Hydralazine-induced lupus syndrome presenting with large pericardial effusion

M.A.R. CHAMSI-PASHA¹, M. BASSIOUNY² and E.S.H. KIM³

From the ¹Department of Cardiovascular Medicine, University of Nebraska Medical Center, Division of Cardiology, 982265 Nebraska Medical Center, Omaha, NE 68198-2265, USA, ²Department of Internal Medicine, Cleveland Clinic, 9500 Euclid Avenue, Cleveland, OH, 44195, USA and ³Department of Cardiovascular Medicine, Cleveland Clinic, 9500 Euclid Avenue, Cleveland, OH, 44195, USA

Address correspondence to Mohammed A.R. Chamsi-Pasha, University of Nebraska Medical Center, Division of Cardiology, 982265 Nebraska Medical Center, Omaha, NE 68198-2265, USA. email: drpasha.moh@gmail.com

Learning Point for Clinicians

Hydralazine-induced lupus syndrome is uncommon, but timely diagnosis is needed in life-threatening illnesses. Severe organ involvement is exceedingly rare; however, pericardial tamponade has been reported. Patients on chronic hydralazine should be monitored for signs and symptoms of systemic lupus erythematosus periodically. If they develop HILS, immediate discontinuation of hydralazine is warranted to prevent serious complications.

Introduction

Hydralazine-induced lupus syndrome (HILS) was first reported in 1953.¹ The syndrome occurs in 5–10% of patients taking hydralazine, presenting with arthralgia, myalgia, fever and serositis. Severe cardiac involvement is rare; and only three cases of tamponade were reported.¹–³ We describe an unusual case of impending cardiac tamponade in a patient taking hydralazine and coumadin.

Case presentation

A 68-year-old female presented with exertional dyspnea and pleuritic chest pain of 1 week duration. The pain was increased by recumbancy and deep inspiration, and relieved by leaning forward. Associated symptoms included low grade fever but no joint pain or rash. She had paroxysmal atrial fibrillation on coumadin, and hypertension treated with hydralazine 100 mg three times daily for years.

Physical examination revealed a blood pressure of 140/80 with 10 mmHg paradox, sinus tachycardia of 115 bpm, respiratory rate of 20 per minute and normal body temperature. The jugular venous pressure was elevated at 10 cm, and heart sounds were faint with no pericardial rub. Labs revealed anemia (hemoglobin of 6.7 g/dl), with normal white and platelet counts. International normalized ratio (INR) was supratherapeutic at 6.8 (ref, 0.8–1.2). Twelve-lead EKG showed sinus tachycardia, low voltage QRS complexes (Figure 1A). Transthoracic echocardiogram revealed a large circumferential pericardial effusion (Figure 1B and C) with features of impending tamponade and respirophasic variation in the mitral and tricuspid valves of 31 and 50%, respectively. There was no diastolic collapse, but inferior vena cava was dilated at 2.3 cm. Transfusion of fresh frozen plasma was started, and the patient underwent surgical subxiphoid pericardial window. The fluid was hemorrhagic in nature with negative infectious and malignancy workup. Pathology showed evidence of organizing fibrinous exudate, consistent with chronic pericarditis. Erythrocyte sedimentation...
rate was 105 mm/h and C-reactive protein was 11.7 mg/dl. Serologies revealed anti-nuclear antibody (ANA) of 11.5 OD ratio (normal < 1.5 OD ratio), anti-chromatin antibodies 1.7 Al (normal < 1.0) and anti-histone antibodies 5.4 U (normal < 1.0). A diagnosis of HILS was established, and patient was started on high-dose prednisone after stopping hydralazine. She made an excellent recovery with resolution of ef- fusio on repeat echocardiograms.

Discussion

HILS commonly presents with arthritis, myalgia, fever and constitutional symptoms. The incidence is dose dependent; with 10.4% of patients on 200 mg of hydralazine develop it after 3 years treat- ment in slow acetylator patients. Following the publication of A-HeFT trial, there has been an in- crease in the amount of hydralazine prescribed to patients with heart failure. Although the total daily dose of hydralazine used in A-HeFT was just above 200 mg daily, there is good evidence to suggest that these patients and those receiving lower doses are not free from the risk of development of HILS.

Pericarditis occurs infrequently in HILS (<5%) as compared to 20% in the spontaneously occurring systemic lupus erythematos. Anti-his- tone antibodies, a distinctive marker for drug-induced lupus, and ANA are present in >95 % of the syndrome.

Discontinuation of drug inducing lupus is a crit- ical part in the management. Corticosteroids and immunosuppressive therapies are only required in life-threatening cases of HILS.

A timely diagnosis is needed in critical organ ill- nesses, and further diagnostic testing is underscored as early treatment is mandatory.

In conclusion, we present a life-threatening case of HILS in a patient on coumadin, which required timely diagnosis and urgent treatment.

Conflict of interest: None declared.

References


