who developed it. In pediatric age groups spontaneous pulmonary air leak syndrome usually develop secondary to interstitial lung disease, ARDs or immunocompromised patients i.e. post stem cell transplants.

There was no reported cases that associate severe malnutrition and spontaneous air leak, which is the case in our patient. Severe malnutrition has been closely linked to reduced lung surfactant production and impaired extracellular matrix, patchy atelectasis, interstitial edema, and inflammation with increased amount of polymorphonuclear cells. Under-nutrition diminished diaphragmatic muscle mass and respiratory muscle strength that led to reduced muscular output, prolonged the decay of inspiratory muscle pressure during expiration, and increased chest wall resistive and elastic mechanical components. Nutritionally deprived patients had significantly less lipid and protein as well.

Histopathological findings of emphysematous - parenchyma (increased size of air spaces distal to the terminal bronchiole with thinning and partial destruction of septal wall) with decreased elasticity and increase susceptibility of tissue injury and poor lung compliance resulting on fragile alveoli and air can easily leaked to the pleura resulting in pulmonary air -leak.

Conclusion

In this case report we are linking the effect of severe malnutrition with spontaneous pulmonary air-leak and trying to spotlight the association and anticipation of air leak syndrome in any patient with severe malnutrition and to look for it before respiratory symptoms appears. Management of air leak is basically supportive with chest tubes in addition good gentle modality of ventilation, early detection plays major role in management and prognosis.

Consent

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

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Conflicts of Interest

None.

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Contribution of Authors

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

Literature review: I.M, Z.D, R.M.

Conceptualization: I.M, F.Z, Z.D.

Intellectual inputs for improvement of manuscript: F.Z, Z.D, I.A.

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

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