HYPEREOSINOPHILIC SYNDROME – AN UNCOMMON CAUSE OF STROKE

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Abstract

Hypereosinophilic syndrome (HES) represents a rare cause for cerebral infarction and inflammatory neurological disorders. Complex mechanisms including a local hypercoagulability by eosinophilic granules and a direct damage to the endothelial cells, leading to alterations of the microcirculation seem to be involved.

Case Report

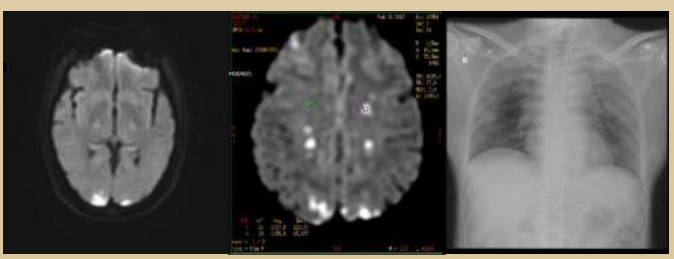
A 50 years old lady known case of bronchial asthma, hypertension brought to our hospital with left hemiparesis, ataxia and paresthesia after 24 hours of onset of symptoms. On examination. She was conscious, and well oriented and had a NIHSS of 5.

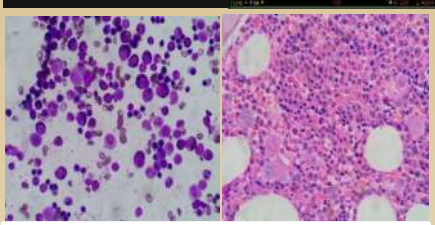
MRI showed multiple infarcts in bilateral frontoparietal lobes, occipital lobe with involvement of right thalamus and cerebellum. ECD showed increased IMT in left CCA. TCD was un remarkable.

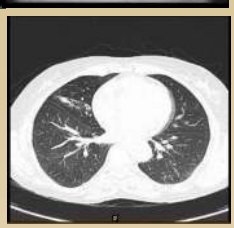
Her chest X-ray showed inhomogeneous opacities in bilateral upper zones, CT-Chest showed small volume prevascular, precarinal nodes, old chronic changes-Possibility of eosinophilic lung disease.

Cardiovascular system examination 2D Echo was normal, Raised Troponin-I of 6.7ng/ml. CT-Coronary Angiography showed normal vessels with papillary muscle hypertrophy. Holter Analysis was normal. Her USG Abdomen , LFT and renal parameters were normal.

Blood investigations: Hemoglobin 10.5gms%, PCV-30.80%, Total Leucocytes Count-26,610cells/cu mm, Eosinophils-41.00% peripheral smear showed Leukocytosis with eosinophilia, Absolute Eosinophil count(AEC)-10,307cells/mL, Smear study for microfilaria and malarial parasite were negative and stool microscopy for the parasite did not showed any ova, cysts. Bone marrow examination showed prominence of eosinophilic precursors. Serum Anti β2 glycoproteins (IgM-8.80 RU/mL, IgG- <1.80 RU/mL), were normal. ANA by IF, ANCA by IF, C-ANCA, P-ANCA were Negative, Various examinations to find cause were unremarkable. Patient was diagnosed with Hypereosinophilic syndrome(HES). Patient was put on corticosteroids.







BONE MARROW BIOPSY & ASPIRATION - Fig A
Leukocytosis with eosinophilia
Fig B. Large number of eosinophilic granules.

Discussion

HES is very rare and is characterized by the presence of Hypereosinophilia (absolute eosinophil count >1500cells/mL on 2 examinations >1 month apart, and/or findings of tissue Hypereosinophilia) with evidence of eosinophil-mediated target organ damage with all other potential causes of Hypereosinophilia excluded. Therefore, it is reasonable to perform a complete workup to exclude common causes of eosinophilia, such as parasitic infections, atopy/allergic disorders, malignancy and collagen vascular diseases. The presence of a known cytogenetic defect (such as FIP1L1-PDGRFa, PDGRFb and FGFR1 rearrangements) distinguishes the group of diseases as myeloid neoplasms associated with eosinophilia.

Complex mechanisms including a local hypercoagulability by eosinophilic granules as well as a direct damage to endothelial cells, leading to alterations of the microcirculation seem to be involved. The most common manifestations of Hypereosinophilic syndrome are pulmonary, skin, gastrointestinal, cardiac disorders and neurologic lesions .The following have been recorded so far: mononeuritis multiplex, sensory polyneuropathy, radiculopathy, myalgia, myositis and polymyositis, neuropathy, ataxia, paraplegia, ophthalmologic abnormalities, optic neuritis, hemiplegia-hemiparesis, spastic quadriplegia, seizures, meningitis, stroke[1],

Neurological manifestations are often observed in HES 65%, and comprise three types[2]. The first type has a cardiac origin, with thromboembolic causing multiple embolic strokes. The second type comprises one distinct encephalopathy and the third type comprises peripheral neuropathies, which occur in half of the HES patients .Cardio embolism (endomyocardial fibrosis) would coexist with impaired washout (perfusion disturbance due to high eosinophil count and/or eosinophil-derived substances), explaining the watershed characteristics of the infarcts[3].

Different treatments, including the use of corticosteroids and cytotoxic, have been investigated for HES with modest success .Today, with the development of new treatments as anti-interleukin-5, imatinib, alemtuzumab and other tyrosinokinase inhibitors, cases that are resistant to corticosteroid therapy should be submitted to other treatment modalities.

Conclusions

Eosinophilic patients can remain asymptomatic for decades, and the feature that defines the syndrome is organ involvement. Timely recognition of idiopathic Hypereosinophilic syndrome may enable aggressive treatment prior to widespread cardio embolism and degranulation that result in devastating cerebrovascular complications. This patient highlights the importance of evaluation and treatment of Hypereosinophilic syndrome presenting with ischemic stroke.

References

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