International Journal of Advances in Medicine
Das A et al. Int J Adv Med. 2024 Nov;11(6):621-623
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: https://dx.doi.org/10.18203/2349-3933.ijam20243070

Case Report

Negative pressure pulmonary edema and angioedema: a fatal correlation

Ananya Das[1], Sahil Kumar[1], Jaideep Pilania[1], Jithesh G.[2]*, Mukesh Bairwa[1]

[1]Department of General Medicine, All India Institute of Medical Sciences, Rishikesh, Uttarakhand, India
[2]Department of Critical Care Medicine, Narayana Health Bengaluru, Karnataka, India

Received: 24 September 2024
Accepted: 15 October 2024

***Correspondence:**
Dr. Jithesh G.,
E-mail: jitheshgkoppam@gmail.com

ABSTRACT

Negative pressure pulmonary edema (NPPE), a non-cardiogenic pulmonary edema resulting from acute or chronic upper airway obstruction, remains a significant challenge in critical care settings due to the morbidity and mortality associated with undiagnosed cases. The most accepted pathophysiology involves the high inspiratory pressure required to counteract the upper airway obstruction, which leads to progressive negative intrapleural pressure. This results in increased pulmonary microvascular pressure, causing alveolar flooding. Among reported cases, laryngospasm in the postoperative period is the most common cause of NPPE. Management involves securing the airway and providing positive pressure ventilation. However, prevention and treatment remain subjects of further research.

Keywords: SLE, Angioedema, Pulmonary edema, Post-obstructive

INTRODUCTION

Negative pressure pulmonary edema (NPPE), sometimes referred to as post-obstruction pulmonary edema, is a form of non-cardiogenic pulmonary edema that results from the generation of high negative intrathoracic pressure needed to overcome upper airway obstruction. The incidence of NPPE in acute upper airway obstruction has been estimated to be around 12%, with laryngospasm during intubation or in the postoperative period after anesthesia being the most common cause.[1,2] However, the data seems underreported due to a lack of proper understanding of the accurate pathogenesis of this clinical entity. The presentation of NPPE can be both immediate and delayed, with no specific indicators of decompensation or prognosis. Therefore, prompt clinical suspicion and subsequent treatment are instrumental in preventing the otherwise inevitable morbidity and mortality associated with this phenomenon. Here, we present a case of a young woman in her twenties who presented with acute upper airway obstruction secondary to angioedema and, unfortunately, succumbed to her illness because of NPPE despite our best possible efforts.

CASE REPORT

A 23-year-old married female from North India, recently diagnosed with systemic lupus erythematosus (SLE) without major organ involvement and not on any immunosuppressive therapy, presented to the emergency department with acute, painless facial swelling, including swelling of the lips and tongue and gradually progressive hoarseness of voice. Urgent medical attention was sought. An urgent ENT consultation was obtained, and a laryngoscopy revealed an edematous epiglottis. The patient's attendants were informed about the potential need for emergency tracheostomy in case of further worsening or onset of stridor. The patient was then transferred to the intensive care unit and started on steroids, antihistamines, and adrenaline infusion for persistent angioedema. Her baseline investigations are shown in the table below (Table 1).

Table 1: Baseline parameters of the patient.

Variables with normal values and units	Patient's values
Hemoglobin (g/dl): (Adult men: 14-18 g/dl, adult women: 12-16 g/dl)	7.8
Total leukocyte count: (5000-10,000/mm^3)	4950
Platelets: (150,000-450,000/mm^3)	35000
Total Bilirubin (mg/dl): (0.3-1.0 mg/dl)	0.3
Direct Bilirubin (mg/dl): (0.1-0.3 mg/dl)	0.2
SGOT (U/l): (5-40 U/l)	169
SGPT (U/l): (7-56)	75
Blood urea (mg/dl): (7-20 mg/dl)	31
Serum creatinine (mg/dl): (0.6-1.2 mg/dl)	0.60
Sodium (mmol/l): (135-145 mmol/l)	134
Potassium (mmol/l): (3.6-5.5 mmol/l)	4.5
Antinuclear antibody (ANA): (<1:40)	2+(1:160)
Anti-double-stranded DNA: (<4IU/ml)	900.4

