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Primary Hypereosinophilic Syndrome and Stroke: Mechanisms and Diagnosis

Primer Hipereozinofilik Sendrom ve İnme: Mekanizma ve Tanı

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Dear Editor,

We would like to express some of our views on the case report by Turğut et al. (1) about a patient with hypereosinophilic syndrome (HES) presenting with stroke, which was accepted for publication in the Turkish Journal of Neurology. First of all, we congratulate the authors for successfully making this rare diagnosis. At this point, we should note that many case reports from our country were also published, but not cited by the authors (2). Stroke presentation is a well-known but extremely rare phenomenon in HES.

Two questions arose in our minds regarding this case report:

Firstly, the diagnosis of HES can be made by detecting eosinophilia in the complete blood count in the blood sample collected immediately in the emergency department, that is, the result is obtained within hours at the latest. But, it took a quite long time to diagnose and initiate treatment in this particular patient. In fact, treatment was just started after development of respiratory distress. Was there any special reason for this significant delay in diagnosis in this particular patient? Examining and sharing the reasons for this unfortunate delay may be helpful for neurologists who manage such patients. A case vignette exemplifying this approach is given in Figure 1.

Our second question is about follow-up duration. Only one-day follow-up information was provided regarding the post-treatment responsiveness and progress. However, as casual readers with specific interest in HES, we wondered the fate of the patient. What kind of treatment was added to the steroid treatment? What did the examinations for the etiology of HES such as bone marrow biopsy show?

We would like to say a few words about the mechanism. The authors stated that HES could cause multiple embolic infarctions and this was important in young people with unexplained stroke. However, the source of the embolism could not be demonstrated in the patient presented herein.

It is important to note that a cardiac magnetic resonance imaging (MRI) is always required in HES for the demonstration of intracardiac thrombi, and transesophageal echocardiography is not sensitive enough in this situation. MRI can document eosinophilic myocarditis or Loffler's fibroblastic endocarditis, resulting in endomyocardial fibrosis and restrictive cardiomyopathy, which has an important role in long-term morbidity. MRI is also critical for documentation of intracardiac thrombus and again is more sensitive than transesophageal echocardiography. This point should not be missed in the work-up for the source of embolism in HES.

While discussing embolism in HES, it is necessary to explain why the distribution of emboli is not random and why the emboli accumulate dominantly in watershed regions. The cranial MRI of this patient did not look like infective endocarditis. The MRI of the patient showed randomly distributed embolism which was contrary to what the authors stated (3). In HES, innumerable watershed microinfarctions are seen, microcirculatory blockage occurs due to impaired wash-out (clearance) of emboli caused by the underlying leukoproliferative disease (4). The microvascular block also leads to endothelial toxicity and/or direct capillary occlusions due to numerous trapped eosinophils in the vasculature. Therefore, its treatment is not at all similar to cardioembolic stroke. This point should not be missed in the treatment plan of HES.

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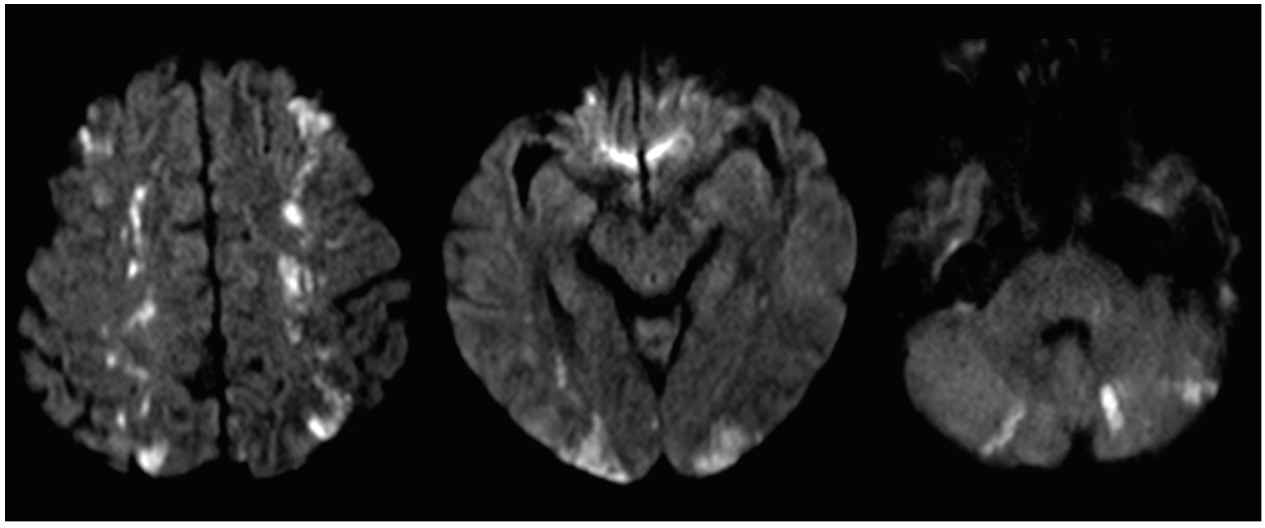

Figure 1. A 50-year-old female patient who was treated for chronic myelocytic leukemia presented with acute abdominal pain, nausea, vomiting, diarrhea, accompanying unconsciousness, and right hemiplegia. On examination, there were stupor, quadriparesis, and diffuse skin rash. Diffusion-weighted imaging performed 270 minutes after the patient entered the emergency room revealed multiple punctuated areas of diffusion restriction in the border regions of the anterior and posterior circulations. A complete blood count available 30 minutes after admission showed that eosinophil count was 18.810 cells/µl. A diagnosis of HES was made 280 minutes after admission to the emergency room
HES: Hypereosinophilic syndrome

Ethics

Informed Consent: Informed consent was obtained for the patient whose magnetic resonance images are shown in Figure 1.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: M.Ç., M.A.T., Design: M.Ç., M.A.T., Data Collection or Processing: M.Ç., M.A.T., Analysis or Interpretation: M.Ç., M.A.T., Literature Search: M.Ç., M.A.T., Writing: M.Ç., M.A.T.

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