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A MALE PATIENT WITH SYNCOPE, ANAPHYLAXIS, AND ST-ELEVATION: HEPATIC AND CARDIAC ECHINOCOCCOSIS PRESENTING WITH KOUNIS SYNDROME

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Abstract—Background: Approximately 1% of emergency department (ED) visits are due to anaphylaxis. Symptoms can include skin rash, facial and laryngeal edema, dyspnea, vomiting, hypotension, and shock. A transient loss of consciousness can also be a manifestation of anaphylaxis. A variety of electrocardiographic changes due to anaphylaxis have been described for Kounis syndrome, also known as allergic angina. **Case Report:** Here we describe the case of a male patient presenting at an ED with syncope, anaphylactic shock, and ST-segment elevation on electrocardiogram (ECG). The diagnostic workup led to the diagnosis of ruptured hepatic echinococcal cyst complicated by anaphylactic shock and syncope. ECG alterations were a manifestation of anaphylaxis, as defined by the type I Kounis syndrome. **Why Should an Emergency Physician Be Aware of This?:** Kounis syndrome represents an underestimated disease. Its prompt diagnosis in an ED has important clinical and therapeutic implications, such as modifications in the anaphylaxis treatment protocol, that is, adrenaline should be avoided because it could worsen vasospasm and myocardial ischemia. © 2016 Elsevier Inc. All rights reserved.

Keywords—anaphylactic shock; non-ischemic ST-elevation; Kounis syndrome; syncope

INTRODUCTION

Approximately 1% of emergency department (ED) visits are due to anaphylaxis (1). Symptoms of anaphylaxis can include skin rash, facial and laryngeal edema, dyspnea, vomiting, hypotension, shock, and loss of consciousness (2). Anaphylaxis can also produce a variety of electrocardiographic changes (3).

Here we describe the case of a patient presenting with syncope, anaphylactic shock, and associated ST-elevation at electrocardiogram (ECG).

CASE REPORT

Our medical emergency team was called to evaluate a 23-year-old man experiencing loss of consciousness. When the emergency team arrived, the patient was lying down on the floor of a stable; he was responsive to verbal stimuli and tachypneic (25 breaths/min) with wheezing. Blood pressure was 80/40 mm Hg and pulse rate was 125 beats/min. ECG monitoring showed sinus tachycardia with ST-segment abnormalities. The patient was

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confused and agitated and reported abdominal pain. A diffuse skin rash with facial edema was present.

With concern for anaphylaxis, the physician administered chlorphenamine 10 mg, hydrocortisone 500 mg, and 1500 mL crystalloid solution. The patient was transferred with an ambulance to the ED with a diagnosis of anaphylaxis with syncope.

Medical history was notable for no major medical conditions and no known allergies. The patient was not currently on any medications. His social history revealed that he worked as a shepherd and had a 75 pack-year history of smoking.

In the ED, the patient's vitals were blood pressure 110/60 mm Hg, pulse rate 95 beats/min, oxygen saturation 100% (inhaled fraction: 0.5), and temperature 37.1°C. The patient appeared alert and with facial edema. Regular S1 and S2 with systolic murmur on Erb's point were audible at cardiac examination. Lung examination revealed bilateral wheezes. The abdomen was soft with hyper-represented bowel sounds. Palpation of the right hypocondrium elicited pain with tenderness. There was no splenomegaly.

A 12-lead ECG performed at ED showed sinus tachycardia, ST-segment elevation in V2-V3 leads, and T-wave inversion in V2-V4 leads (Figure 1). Troponin-I was elevated (0.109 ng/mL, normal value < 0.014 ng/mL). Based on the ECG findings, troponin elevation, and clinical improvement after the initial treatment, no adrenaline was administered. An echocardiogram showed an anechoic mass with hyperechoic capsule (51 × 43 mm). This was located in the medial apical interventricular septum (see Figure 2 for images from computed tomogra-

phy [CT] and magnetic resonance imaging [MRI]). The mass distended the septum and occupied the apical portion of left ventricle. Left and right ventricular function was normal. No significant valvular insufficiencies were found, thus the murmur heard at physical examination was an ejection murmur. From the sub-xiphoid window, a voluminous cystic lesion (diameter 15 cm) was noted in the right hepatic lobe. A thorax-abdomen CT scan confirmed the presence of both myocardial (4.5 cm diameter) and hepatic (15.5 cm) cystic masses, the latter showing discontinuity of the wall (Figures 2–4).