Legend: Hb: haemoglobin; TLC: total leukocyte count; SGOT: serum glutamic oxaloacetic transaminase (AST: aspartate aminotransferase); SGPT: serum glutamic pyruvic transaminase (ALT: alanine aminotransferase); Na: sodium; K: potassium; ANA: antinuclear antibody; Anti-dsDNA: anti-double stranded DNA antibody; DCT: direct coombs test; ICT: indirect coombs test; ESR: erythrocyte sedimentation rate; CRP: C-reactive protein; g/dl: grams per decilitre; mg/dl: milligrams per decilitre; U/L: units per litre; mmol/l: millimoles per litre; mm/hr: millimetres per hour; mg/l: milligrams per litre; IU/ml: international units per millilitre.

On presentation to the ICU, the patient was tachypneic, necessitating positive pressure support via non-invasive ventilation. She was maintaining oxygen saturation and stable vital signs when she suddenly developed a massive amount of pink, frothy sputum, followed by a sudden drop in oxygen saturation. The patient's vitals became unstable, and endotracheal intubation with a 6.5 cm ET tube was attempted but unsuccessful due to the edematous airway. A laryngeal mask airway was then used to secure her airway, stabilizing her vitals temporarily. An urgent bedside X-ray chest was done (Figure 1). A bedside point-of-care lung ultrasound was done, suggesting diffuse B lines in bilateral lung fields without shredding. The ENT department was promptly informed, and an emergency tracheostomy was performed to further secure her airway. Despite these efforts, the patient again developed massive pulmonary edema, leading to sudden cardiac arrest. High-quality cardiopulmonary resuscitation was initiated, but unfortunately, the patient could not be revived and succumbed to her illness the same day.

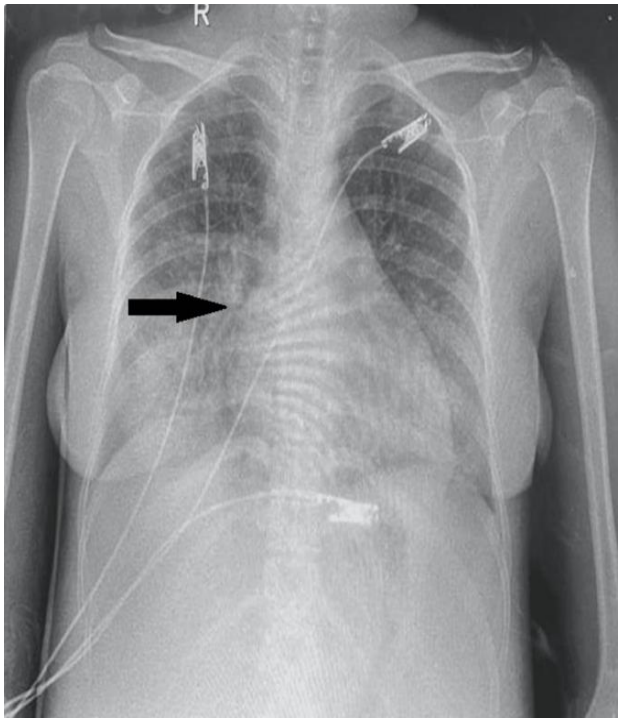

Figure 1: X-ray chest of the patient on presentation.
Legend: X-ray chest on presentation. Arrow (A) showing hilar prominence with cephalization suggestive of pulmonary edema.

