A Case of Hypereosinophilic Syndrome Presenting as Pericardial Effusion, Myocarditis and Ascites

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ABSTRACT

Hypereosinophilic syndrome is a clinical disorder characterized by peripheral eosinophilia and eosinophilic infiltration of multiple organ systems, including the cardiovascular system. The manifestations are variable, but cardiac involvement is the major source of morbidity and mortality, and several case reports have highlighted the various types of cardiac involvement. However, no reported case has simultaneously presented with pericardial effusion, myocarditis, and ascites. We report a case of a 28-year-old woman with hypereosinophilic syndrome involving both the heart and intra-abdominopelvic cavity. (Korean Circ J 2008;38:436-439)

KEY WORDS: Hypereosinophilic syndrome; Myocarditis; Ascites.

Introduction

Hypereosinophilic syndrome (HES) is defined as a state of continuous eosinophilia and eosinophilic infiltration into multiple organs, with no clear causative disease found during broad inspection.1) Cases of infiltration into multiple organs have occasionally been reported in Korea. However, there has been no report of a case where eosinophilic infiltration occurred simultaneously in both the heart and gastrointestinal tract at an early clinical stage. The authors report a case of an HES patient presenting with pericardial effusion, myocarditis, and ascites. A literature review is included.

Case

A 28-year-old woman was admitted to the hospital secondary to right upper quadrant (RUQ) abdominal pain and lower abdominal pain, both of which had suddenly developed one day earlier. The patient had no significant past medical history or family history. She had a mild fever of 37.5°C. Her breath sounds were clear, except for some crackles in both lower lung fields. The patient had tenderness in the RUQ and lower abdomen. The absolute eosinophil count was 1694/mm³. The following diagnostic tests yielded negative results: sputum/urine/blood cultures, stool parasite egg examination, hepatitis B and C markers, VDRL, Human immunodeficiency virus antibody (HIV Ab), Anti-nuclear antibody (ANA), anti-dsDNA, Hantaan virus Ab, leptospiira Ab, O. tsutsugamushi Ab, Human papiloma virus (HPV) genotype, parasite-specific IgG (micro-Enzyme-Linked ImmunoSorbent Assay (micro-ELISA test)), Cysticercus Ab, Paragonimus Ab, Sparganum Ab, and Clonorchis Ab. C-reactive protein (CRP) was 23.4 mg/L, total IgE was 603.4 IU/mL, troponin T was 0.712 ng/mL, and N-terminal pro-B-type natriuretic peptide (NT-proBNP) was 10,674 pg/mL. All of these were increased above the normal reference range. An electrocardiogram showed normal sinus rhythm and diffuse, non-specific ST segment changes. A chest x-ray and computed tomography of the chest revealed cardiomegaly, pericardial effusion, and bilateral pleural effusions (Fig. 1A and B). Computed tomography of the abdomen and pelvis demonstrated ascites in both the gallbladder fossa and pelvic cavity (Fig. 1C and D). Examination of ascitic fluid obtained from culdocentesis showed an exudate (protein 4.3 g/dL, albumin 2.5 g/dL, and lactate dehydrogenase (LDH) 216 IU/L) with many polymorphonuclear leukocytes (PMN). An ultrasound of the abdomen and pelvis demonstrated pelvic ascites, edema of the gallbladder wall, and periportal thickening. A hepatobiliary scan revealed no abnormalities. An echocardiogram demonstrated mild left ventricular dysfunction with an ejection fraction of approximately 45%. It also showed multiple...
regional wall motion abnormalities incompatible with the coronary territory, a moderate pericardial effusion of no hemodynamic significance, and increased thickening of the basal inferoposterior wall (Fig. 2). A coronary angiogram revealed no abnormalities.

An endomyocardial biopsy revealed a markedly increased eosinophilic infiltration of the endomyocardial tissue (Fig. 3).

