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Cardiac tamponade as an initial presentation for systemic lupus erythematosus

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ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic autoimmune inflammatory disease which follows a relapsing and remitting course that can manifest in any organ system. While classic manifestations consist of arthralgia, myalgia, frank arthritis, a malar rash and renal failure to name a few, cardiac tamponade, however, is a far less common and far more dangerous presentation. We highlight the case of a 61 year-old male with complaints of acute onset shortness of breath and generalized body aches associated with a fever and chills in the ER. A bedside echocardiogram revealed a significant pericardial effusion concerning for pericardial tamponade. An emergent pericardiocentesis performed drained 800 mL of serosanguinous fluid. While denying a history of any rash, photosensitivity, oral ulcers, or seizures, his physical examination did reveal metacarpal phalangeal joint swelling along with noted pulsus paradoxus of 15–200 mm Hg. Subsequent lab work revealed ANA titer of 1:630 and anti-DS DNA antibody level of 256 IU/mL consistent with SLE. This case highlights cardiac tamponade as a rare but life-threatening presentation for SLE and raises the need to keep it in the differential when assessing patients presenting with pertinent exam findings.

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1. Introduction

One of the most well-recognized "great imitators" in medicine, systemic lupus erythematosus (SLE) is a chronic autoimmune inflammatory disease with a female predominance and variable manifestations that can involve virtually any organ system followed by a relapsing and remitting course. Joint pain such as arthralgia, myalgia and frank arthritis involving smaller joints of the hands, wrists and knees are the classic initial presentation of SLE. Other hallmarks include malar rash, renal failure, seizure activity and non-specific malaise. While cardiac involvement is a known complication, cardiac tamponade is a rare and potentially lethal initial presentation.

2. Description

Our patient is a 61 year-old man with a history of type 2 diabetes mellitus, hypertension and alcoholic liver cirrhosis who presented with shortness of breath and generalized muscle aches associated with subjective fever and chills. He reported intermittent tenderness of the small joints of the hands and wrists for 1–2 years. He denied having rashes, photosensitivity, oral ulcers, seizures or headaches. Initial vital signs included a heart rate of 123 bpm, blood pressure of 101/62 mm Hg, respiratory rate of 25 breaths/min and a temperature of 37.2 °C. Muffled heart sounds and swelling of bilateral metacarpal phalangeal joints were present. Pulsus paradoxus of 15–20 mm Hg was noted. Jugular Venous Pressure (JVP) was estimated at 6 cm H20. Because the cardiac silhouette was markedly enlarged on chest radiography, a CT of the thorax was obtained which revealed a large pericardial effusion (Figs. 1 and 2). Initial telemetry and EKG findings showed new onset atrial flutter (Fig. 3a). A bedside echocardiogram revealed a large pericardial effusion and inferior vena cava plethora with minimal collapse on inspiration. (Fig. 4) There was late diastolic compression of the right atrium deemed to be secondary to pericardial tamponade. Emergency pericardiocentesis was performed, draining over 800 mL of serosanguinous fluid with subsequent EKG showing resolution of the atrial flutter (Fig. 3b). Possible etiologies for the pericardial included viral, tuberculosis, immunologic disorder, malignancy, thyroid disease or drug-induced as the patient has a history of hydralazine use. Serological testing supported the clinical suspicion for SLE including ANA titer of 1:630 and anti-double-stranded (ds) DNA of

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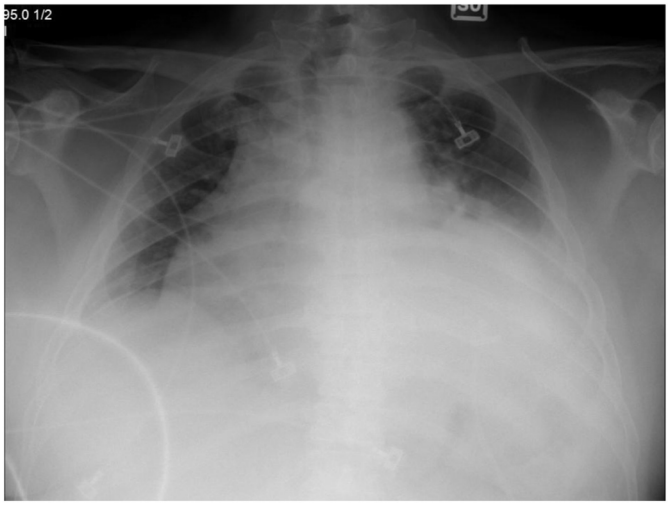

Fig. 1. Portable chest X-ray, antero-posterior view showing enlarged cardiac silhouette.

