

Acute thrombotic complication of essential thrombocythemia in a young adult

Esansiyel trombositeminin genç erişkindeki akut trombotik komplikasyonu

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To the Editor,

We read with great interest the recent publication by Arkan et al. on central retinal artery occlusion (CRAO) in a newly diagnosed patient with essential thrombocythemia (ET), and their conclusion that ET is among the underlying hematologic disorders in patients with CRAO [1]. ET is a myeloproliferative disorder associated with an increase in abnormal platelets that causes both hemorrhagic and thrombotic pathology. Some of its systemic complications include deep vein thrombosis, pulmonary emboli, myocardial infarcts, and renal vessel thrombosis [2]. We would like to share our experience concerning the causative relationship between ET and CRAO.

A 29-year-old male presented with acute visual loss in his right eye. Upon examination his right eye visual acuity (VA) was counting fingers at 0.5 m and the relative afferent pupillary defect was positive in the same eye. Fundus examination revealed retinal edema and a cherry-red spot appearance of the

macula with narrowed vessels, which were compatible with the diagnosis of CRAO. Ocular massage, anterior chamber paracentesis, and systemic therapy with a carbonic anhydrase inhibitor and mannitol were initiated. At presentation the patient's routine laboratory test results were normal, except for thrombocytosis (platelet count: $1.278 \times 10^9/L$); following this result, the patient was referred to our clinic.

The patient had a history of diabetes mellitus type 1 (controlled with insulin). The patient's platelet count was $1.324 \times 10^9/L$, which was in accordance with the peripheral blood smear that showed an increase in the number of platelets and clumps, and bone marrow biopsy showed an increase in the number of megakaryocytes. The white blood cell count was $11.6 \times 10^9/L$ and hemoglobin was 15.5 g/dL. Physical and ultrasonographic examination showed splenomegaly. Hematological results for prothrombin time, partial thromboplastin time, fibrinogen, antithrombin III, protein C, protein S, activated protein C resistance, lupus anticoagulant, antinuclear antibody, and the erythrocyte sedimentation rate

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were unremarkable. The homocysteine level was 17.9 (normal range: 5.4-16.2 $\mu\text{mol/L}$). Factor V Leiden 1691 G>A and factor II 20210 G>A were normal.

Conventional cytogenetic analysis of a bone marrow specimen was normal. The patient was heterozygous for JAK2 V617F mutation, based on real time PCR analysis. The final diagnosis of ET was then established. Therapeutic thrombopheresis was performed once, followed by low-dose acetyl salicylic acid and hydroxyurea treatment. At the 1-month follow-up the patient's platelet count was within the normal range, but his VA had not improved completely. Maintenance therapy was planned for the appropriate dose and duration.

ET is associated with systemic and ocular thrombotic and embolic complications. The risk for thrombosis is highest in patients with a history of thrombosis and in those aged >60 years [3]. Although ET is primarily considered a disorder of middle-advanced age, it has been observed in children and young adults; however, the actual risk for thrombosis in young patients has not been clearly established [4]. We think that this complication should be kept in mind, especially in young adult patients with acute visual loss.

Ethical Consideration

Written informed consent was obtained from the patient.

Conflict of interest statement

The authors of this paper have no conflicts of interest, including specific financial interests, relationships, and/or affiliations relevant to the subject matter or materials included.

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