The patient was admitted with a diagnosis of anaphylactic shock secondary to ruptured hepatic echinococcal cyst.

CT scan of coronary arteries was performed to rule out coronary artery disease or compression *ab extrinseco* and showed normal coronary arteries and no signs of compression. Cardiac MRI confirmed the presence of a 5 × 4 cm cystic lesion localized in the interventricular septum producing a complete systolic obliteration of the medio-apical left ventricle (Figures 2 and 4). No pathologic enhancement of the myocardial wall was found.

ECG recordings during the hospitalization showed normalization of ST-segment 24 h and T-wave inversion in V2-V6 (Figure 5). T-troponin levels normalized after 72 h.

Diagnosis of echinococcosis was confirmed by serologic tests (enzyme-linked immunosorbent assay). Albendazole 400 mg twice a day was prescribed, together with chlorphenamine and bisoprolol.

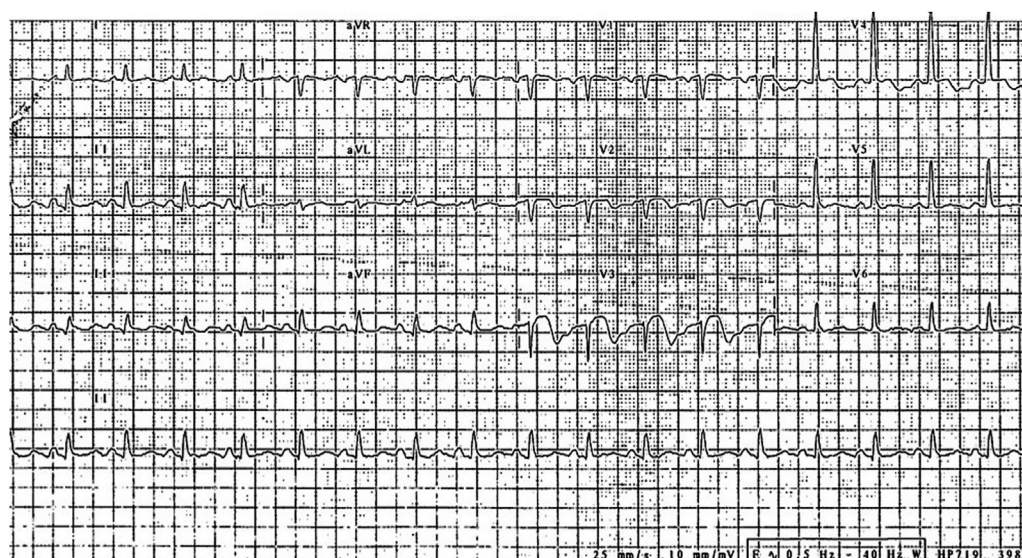


Figure 1. 12-Lead electrocardiography indicated sinus tachycardia, ST-segment elevation in V2-V3, and T-wave inversion in V2-V4.

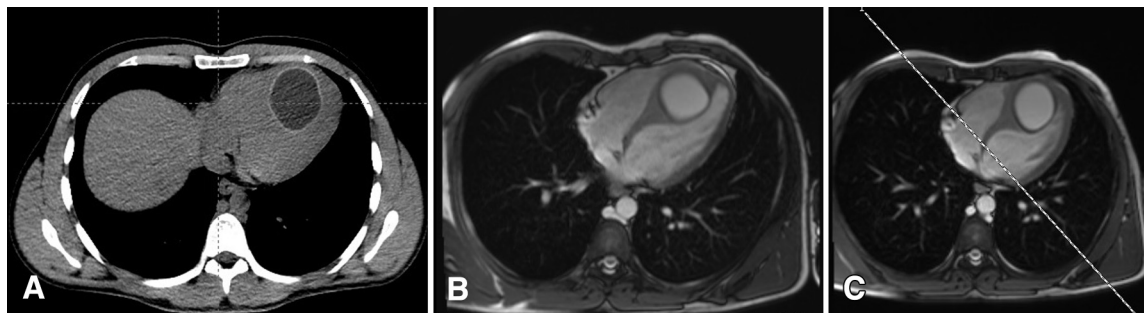


Figure 2. Thoracic axial scan. Echinococcal cyst located in the medial apical interventricular septum. (A) Computed tomography scan view. (B) Magnetic resonance imaging (MRI) scan during diastole. (C) MRI scan during systole with subtotal obliteration of the apex and medio-ventricular space.

The patient was scheduled for surgical treatment of both hydatid cysts. He is now in good general condition and has a normal quality of life.

