Case Report

Hypereosinophilic syndrome presenting with junctional rhythm

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Abstract

Hypereosinophilic syndrome is a rare disorder with an incidence rate of 0.36 to 6.3 per 100,000 persons. It is characterized by a sustained overproduction of eosinophils, peripheral eosinophilia, and organ damage by eosinophils. Cardiac involvement is the major cause of death in patients with hypereosinophilic syndrome. Clinical presentations of cardiac involvement include heart failure, intracardiac thrombus, myocardial ischemia, arrhythmia, and pericarditis. Most of these patients present with chest tightness and dyspnea. Arrhythmia is seldom mentioned in these cases. We report an 80-year-old man who presented to the emergency department with junctional bradycardia.

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1. Introduction

Hypereosinophilic syndrome (HES) is a rare disorder characterized by a sustained overproduction of eosinophils, peripheral eosinophilia, and eosinophilic tissue infiltration.1 HES has various clinical presentations, ranging from fatigue and other nonspecific complaints to fatal cardiac and neurological involvement. Cardiac involvement is the major cause of morbidity and mortality in HES.2 The most common symptom of cardiac involvement in HES patients is heart failure, followed by chest pain and systemic thromboembolic events. We report a patient who presented with junctional rhythm in the emergency department (ED) and was subsequently diagnosed with HES.

2. Case report

An 80-year-old man presented to the ED with sudden-onset chest tightness and dyspnea for 5–6 hours. He had a history of hypertension, diabetes mellitus with nephropathy (2 years), chronic kidney disease (stage IV), and duodenal ulcer with irregular medication control. He denied coughing, cold sweats, fever, tarry stools, abdominal pain, and radiating pain. His blood pressure was 116/71 mmHg, his heart rate was 42 beats/min (BPM), and his respiratory rate was 20/min. There were no other remarkable findings during physical examination except for the low heart rate. Electrocardiography (EKG) showed junctional bradycardia, without significant ST-T changes (Fig. 1). The initial laboratory examination revealed a white blood cell count of $33.38 \times 10^3/\mu \text{L}$, with $32\%$ eosinophils (absolute eosinophil count $10.681 \times 10^3/\mu \text{L}$), creatinine 5.4 mg/dL, creatine kinase-MB 1.2 ng/ml, troponin I 0.037 ng/ml, and glucose 238 mg/dL. Results for the hematocrit, coagulation function, electrolyte levels, thyroid function, adrenal function, and other routine biochemical...
analyses were within reference levels. Chest radiography revealed an enlarged heart with Kerley B lines. Based on the symptomatic junctional bradycardia, atropine and dopamine were administered in the ED with no obvious response. The patient was then admitted to the coronary care unit for monitoring and diagnostic evaluation. Dopamine was administered at 10 μg/kg/min. His heart rate was approximately 40–45 BPM. The results of transthoracic Doppler echocardiography showed four chambers with no abnormalities and four functioning valves. Cardiac enzyme analysis showed creatine kinase-MB 13.1 ng/ml and troponin I 6.097 ng/ml. A TI-201 myocardial perfusion scan showed no abnormalities. On the basis of these results, HES was suspected. Prednisolone (0.5 mg/kg per day) was prescribed, and additional diagnostic examinations were performed. Examination of blood and stool showed no evidence of an allergic reaction or parasite infection. The results from serological analysis of autoantibodies to nuclear antigen, complement 3, complement 4, antiextractable nuclear antigen, rheumatoid factor, and immunoglobulins G, A, E, and M were within reference limits. Bone marrow biopsy results revealed no evidence of hematological malignancy. An EKG showed normal sinus rhythm 2 days after admission, and the heart rate was approximately 70 BPM (Fig. 2). Dopamine was discontinued. He was discharged on day 13. One year later, his white blood cell count was 20,000 to 25,000 cells/μL, with 5% to 10% eosinophils, and he had no episodes of bradycardia.