The patient was treated with oral glucocorticosteroids (prednisolone, 40 mg once daily) starting on day 5 of admission, and her symptoms and eosinophilia improved. A follow-up examination consisting of a chest x-ray, an
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Echocardiogram, and an abdominopelvic CT showed marked improvement of her condition (Fig. 4). The patient is now well and has been followed continuously in the outpatient clinic for the past 10 months.

Discussion

In 1975, Chusid et al.\textsuperscript{2} from the National Institutes of Health (NIH) defined the criteria for the diagnosis

Fig. 3. Endomyocardial biopsy revealed a markedly increased number of eosinophils infiltrating the endomyocardial tissue. A: H & E stain, ×100. B: H & E stain, ×400. H & E: hematoxylin and eosin.

Fig. 4. Follow-up images. A: chest X-ray showed no cardiomegaly or pleural effusion. B: echocardiography showed that the pericardial effusion and abnormal echogenicity of the posterolateral wall had resolved. C, D: abdominal CT demonstrated marked improvement of ascites in the gallbladder fossa and pelvic cavity.
of hypereosinophilic syndrome. First, the patient must have persistent eosinophilia of more than 1,500/mm³ in the peripheral blood for at least six months, or the eosinophil count (related to the symptoms of HES) must be consistently over 1,500/mm³ six months before death. Second, parasitic infection, allergic disease, and other possible causes of the clinical signs and symptoms must not be present. Third, there must be signs and symptoms of organ system involvement or dysfunction that can be directly correlated to the eosinophilia, or are otherwise unexplained in the clinical setting. The signs and symptoms of HES are related to involvement of multiple organ systems. The heart is the second most frequently infiltrated organ after the bone marrow, and eosinophilic infiltration into the heart is the most important determinant of mortality. Of 51 patients with HES evaluated at the Mayo Clinic, 29 had the characteristic echocardiographic findings. These findings included limited motion of the posterior mitral leaflet (resulting in mitral regurgitation of varying severity) in combination with thickening of the inferobasal left ventricular wall, endocardial thrombotic-fibrotic lesions, and biventricular apical obliteration by thrombus. In our patient, echocardiography showed thickening of the basal postero inferior wall without fibrosis or thrombosis, which suggested an early stage of cardiac involvement. Advancements in echocardiographic techniques and therapy have allowed for early diagnosis and control of cardiac complications, and the prognosis of patients with HES has subsequently improved. Our patient had no notable cardiac symptoms, as she was diagnosed at an early stage. Her prognosis is good because she underwent immediate treatment and experienced rapid improvement of symptoms. Gastrointestinal tract symptoms associated with HES vary depending on the layer of the gut wall into which the eosinophils infiltrate. Ascites and peritonitis are associated with infiltration into the subserous layer, and HES can be diagnosed based on increased eosinophils in ascites fluid. Our patient presented with ascites and peritonitis-like symptoms, and she rapidly improved after corticosteroid therapy was instituted. We believe eosinophils infiltrated the subserous layer of the gastrointestinal tract based on the rapid improvement of clinical symptoms and ascites after corticosteroid treatment and based on the fact that there were many polymorphic neutrophil (PMNs) in the ascitic fluid. Because HES is accompanied by infiltration into various organs, its treatment should focus on the control of organ dysfunction and eosinophilia. If there is no organ dysfunction, periodic follow-up is conducted every three to six months, with no special treatment indicated. If organ dysfunction is noted, then steroids (prednisone 1 mg/kg/day) are given every day for one to two weeks, followed by the same dosage given every other day for the next three months.

Our patient exhibited various clinical symptoms attributable to multi-organ infiltration. There have been no other reports in Korea of patients with simultaneous infiltration into both the heart and intra-abdominopelvic cavity. The possibility of HES must not be ignored when a patient presents with peripheral eosinophilia and pathology of multiple organs, for immediate treatment can be lifesaving.

REFERENCES