



# A Case with Two Rare Etiologies of Ischemic Stroke: Eosinophilic Granulomatosis with Polyangiitis and Syphilis

## İki Nadir Etiyolojinin Birlikte Olduğu Bir İskemik İnme Olgusu: Eozinofilik Granülomatöz Polianjit ve Sifiliz

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### ABSTRACT

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg-Strauss syndrome, is a rare disease with asthma, eosinophilia, and systemic vasculitis. EGPA may be among the rare causes of ischemic stroke. Neurosyphilis, on the other hand, is a disease that can mimic many diseases due to its wide clinical symptoms, and the diagnosis is often delayed or missed. Some of these patients present to the emergency services with acute stroke and are only diagnosed after suffering a cerebrovascular complication. We present this case, because two rare causes of ischemic stroke are together.

**Keywords:** Eosinophilic granulomatosis with polyangiitis; intravenous thrombolysis; ischemic stroke; stroke etiology; stroke; syphilis; vasculitis.

### ÖZET

Polianjit ile birlikte olan eozinofilik granülomatöz, önceki adıyla Churg-Strauss sendromu, nadir rastlanılan; astım, eozinofili ve sistemik vaskülit ile seyreden bir hastalıktır. Eozinofilik granülomatöz polianjit iskemik inmenin nadir nedenleri arasında gösterilebilir. Nörosifiliz ise geniş klinik belirtileri nedeniyle birçok hastalığı taklit edebilen ve sıklıkla tanısı atlanılan bir hastalık olup, bu hastaların bir kısmı günümüzde acil servislere akut inme ile gelmekte ve ancak bu şekilde tanı almaktadır. Buradaki olgu iskemik inmenin iki nadir nedeni bir arada olduğu için sunuldu.

**Anahtar sözcükler:** Eozinofilik granülomatöz polianjit; inme; inme etiyolojisi; intravenöz tromboliz; iskemik inme; sifiliz; vaskülit.

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly known as Churg-Strauss syndrome, is a rare disease with asthma, eosinophilia, and systemic vasculitis. Most patients develop vasculitis with diffuse granulomatous involvement in small and medium vessels after 3 years of the initial symptoms of the disease.<sup>[1]</sup> Although EGPA is mostly associated with involvements such as lung, heart, liver, spleen, and kidney; in the study conducted on 187 patients with EGPA, 41 (22%) patients had cerebrovascular involvement.<sup>[2]</sup>

Neurosyphilis is a disease that can mimic many diseases due to its wide clinical symptoms and is often missed. Some of these patients come to the emergency departments with acute stroke and they are only diagnosed in this way.<sup>[3]</sup>

### Case Report

A 63-year-old right-handed male patient was admitted to our emergency department with the complaint of weakness in the left arm, which started 4 h ago. It had been learned that he has treated for 3 years with the diagnosis of EGPA.

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While he was examined for rash and cough in 3 years ago; pulmonary nodule, IgE increase, and pulmonary effusion were determined. He was treated with a high dose of steroids, a course of rituximab, and endoxan with the diagnosis of EGPA. He used 3 g of endoxan and two doses of rituximab adding up to 1 g totally. On discovered being anti-HbcIgG (+), he used prophylactic Tenoviral 2 years ago. On admission, he was only using prednisolone 10 mg/day.

The patient was awake and alert. His speech was dysarthric. Cranial nerves were normal. On motor examination, the right upper extremity muscle strength was 1/5, left upper extremity muscle strength was 5/5, and lower extremities muscle strength was 5/5. Sensorial examination was intact. Cerebellar examination and gait were normal. No pathologic reflexes were discovered.

No hemorrhage was observed in the cranial computed tomography (CT). Craniocervical CT angiography did not reveal any major vascular pathology requiring endovascular intervention. National Institute of Health Stroke Score (NIHSS) was 6. The patient was treated with alteplase 0.9 cc/kg. Acetylsalicylic acid (ASA) 300 mg/day was added to the treatment of the patient who did not have hemorrhage on the control CT taken at the 24th h of thrombolytic therapy (Fig. 1). Diffusion-weighted imaging magnetic resonance imaging revealed a left middle cerebral artery ter-

