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Case Report / Olgu Sunumu

Idiopathic Hypereosinophilic Syndrome as a Rare Cause of Stroke: A Case Report

İnmenin Nadir Bir Nedeni, İdiyopatik Hipereozinofilik Sendrom: Olgu Sunumu

Esra Turğut[1], Murat Alpua[1], Selim Yalçın[2], Oya Coşkun[3], Mirace Yasemin Karadeniz Bilgili[4], Ergin Ayaşlıoğlu Açıkgöz[5], Özlem Coşkun[1]

[1]Kirikkale University Faculty of Medicine, Department of Neurology, Kirikkale, Turkey
[2]Kirikkale University Faculty of Medicine, Department of Medical Oncology, Kirikkale, Turkey
[3]Kirikkale University Faculty of Medicine, Department of Psychiatry, Kirikkale, Turkey
[4]Kirikkale University Faculty of Medicine, Department of Radiology, Kirikkale, Turkey
[5]Kirikkale University Faculty of Medicine, Department of Infectious Diseases, Kirikkale, Turkey

Abstract

Hypereosinophilic syndrome (HES) is a rare hematological disease that causes organ damage by eosinophil infiltration in the tissue with increased eosinophil production in the bone marrow. HES is a rare but important cause of stroke. Central nervous system involvement findings can be serious and life-threatening. Eosinophil values should be examined as the cause of stroke, and hypereosinophilia should be suspected, especially in young patients with no etiology. Reported herein is a case of a 47-year-old female patient who was followed up with the diagnosis of acute cerebrovascular disease due to idiopathic HES.

Keywords: Cerebrovascular accident, hypereosinophilic syndrome, ischemic stroke, hypereosinophilia

Öz

Hipereozinofilik sendrom (HES) kemik iliğinde eozinofil üretiminin artması ve dokuların eozinofillerle infiltrasyonu ile seyreden nadir bir hematolojik hastalıktır. Merkezi sinir sistemi bulguları ciddi ve hayatı tehdit edici olabilir. İnme nedeni olarak eozinofil değerleri takip edilmeli ve özellikle etiyolojisi olmayan genç inme nedeniyle izlenen hastalarda hipereozinofili akla gelmeli. Bu olgu sunumunda idiyopatik hipereozinofili sendromuna bağlı akut serebrovasküler hastalık tanısı ile izlenen 47 yaşında bir kadın hastayı sunuyoruz.

Anahtar Kelimeler: Serebrovasküler hastalık, hipereozinofili sendromu, iskemik inme, hipereozinofili

Introduction

Hypereosinophilic syndrome (HES) is a rare hematological disease that causes organ damage by eosinophil infiltration in the tissue with increased eosinophil production in the bone marrow. The following criteria are taken into consideration in the diagnosis: Eosinophil count >1500 cells/mcl that lasts longer than 6 months; the absence of diseases that can cause eosinophilia, such as allergic diseases and parasitic infections; and the presence of signs and symptoms of organ manifestations. The most frequently involved organs are the heart, skin, lungs, and central and peripheral nervous systems. Less frequently involved are the liver, eyes, and gastrointestinal system (1,2).

Herein, presented is a 47-year-old female patient who was followed up with the diagnosis of acute cerebrovascular disease due to idiopathic HES.

Case Report

A 47-year-old female patient with hypertension consulted our clinic with complaints of weakness, sleepiness, double vision, and right hand weakness for one day. A neurological

Address for Correspondence/Yazışma Adresi: Esra Turğut MD, Kirikkale University Faculty of Medicine, Department of Neurology, Kirikkale, Turkey
Phone: +90 542 732 58 69 E-mail: dresracicekturgut@gmail.com ORCID: orcid.org/0000-0002-7166-6649
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examination revealed that she was lethargic, oriented, and cooperation was limited. Muscle strength was 3/5 on the right upper extremity distally, 4/5 proximally, and 5/5 on the other 3 extremities. Pathological reflexes were not found. The diffusion-weighted images detected acute-subacute infarcts in the bilateral frontoparietal border areas, bilateral thalamus and basal ganglia, right mesencephalon, right temporal and bilateral occipital lobe, and bilateral cerebellar hemisphere (Figure 1). She was hospitalized with the diagnosis of acute ischemic cerebrovascular disease, and antiaggregant therapy was started. The patient was evaluated by the department of cardiology to exclude the diagnosis of possible infective endocarditis (IE) due to existing magnetic resonance imaging findings in the patient who was considered to have a young ischemic stroke, and IE was not detected in the patient who underwent trans-esophageal echocardiography. The carotid and vertebral Doppler ultrasonography and neck and brain angiography examinations were normal. No pathology was found in routine blood tests, except for increased sedimentation, elevated C-reactive protein, eosinophilia, and elevated lipid panel. The genetic panel test and vasculitis markers were normal. It was noteworthy that the eosinophilia persisted during patient follow-up. The parasitic disease was not detected in the patient when evaluated by the infectious diseases department in terms of possible parasitic infectious diseases. The current cerebrovascular event of the patient was thought to be due to eosinophilia. The patient was consulted with the hematology department, and hypereosinophilia was also present in the peripheral smear test. When the patient's general medical condition worsened and respiratory distress began, the patient was started on a methylprednisolone infusion at 1 g/day. On the first day of treatment, marked improvement was noted in both the general medical condition and eosinophil values of the patient (Graphic 1).