256 IU/mL. The patient's condition improved using diltiazem and metoprolol tartrate for blood pressure and heart rate control. He was discharged with a prednisone taper and initiation of plaquenil as well as outpatient follow-up with the rheumatology clinic.

3. Discussion

Cardiac tamponade is a medical emergency resulting from accumulation of fluid in the pericardial space that can impair cardiac function. Complications including pulmonary edema, shock and death may result. Therefore, early recognition and intervention is crucial. Cardiac tamponade commonly present with chest pain, shortness of breath, and tachycardia. Our case is unique in that atrial flutter was a manifestation of tamponade. While the classic electrocardiographic findings include low voltage and electrical alternans, atrial flutter has not been described [1]. Furthermore, it is unusual for cardiac tamponade to occur as the initial presentation of SLE.

SLE is a connective tissue disorder with numerous cardiac manifestations including myocarditis, pericarditis and endocarditis [2]. Pericardial effusions are a common finding among SLE patients, with a lifetime occurrence of about 50%. However, cardiac tamponade is a rare finding with a lifetime prevalence of 1% [3]. To our knowledge, only a few instances of cardiac tamponade as the initial presentation of SLE have been described.

The patient's age and gender do not fit the classic female patient between 10 and 50 years of age [4]. One study noted distinct clinical and biological manifestations in patients diagnosed at age 65 or older that differed from the classical presentation. Pericarditis was found to be the most common cardiac manifestation in this group. No reports of cardiac tamponade were noted [5].

The possibility of drug-induced lupus was initially considered as the patient had a history of hydralazine usage. A retrospective study suggested exposure to hydralazine may manifest in lupus diathesis later on in life because of a latency that is unmasked due to other stressors or medical conditions [6]. This initial presentation of SLE may be underappreciated, and early diagnosis could help to better understand the spectrum of manifestations of SLE. Unlike other etiologies of tamponade, connective tissue disorders predispose patients to recurrent serositis. An observational cohort study showed recurrence in 18.7% of patients during their lifetime [7]. If patients remain unaware of their underlying condition, they are at even higher risk for recurrence of medical emergencies such as cardiac tamponade in the future.

In conclusion, cardiac tamponade as an initial presentation of SLE is a rare, but life-threatening condition that should be kept in mind as a possible differential.

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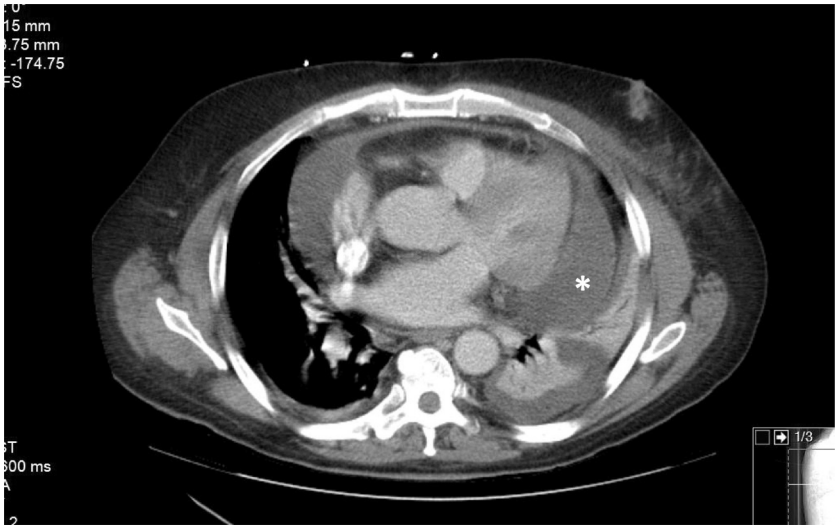

Fig. 2. Chest CT showing pericardial effusion (*).

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