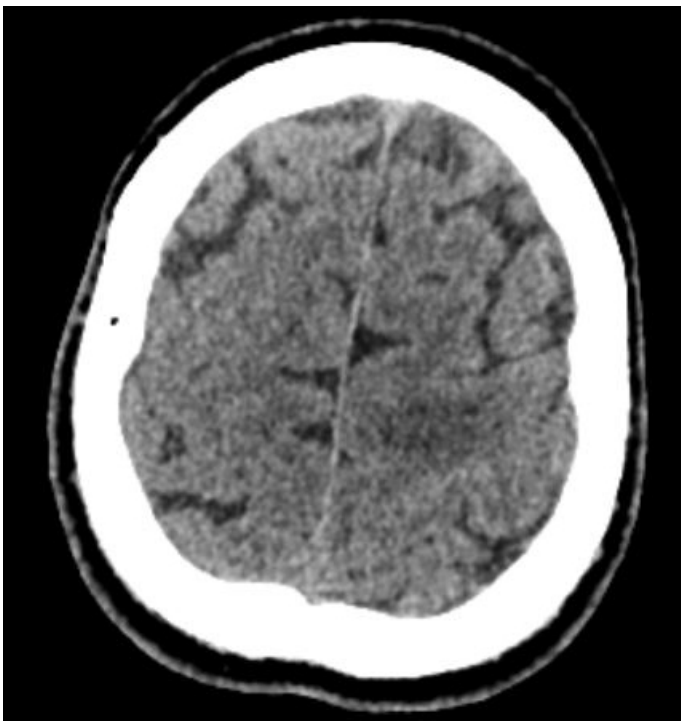


Figure 1. No hemorrhage on the control computed tomography taken at the 24<sup>th</sup> h.

ritory infarction (Fig. 2). At the 24<sup>th</sup> h of the patient, the NIHSS was 4.

The lipid profile and HbA1c values were found within normal limits. No cardioembolic focus was found in the cardiac examination. During observations, all of the fasting glucose measurements were within normal limits. All of the blood pressure measurements were within normal limits as well. The patient's body mass index was within the healthy weight range. None of the observations revealed apparent risk factors for atherosclerosis.

Venereal disease research laboratory (VDRL) which is a part of our routine stroke screening test was positive. The patient had no history of suspected sexual contact or blood transfusion. Treponema pallidum hemagglutination assay (TPHA) was requested for a definitive diagnosis and it was positive as well. Penicillin G 2400000 unite/week treatment was started.

The patient with clinical improvement was discharged with ASA 100 mg 1x1, prednisolone 10 mg/day and penicillin treatment.

## Discussion

In the literature, there are studies reporting that people with EGPA have increased frequency of neurovascular disease.<sup>[4]</sup> It is also known that vasculitis such as EGPA cause an increase in intracardiac thrombus, diastolic dysfunction, and valve pathologies. Therefore, systemic vasculitic diseases can be both the cause of stroke with the direct neurovascular damage or by increasing the frequency of cardioembolic stroke with the cardiac pathologies they cause.<sup>[5]</sup> The high blood sugar level that may occur secondary to long-term steroid use may also increase the frequency of small vessel disease in patients.

Syphilis, on the other hand, may occur with clinical presentation in the form of acute ischemic stroke in some patients. Meningovascular neurosyphilis frequently occurs in 5 or 12 years of infection and affects 3% of untreated patients. It mostly affects middle cerebral artery territory.<sup>[1]</sup> Patients who are not diagnosed are unable to receive appropriate treatment; therefore, in the future, they apply to the hospital with more severe clinical pictures. Without treatment the disease can progress over years through a series of clinical stages and lead to irreversible neurological or cardiovascular complications.<sup>[6]</sup> For this reason, it is important to perform syphilis screening in patients who belong to the young stroke group, who have a history of unprotected sex-