DISCUSSION

The present case describes the occurrence of Kounis syndrome in a patient with anaphylactic shock secondary to the rupture of a hepatic echinococcal cyst. Kounis syndrome is defined as the occurrence of acute coronary syndromes (i.e., coronary spasm, acute myocardial infarction, and stent thrombosis) in the setting of anaphylaxis (3).

Echinococcosis is a zoonosis caused by four species of echinococci. *Echinococcus granulosus* is the most common cause of human disease (2). Dogs are the definitive hosts, while humans represent intermediate host (2). Although any localization is possible, the most common are hepatic and lung cysts. Cardiac localization is rare (0.5%–3% of cases) and interventricular septum is involved in only 4% of cardiac localizations (4).

This healthy 23-year-old male reported a sudden abdominal pain and then he started feeling facial flush and dyspnea. He was unable to recall other potential symptoms because he had a loss of consciousness, as reported by his coworker who was interviewed by the

medical emergency team. Rupture of hepatic cyst should be considered among the differential diagnoses for acute abdominal pain followed by symptoms of shock. It has been reported that strenuous physical activity can lead to rupture or partial leak of echinococcal cysts (2).

After the initial treatment of anaphylaxis, a 12-lead ECG showed the presence of ST-elevation. Blood work indicated moderately increased troponin levels. After 24 h, the ST-elevation normalized but T-wave inversion remained. These findings deserve additional interpretation, as detailed next.

First, although ECG changes could indicate a myocardial ischemia, coronary arteries were normal at the coronary CT scan and myocardial wall was not showing pathologic enhancement at the MRI.

Second, ECG changes could be attributed to the presence of myocardial cyst. In this regard, cardiac hydatidosis can produce a variety of ECG changes ranging from no alterations, to T-wave inversion, and alterations of ST-segment (5–9). However, unlike in this patient, cardiac enzymes are usually normal.

Our patient showed the presence of two distinct ECG patterns: ST-segment elevation during anaphylactic shock and T-wave inversion after 24 h. The ST-segment elevation is consistent with type I Kounis syndrome caused by anaphylaxis (3). On the other hand, T-wave

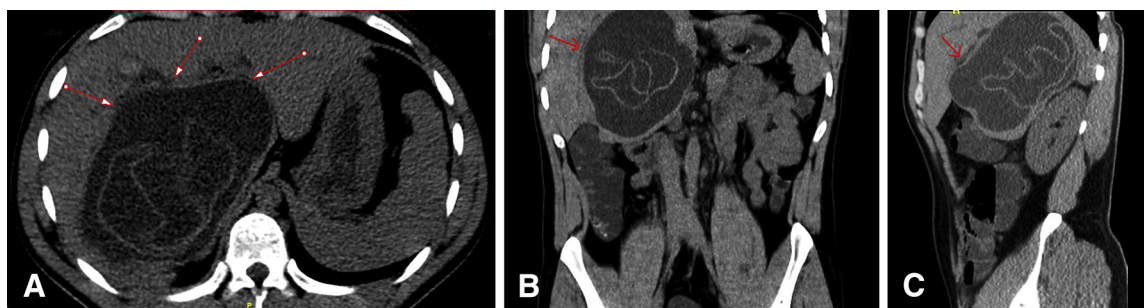


Figure 3. Computed tomography scan of the abdomen. Axial (A), coronal (B), and sagittal (C) images showing a hypodense lesion measuring 15 cm in greatest dimension, located in the right hepatic lobe. There are multiple thin, undulating internal septations, and signs of discontinuity of the cystic wall (arrows). No internal calcifications are visualized.