3. Discussion

HES is a rare disease with an estimated prevalence of 0.36–6.3 per 100,000 persons. HES is defined as blood eosinophilia greater than 1500 cells/μL for longer than 6 months with symptoms of organ involvement in the absence of identifiable causes of hypereosinophilia such as parasite infections and allergic reactions. Our patient presented...
with chest tightness and dyspnea, and clinical findings included junctional bradycardia, elevated troponin I, and marked hypereosinophilia. Certain clinical disorders may result in hypereosinophilia with myocardial injury, including HES with cardiac involvement, eosinophilic leukemia with cardiac injury, vasculitis and granulomatous diseases, drug reactions, transplant rejection, and acute coronary syndrome with coexisting HES. Eosinophilic leukemia was ruled out by bone marrow biopsy in our patient. Autoimmune disease, such as Churg–Strauss syndrome, was unlikely because of the absence of asthma, paranasal sinusitis, histological proof of vasculitis, evidence of neuritis, and negative results for serologic markers of autoimmune disease. Drug reactions and transplant rejection were rejected based on the patient’s medical history. Acute coronary syndrome was also ruled out based on a negative TI-201 myocardial perfusion scan. Therefore, the diagnosis was HES with cardiac involvement.

A junctional rhythm initiates within the atrioventricular nodal tissue. In autonomic tone changes or sinus node disease, inappropriate slowing of the sinus node can occur, inducing junctional rhythm. The possible causes of junctional rhythm include sick sinus syndrome, myocardial injury such as cardiac surgery, inflammatory processes involving the conducting system, sinoatrial node ischemia, effects of drugs such as digitalis, and electrolyte imbalance such as hyperkalemia and hypercalcemia. In our patient, drug effects and electrolyte imbalance were ruled on the basis of the patient’s history and initial blood tests. His EKG showed normal sinus rhythm after steroid treatment, and there were no episodes of bradycardia after a 1-year follow-up. Thus, sick sinus syndrome was not favored. Sinoatrial node ischemia was also ruled out based on a negative TI-201 myocardial perfusion scan. So, myocardial injury involving the conducting system was the most likely cause of the junctional rhythm in this case.

The symptoms and signs of HES depend on the degree of end-organ eosinophilic damage. One retrospective study of 188 patients reported that dermatologic presentation was the most frequently reported initial symptom in these patients. However, only 5% of patients presented with cardiac involvement. Nonetheless, cardiac involvement has been shown to be the major cause of morbidity and mortality associated with HES. Clinical presentations of cardiac involvement include heart failure, intracardiac thrombosis, myocardial ischemia, arrhythmia, and pericarditis. Chest pain and dyspnea are the most common symptoms in these patients. EKG results show ST elevation, ST depression, atrial flutter, bundle branch block, and other nonspecific findings. Consequently, patients may be misdiagnosed with acute myocardial infarction. In a retrospective study, Kawano et al reported seven cases over a 27-year period. EKG abnormalities were corrected in two of the seven patients following treatment. Our patient presented with chest tightness and dyspnea, with initial EKG results indicating junctional bradycardia. His EKG showed a corrected sinus rhythm after corticosteroid use.

Specific therapy is not needed in patients with eosinophilia without evidence of end-organ damage. However, HES patients with vital organ involvement require treatment. Corticosteroids remain the first line of treatment for most patients. High-dose prednisolone (1 mg/kg per day) as the initial regimen is recommended in some studies. If patients are refractory to corticosteroids or cannot tolerate them, alternative therapies can be considered. These alternatives include cytotoxic agents, such as hydroxyurea, and immunomodulatory agents, such as alemtuzumab and cyclosporine.

In conclusion, cardiac involvement is the major cause of death in patients diagnosed with HES. HES must be considered in patients with hypereosinophilia in the absence of identifiable causes. Therefore, HES with cardiac involvement should be included in the differential diagnosis of patients reporting chest tightness and dyspnea with associated hypereosinophilia. Corticosteroids should be initiated in these cases.

References