LETTER TO THE EDITORS

Dissection of the posterior inferior cerebellar artery in the hypereosinophilic syndrome

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Dear Sirs,

Hypereosinophilic syndrome (HES) is a rare disorder, and to set the diagnosis patients should have persistently elevated eosinophil counts over 1,500/mm³ for 6 months or evidence of end organ damage from eosinophilia, and other causes should be excluded, such as a reactive phenomenon or underlying haematological malignancy [2, 13].

Different types of HES exist: T-lymphocytic variants (L-HES); Myeloproliferative variants (M-HES); Familial HES; Idiopathic HES; and HES associated with vasculitis (like Churg–Strauss vasculitis) [6].

In patients with HES stroke, encephalopathy and neuropathy are frequent neurologic complications. All strokes reported in HES patients had thrombo-embolic causes, usually cardiac embolism [4, 8, 9].

We here report a young patient with L-HES with a stroke due to a dissection of the posterior inferior cerebellar artery (PICA).

An 18-year-old female with hypereosinophilic syndrome (L-HES) presented with sudden numbness on the left side of her body and face. She also had left-sided hearing loss and mild headache. The diagnosis of L-HES was made at the age of 15 years, when she experienced swelling of her legs with erythema and itching, arthralgia, Raynaud phenomenon, hepato-megaly and splenomegaly with splenic infarctions. Laboratory analysis showed eosinophilia (3.87 × 10⁹/L, ref. range 0–0.5 × 10⁹/L) and a strongly activated (CD25+, HLADR+) clonal CD4 T-cell population immunophenotypically characterized as CD4+, CD2hi, CD5+, CD7±, TCRab+, Vbeta4dim. A biopsy revealed eosinophilic fasciitis. After the start of prednisone (60 mg/d) most complaints and the extreme eosinophilia disappeared. With the addition of interferon (IFN) α s.c. the prednisone could be tapered. At the time of presentation she used prednisone 5 mg/d and IFNα 4 × 10⁶ U/d. The number of aberrant T-cells was stable around 0.500–1.000 × 10⁹/L.

On admission her blood pressure was 135/85 mmHg. There was diminished pain perception, with intact vibration and proprioception, at the left side of body and face. There was no motor deficit and tendon reflexes were normal.

Tests for anti-nuclear antibodies, anti-neutrophil cytoplasmic antibodies, vasculitis and blood coagulation disorders revealed no abnormalities.

Brain MRI showed restricted diffusion in the right medulla compatible with infarction in the PICA territory. MRA showed a dissecting aneurysm at the origin of the right PICA (Fig. 1). Transthoracic echocardiogram and ECG were normal. An audiogram registered a left-sided perceptive hearing loss.

During the episode, eosinophil count was slightly elevated without other clinical signs of active HES. In view of the stable L-HES condition we did not change the prednisone/IFNα regimen to lower the eosinophil count further. We started aspirin 300 mg, dipyridamol at two times...
200 mg and simvastatin 40 mg for secondary prevention. The following days the hemisensory disturbances improved and had 1 week after initial presentation completely disappeared. Three months and 1 year later, brain MRI still showed the PICA aneurysm of 10 mm. In consultation with the neurosurgeon to discuss the possibilities for an operation, no options for intervention were found. The patient continued to recover without any residual symptoms.

We describe a patient with L-HES who presented with a PICA dissection leading to an ischaemic stroke. Spontaneous arterial dissection is a frequent cause of cerebral ischaemia in young patients, in contrast to strokes described in patients with HES that were all on the basis of cardioembolic stroke [8, 12].

The lymphocytic variant of HES results from increased secretion of eosinophilic cytokines by T lymphocytes. Though eosinophilia is usually less severe than in the myeloproliferative variant, it is harder to treat. The myeloproliferative variant results from clonal expansion of eosinophilic precursors due to various mutations, like the FIP1L1 gene translocation to platelet-derived growth factor receptor α (PDGFRα) gene [12].

Eosinophilia can damage the endocardium and myocardium, rendering the heart a potential source of emboli. Eosinophils contain multiple lytic substances. These lytic substances may result in destruction of the tunica media. Another explanation for vascular damage by eosinophils is the release of major basic protein, which has been isolated from the granules of eosinophils and has been shown to be cytotoxic [3]. The release of this protein may damage collagen, elastin and smooth muscle cells. This damage predisposes to aneurysm formation or spontaneous intimal dissection of coronary arteries and can lead to cardiac emboli or sudden cardiac death [10, 11]. A similar mechanism may play a role in intracranial dissection, as in our patient. Since intracranial dissection in L-HES has not previously been described an unfortunate co-incidence of the two cannot be ruled out.

Conservative therapy with antiplatelet agents is considered in non-ruptured intracranial dissections [14].
Follow up brain MRA should be considered in these patients to evaluate possible growth of the aneurysm [1]. Ruptured dissecting aneurysms usually need more aggressive, interventional treatment, such as endovascular coiling or stenting and has been performed with a good outcome in the majority of patients [5, 7]. In stroke patients with eosinophilia lowering the eosinophil count to prevent local thrombogenicity with steroids and nonspecific immune suppression is advised [12].

To our knowledge this is the first report of dissection of the PICA associated with L-HES. Arterial wall fragility may be a consequence of eosinophilic infiltration. Thus, besides cardiac embolism, cerebral artery dissection should also be considered as a cause of ischaemic stroke in HES patients.

Conflict of interest None declared.

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