A voluntary informed consent form was obtained from the patient before the preparation of this case report.

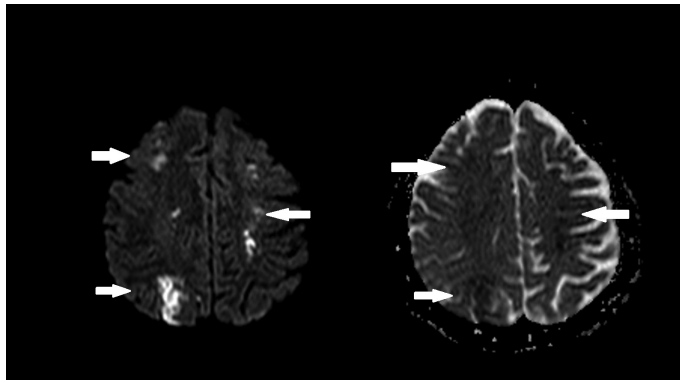
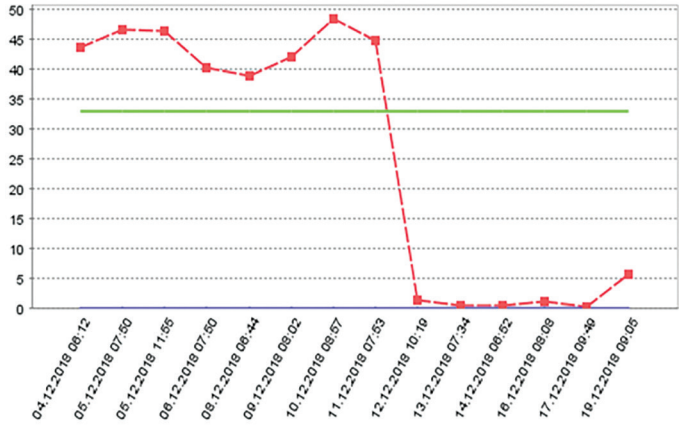


Figure 1. Lesions compatible with acute ischemia in bilateral watershed zones in DWI and ADC

DWI: Diffusion-weighted images, ADC: Apparent diffusion coefficient



Graphic 1. On the first day of treatment, marked improvement was noted in both the general medical condition and the eosinophil values of the patient

Discussion

HES, is a rare but highly specific neurovascular syndrome (3) and is a fast-progressing disease that can cause heterogeneous and multiple organ damage with various symptoms (4). The disease is more common in men and ages 20-50 years (1,2). The granular proteins in active eosinophils cause clotting and fibrinolysis, which results in thrombosis after endothelial injury. Neurological involvement may be in the form of encephalopathy, brain infarction, and sensory polyneuropathy (5). Cerebral infarction develops with endomyocardial thromboembolism due to vascular endothelium toxicity after the release of mediators by eosinophils (3,4,5,6). Infarcts are small and can develop in the arterial border areas only and later in large cortical and subcortical regions.

The exact pathogenesis of encephalopathy is unknown, but encephalopathic changes are associated with significantly higher eosinophil rates and infarctions in the cerebral-arterial border zone areas.

Another neurological dysfunction described in patients with HES is peripheral-neuropathies and is the neurological finding that occurs in approximately half of the patients with HES (7).

The first treatment option should be a high-dose corticosteroid (intravenous methylprednisolone treatment at 1 mg/kg/day) for conditions such as cardiac and neurological involvement and marked eosinophilia. Patients who do not respond to the corticosteroid therapy are administered imatinib. Vincristine and a much higher dose of corticosteroids can be administered to patients with life-threatening rapidly progressing conditions at 10-20 mg/kg/day. Other drugs that can be used in the treatment of HES include interferon, etoposide, chlorambucil, hydroxyurea, and cyclosporine (8,9).

In this case report, we presented a case and the treatment that was followed up in our clinic due to HES-related cerebrovascular disease. Hypereosinophilia should be suspected to be a cause of cerebrovascular disease especially in patients with ischemic lesions at a young age with borderline zones and multiple locations.

The characteristic features of a stroke due to HES are the emergence of multiple ischemiae in different vascular areas. HES is a syndrome that should be kept in mind that can change the treatment options in patients with bilateral, multiple localization, and border area infarction with unknown etiology. This case report aimed to emphasize that HES can cause multiple embolic infarcts.

Ethics

Informed Consent: A voluntary informed consent form was obtained from the patient before the preparation of this case report.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: M.A., Concept: S.Y., Design: O.C., Data Collection or Processing: M.Y.K.B., Analysis or Interpretation: Ö.C., Literature Search: E.A.A., Writing: E.T.

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