DISCUSSION

NPPE is a common yet relatively underreported cause of acute respiratory failure resulting from intense inspiratory effort against an obstructed airway, which may be due to upper airway infection, tumor, or laryngospasm.[3] Moore initially reported NPPE in 1927 in spontaneously breathing dogs exposed to a resistive load.[4] The relationship between pulmonary edema and upper airway obstruction was later described by Capitanio et al who documented the phenomenon in two children with epiglottitis and croup.[5] NPPE has been categorized into type 1 and type 2. Type 1 NPPE is an acute condition that occurs after sudden airway obstruction, while type 2 NPPE typically follows the relief of chronic airway obstruction, often seen in children.[6] The incidence of NPPE varies widely, from as low as 0.1% to as high as 12%, suggesting that many cases go unreported.[7-10] The pathophysiology of NPPE involves highly negative intrathoracic pressure, leading to a dramatic increase in systemic venous return to the heart and a simultaneous drop in cardiac output. This disrupts alveolar cell junctions and increases pulmonary capillary pressure while lowering intra-alveolar pressure, causing fluid to rapidly move into the interstitial and alveolar spaces. When the fluid exceeds a critical level, massive alveolar flooding and pulmonary edema occur. In type 2 NPPE, the development of auto-PEEP (positive end-expiratory pressure) and an increase in end-expiratory lung volume due to chronic obstruction can suddenly return to baseline upon relief of the obstruction, creating negative intrapulmonary pressure and resulting in fluid transudation into the interstitium and alveoli.[11] Diagnosing

NPPE requires strong clinical suspicion and a thorough understanding of its pathophysiology. Clinical presentation includes sudden hypoxia, tachypnea, tachycardia, agitation, and massive pink frothy sputum.[12] The diagnosis is primarily clinical and supported by chest radiographs and computed tomography. There are no proven interventions to prevent NPPE. However, prompt diagnosis and timely intervention can prevent morbidity and mortality. It is crucial to monitor patients postoperatively for signs of laryngospasm. Treatment typically involves maintaining a patent airway, oxygen supplementation with positive pressure ventilation, and, in some cases, diuretics, although their efficacy is yet to be proven. In our case, the condition was identified promptly and managed timely, but unfortunately, the patient could not be saved despite the best efforts. Nonetheless, data indicates that NPPE usually resolves rapidly within 12-48 hours when recognized and treated early.

CONCLUSION

NPPE remains an underreported yet significant cause of acute respiratory failure, often developing rapidly after airway obstruction. Despite timely recognition and intervention, this case highlights the high morbidity and mortality associated with NPPE, underscoring the need for continued research into its prevention and improved management strategies in critical care settings.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Lemyze M, Mallat J. Understanding negative pressure pulmonary edema. Intensive Care Med. 2014;40(8):1140-3.
2. Louis PJ, Fernandes R. Negative pressure pulmonary edema. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2002;93(1):4-6.
3. Bhattacharya M, Kallet RH, Ware LB, Matthay MA. Negative-pressure pulmonary edema. Chest. 2016;150(4):927-33.
4. Rupress.org. Available at: https://rupress.org/jem/article-abstract/45/6/1065/31677/THE-RESPONSE-TO-RESPIRATORY-RESISTANCE-A?redirectedFrom=fulltext. Accessed on 20 September 2024.
5. Capitanio MA, Kirkpatrick JA. Obstructions of the upper airway in children as reflected on the chest radiograph. Radiology. 1973;107(1):159-61.
6. Dicpinigaitis PV, Mehta DC. Postobstructive pulmonary edema induced by endotracheal tube occlusion. Intensive Care Med. 1995;21(12):1048-50.
7. Tami TA, Chu F, Wildes TO, Kaplan M. Pulmonary edema and acute upper airway obstruction. Laryngoscope. 1986;96(5):506-9.
8. Park H, Nam S, Jang YJ, Ku S, Choi S-S. Negative pressure pulmonary edema in a patient undergoing open rhinoplasty: A case report. Medicine (Baltimore). 2021;100(1):e24240.
9. Tebay A, Bouti K, Tebay N. Œdème pulmonaire à pression négative après une cholécystectomie-à propos d'un cas. Rev Pneumol Clin. 2017;73(5):267-71.
10. Xiong J, Sun Y. Negative pressure pulmonary edema: a case report. BMC Anesthesiol. 2019;19(1):10.
11. Zumsteg TA, Havill AM, Gee MH. Relationships among lung extravascular fluid compartments with alveolar flooding. J Appl Physiol. 1982;53(1):267-71.
12. Budhathoki A, Wu Y. Negative pressure pulmonary edema: A case report. JNMA J Nepal Med Assoc. 2020;58(227):491-3.

Cite this article as: Das A, Kumar S, Pilania J, Jithesh G, Bairwa M. Negative pressure pulmonary edema and angioedema: a fatal correlation. Int J Adv Med 2024;11:621-3.