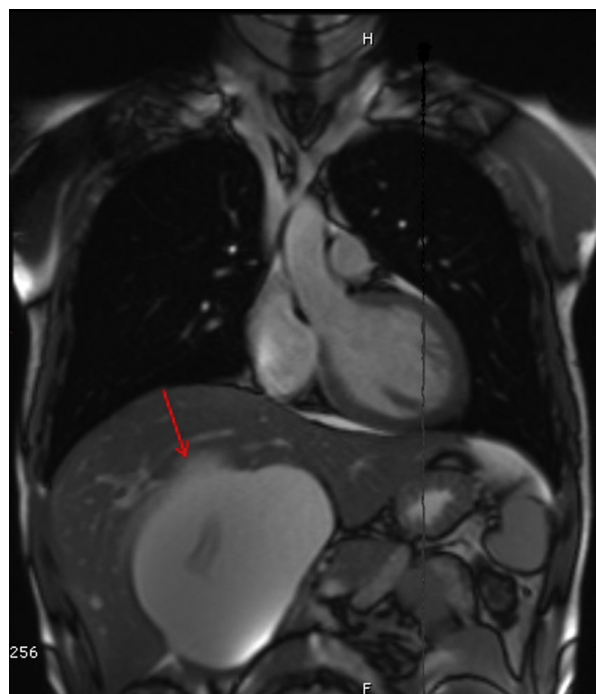


Figure 4. Magnetic resonance imaging scan of thorax and abdomen. Coronal T2-weighted image showing a large hyperintense capsulated cyst. The hyperintensity located around the cyst (arrow) is consistent with leakage of wall cyst.

inversion should be considered as the ECG manifestation of hydatidosis of the interventricular septum.

Interestingly, Alansari and Alsanouri described the appearance of ECG changes during anaphylactic shock occurring in the setting of surgical removal of a hepatic hydatid cyst, in the absence of cardiac hydatidosis (10).

Kounis syndrome, firstly described by Kounis and Zavras in 1991, represents an underdiagnosed clinical entity manifesting as acute coronary syndromes associated with anaphylactic reactions (11). Literature reports from all continents have described this syndrome in patients of any age and both sexes, in association with a variety of allergens, ranging from dietary to pharmacological agents (3). The pathophysiology of Kounis syndrome starts with mast cell degranulation and activation of the allergic cascades. The release and production of pro-inflammatory mediators, such as histamine, proteases, arachidonic acid products, and cytokines, leads to platelet activation and myocardial ischemia (3,12). The type I variant is characterized by coronary artery vasospasm with or without myocardial necrosis. As in the present case, this variant appears in patients with no coronary artery disease (3,11,12). The type II variant manifests itself in those patients with asymptomatic coronary disease. Inflammation produces coronary vasospasm and plaque destabilization with thrombus formation and myocardial infarction (3,11,12). Finally, type III Kounis syndrome affects patients already treated by coronary artery stent in whom eosinophils and mast cells can be evidenced by microscopy (3,11,12).

To the best of our knowledge, this represents the first description of Kounis syndrome occurring during anaphylaxis induced by a ruptured echinococcal cyst.

WHY SHOULD AN EMERGENCY PHYSICIAN BE AWARE OF THIS?

Diagnosing Kounis syndrome in an ED setting has important implications for clinical practice. If fact, its diagnosis requires a change in the treatment of anaphylaxis, that is,

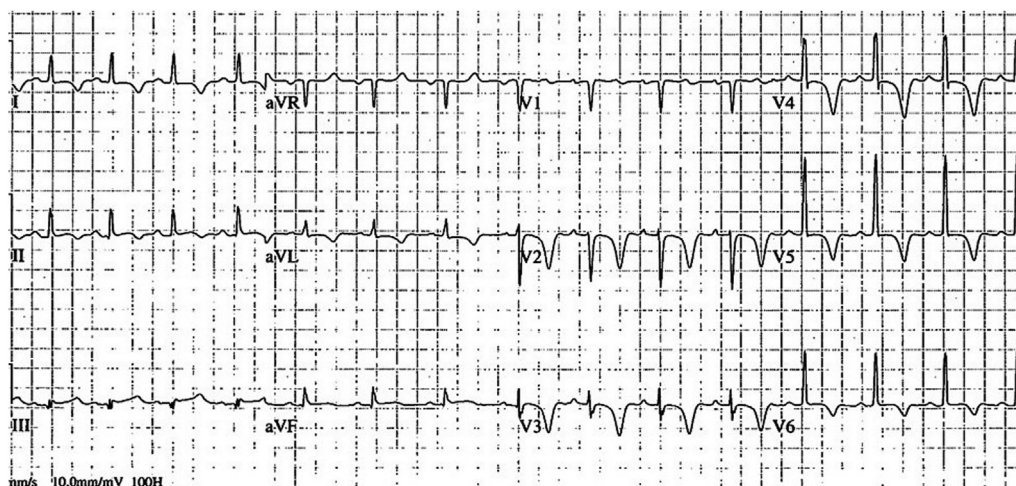


Figure 5. 12-Lead electrocardiography recorded 24 h after admission to the emergency department indicated the regression of ST-segment elevation and the presence of T-wave inversion in I, aVL, and V2-V6.

adrenaline should be avoided as it can worsen vasospasm and myocardial ischemia (3). Thus, a 12-lead ECG should be performed in every patient referring to ED for allergy or anaphylactic symptoms.

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