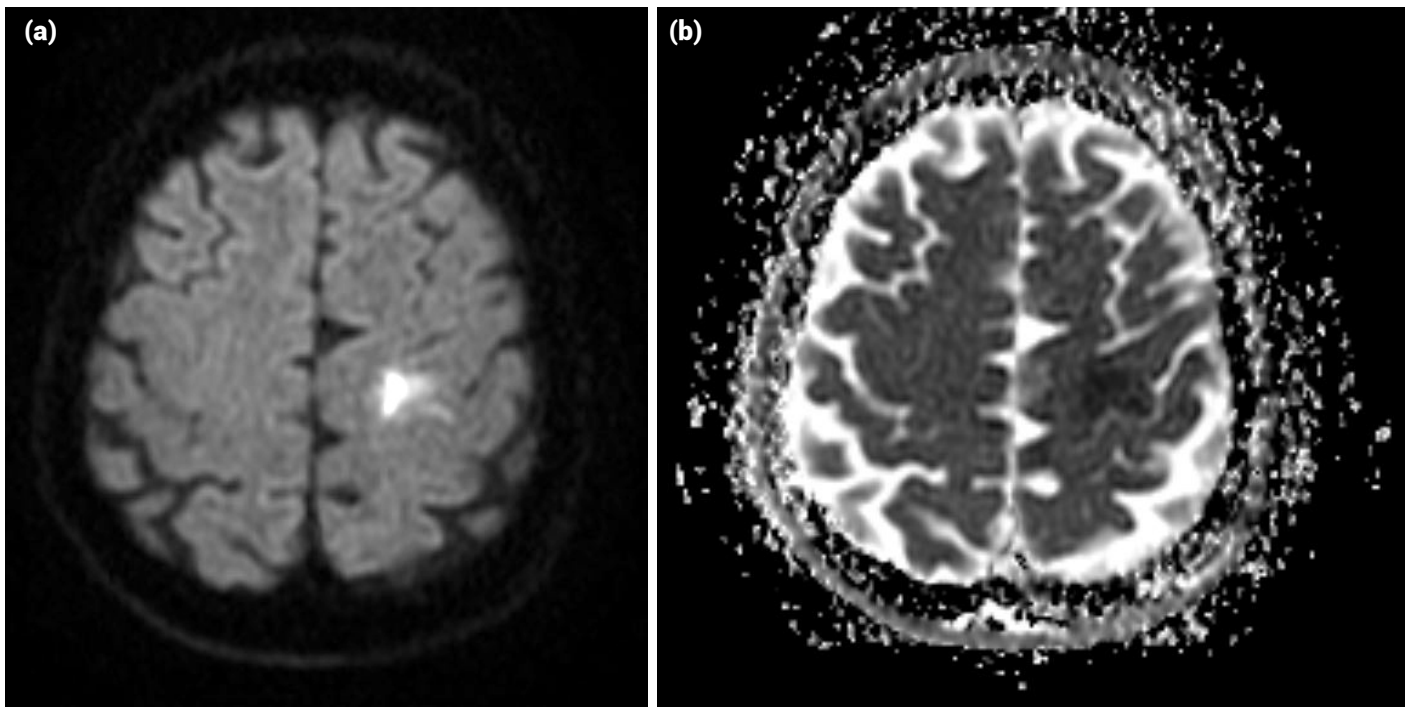


Figure 2. Left middle cerebral artery infarction on diffusion-weighted magnetic resonance imaging.

ual contact and also to the middle-aged and elderly patients without apparent etiologies.<sup>[7]</sup> VDRL is recommended as a screening tool; however, it has a high false positive rate. Especially in cases of pregnancy, older age and autoimmune diseases, the rate of VDRL false positivity rate increases; therefore, TPHA test should be ordered to confirm the diagnosis of syphilis.<sup>[8]</sup>

In this case, the patient has no conventional risk factors such as hypertension, diabetes mellitus, or hyperlipidemia. Large artery atherosclerosis or cardioembolic pathology were not detected. The patient was being treated for vasculitis and he was diagnosed with syphilis during the follow-up. Lack of the conventional risk factors and the fact that the patient had two of the rare causes according to the TOAST classification led us to the conclusion that the reason of the ischemia laid in these two rare etiologies.

This case has been presented to highlight the combination of two rare causes in the “other etiological causes” group, which is seen below 1% according to the TOAST classification.

#### Disclosures

**Informed consent:** Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

**Peer-review:** Externally peer-reviewed.

**Conflict of Interest:** None declared.

**Authorship Contributions:** Concept – A.A.; Design – I.K.A.; Supervision – E.G.; Materials – I.S.; Data collection &/or processing – A.A.; Analysis and/or interpretation – I.S.; Literature search – A.A.; Writing – A.A.; Critical review – I.S.

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