

CASE REPORT

Hamman's Syndrome Accompanied by Diabetic Ketoacidosis; a Case Report

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Abstract: Hamman's syndrome is an uncommon clinical entity characterized by an idiopathic spontaneous pneumomediastinum as a result of a sudden increase in intra-alveolar pressure. It can be triggered by repeated vomiting or Kussmaul breathing associated with diabetic ketoacidosis (DKA). Careful attention to this particular condition is needed to avoid under-diagnosis and to provide optimal management. Herein, we report a case of an 18-year-old man complaining of chest discomfort and progressive weight loss, ultimately diagnosed with Hamman's syndrome secondary to DKA. The patient's symptoms disappeared after intravenous fluid and insulin administration, while his pneumomediastinum resolved following conservative treatment. Our report highlights the importance of recognition of the links between pneumomediastinum as a cause of chest pain in patients with DKA.

Keywords: Hamman's syndrome; Diabetic ketoacidosis; Mediastinal Emphysema

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1. Introduction

Hamman's syndrome, a spontaneous pneumomediastinum and pneumopericardium, accompanied by diabetic ketoacidosis (DKA), is a rare clinical entity (1). Although the pathogenesis of Hamman's syndrome caused by DKA is multifactorial, intra-alveolar pressure can be increased by Kussmaul breathing or repeated vomiting, which may lead to alveolar rupture (2,3). The vulnerability of alveolar walls due to malnutrition may influence the development of pneumomediastinum (3). Lack of knowledge about Hamman's syndrome secondary to DKA may lead to underdiagnose or unnecessary workups and surgical treatment (3,4). Herein, we report an 18-year-old man who was diagnosed with Hamman's syndrome accompanied by DKA. We share our experience and a literature review on etiology and pathogenesis to increase familiarity with this syndrome among physicians. Physicians should be aware of the possibility that pneumomediastinum

can be induced by DKA associated with type I diabetes mellitus.

2. Case presentation

An 18-year-old man, a college student with no medical history, came to the emergency department with two days of chest discomfort and history of marked body weight loss of 10 kg in two months. He did not take sugar-sweetened soft drinks daily. The patient reported polyuria and general fatigue for one week prior to admission.

His height and weight were 165.0 cm and 63.9 kg, respectively. The patients' vital signs were as follows: blood pressure 153/104 mmHg, heart rate 121 beats/minute, and respiratory rate of 26 breaths/minute. He had no cold sweating, no pallor, and no cyanosis. On physical examination, his chest was clear on auscultation with normal heart sound, and without chest wall tenderness on his abdomen. Subcutaneous emphysema was not observed. He was severely dehydrated with poor skin turgor and marked delay in capillary refill >2s. Significant laboratory results included a blood pH of 7.055, a blood glucose level of 901 mg/dL, white blood cells (WBC) 20,320 cells/ μ L, hemoglobin 16.2 g/dL, hemoglobin A1c 18%, serum potassium level of 6.1 mmol/L, blood urea nitrogen 20.6 mg/dL, and serum creatinine 0.93 mg/dL. Anti-

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insulin, anti-islet antigen-2, and anti- glutamic acid decarboxylase antibody tests were negative. Ketone bodies in his serum and urine were strongly positive. The other laboratory test results were unremarkable.

Chest X-ray revealed pneumomediastinum and supraclavicular subcutaneous emphysema without pneumothorax. A chest computed tomography (CT) revealed a few air bubbles in pneumomediastinum without pneumothorax or esophageal rupture (Figure 1A). No abnormal findings were noted in the trachea, bronchi, or lung parenchyma.

The patient was diagnosed with DKA associated with type I diabetes mellitus and pneumomediastinum. It was not known that our patient was diabetic until DKA and pneumomediastinum were found.

The patient was admitted to the intensive care unit for glycemic control, and hydration with normal saline and an intravenous regular insulin infusion were initiated. Intravenous insulin was transitioned to a subcutaneous insulin regimen within 24 hours of initiation due to improvement of glycemic control and the recalcitrant acidosis. We arranged a consultation with a diabetologist to review and control the disease.

The pneumomediastinum was treated conservatively. His chest discomfort disappeared and chest X-ray showed no signs of worsening amount of gas in the neck and mediastinum on day 2 of hospitalization.

Following resolution of DKA, CT scan showed that the pneumomediastinum had disappeared on day 12 of hospitalization (Figure 1B). He was discharged from the hospital on day 13 with an appointment for diabetic follow-up, including a chest radiograph.

Since his discharge from hospital, he has not experienced a recurrence of pneumomediastinum during the one-month follow-up, having fully recovered with adequate education regarding the management of insulin at home.

3. Discussion

The average age of Hamman's syndrome patients with DKA is 20 years old. The average time period since diagnosis of diabetes mellitus is 7.2 years with average blood glucose levels of 638 mg/dl on presentation (2).

Pneumomediastinum is rare and happens when air leaks into the connective tissue and finally enters the mediastinal space into the chest cavity from the bowel, airways, or lungs. It can result from physical trauma or other events including mechanical ventilation, esophageal perforation, hyperemesis, dental extraction, and obstetric delivery. These conditions sometimes become very severe and fatal. Although rare, pneumomediastinum accompanied by DKA is a benign, self-limiting condition (2).

In general, patients with pneumomediastinum usually

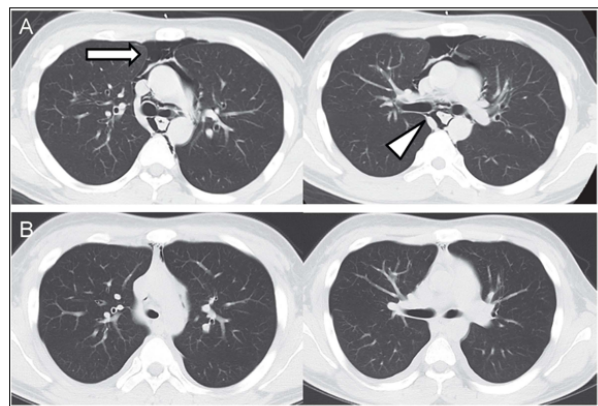


Figure 1: Chest computed tomography (CT) scan on admission demonstrated pneumopericardium (white arrow) and pneumomediastinum (arrowhead) (A). CT showed that pneumomediastinum disappeared on day 12 of hospitalization (B).

manifest extreme pain in the central chest. Other symptoms include voice distortion, labored breathing, and subcutaneous emphysema, specifically affecting the chest, neck, and face. However, most of the clinical signs of pneumomediastinum complicated with DKA are likely to have subsided by the time metabolic control has been achieved (2). Since the symptoms of pneumomediastinum caused by esophageal rupture are similar to those of DKA with pneumomediastinum, the exclusion of esophageal rupture, which has a high mortality rate, is the primary task in the diagnosis of DKA complicated by pneumomediastinum (5). Of note, perforation of the esophagus due to increased intraluminal pressures following violent vomiting is sometimes seen in DKA (1,6).

The precise pathophysiology of pneumomediastinum associated with DKA remains to be explained. Mainly, Kussmaul breathing, the compensatory respiratory mechanism in metabolic acidosis, raises alveolar pressure, predisposing the alveoli to rupture. DKA is often linked to severe vomiting caused by acidosis, which also increases intrathoracic pressures. Gastroparesis accompanied by diabetic mellitus may exacerbate vomiting. Our patient presented tachypnea, which may have been caused by increased expiratory efforts related to acidosis and ketotic hyperventilation. However, our patient did not complain of vomiting. Hu et al. recently reported that fibrotic changes in the lungs of individuals with poorly-controlled diabetes predispose the alveoli to rupture at lower intrathoracic pressures (7). In addition, diabetic malnutrition may influence lung mechanics by increasing surface forces and decreasing tissue elasticity (8). A previous experimental study indicated that starvation resulted in decreased surface density of lamellar bodies and mitochondria in a rat model (8). Although speculative, all

these phenomena might be involved in the development of Hamman's syndrome.

Of interest, there is a slight predominance of male patients with Hamman's syndrome (71%) due to men's greater average muscle mass and their ability to create intrathoracic pressures higher than those of women (2).

Pneumomediastinum can be diagnosed based on chest radiography. Chest CT is required to see mediastinitis or esophageal rupture.

Conservative management is required with follow-up. Drainage of subcutaneous air or skin incisions may be needed in symptomatic patients with respiratory distress. Gastroesophageal endoscopy is only indicated when other pathologies are being considered (6). In general, Hamman's syndrome is a benign entity with good prognosis.

This case had several important points worth discussing. We reached an early diagnosis using non-invasive and inexpensive examinations by avoiding gastroesophageal endoscopy and/or surgical interventions. The clinical symptoms improved spontaneously after the intensive intravenous insulin therapies without invasive management. However, this disorder can have significant mortality if misdiagnosed or mistreated, which is almost 100% without insulin therapy (9). Our report highlights the importance of recognition and management of pneumomediastinum induced by DKA associated with type I diabetes mellitus.

4. Conclusion

Emergency physicians should be aware of the possibility that pneumomediastinum can be induced by DKA associated with type I diabetes mellitus.

5. Declarations

5.1. Acknowledgments

None.

5.2. Authors' contributions

All authors met the criteria for authorship based on the International Committee of Medical Journal Editors' recommendations.

5.3. Consent for publication

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images. A copy of the consent form is available for review by the Editor-in-Chief of this journal.

5.4. Funding and supports

There is no funding to declare.

5.5. Competing interests

The authors declare that they have no competing interests.

5.6. Data availability

The data used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

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