Hypereosinophilic Syndrome: A Catastrophic Pediatric Case

Adriana Bello [1*], Andres Casanova [1], Andrea Arias [1], Theodosia Kalfa [2], Steven Kussick [3]

[1] Centro Medico Docente la Trinidad, Caracas, [2] Cincinnati Children's Hospital, Cincinnati OH, [3] PhenoPath Laboratories, Seattle WA

Introduction



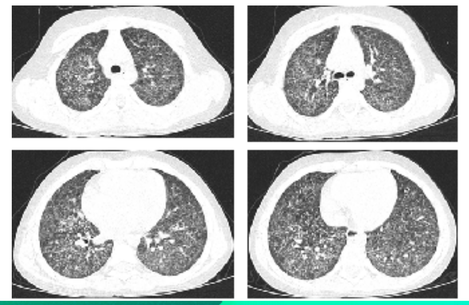
Hypereosinophilic syndrome (HES) is a persistent blood eosinophilia, absence of an underlying cause and characterized by organ damage.

We present a boy with extreme, refractory eosinophilia and end-organ complications.

The persistence, severity and outcome suggested an aggressive myeloid neoplasm.

Patient

A 6-year-old boy presented with fever and respiratory symptoms. A blood count revealed hyperleukocytosis of 200×10^9/L with 90% eosinophils. Fluorescent in-situ hybridization on the blood was negative for FIP1L1/PDGFRA, PDGFRB, FGFR1, and CBFB. Parasitic and infectious etiologies were ruled out.



Interventions

He was started on glucocorticoids and hydroxyurea as well as imatinib and a trial of antihelmintics. Signs of end-organ damage included 1) CNS hemiparesis, dysarthria, and MRI-documented small-vessel and white matter abnormalities; 2) myocarditis by cardiac ultrasound; and 3) respiratory distress with interstitial infiltrates.

Bone marrow evaluation ruled out leukemia, an abnormal T-cell population, and occult acute lymphoblastic leukemia. Marrow biopsy showed dysmorphic megakaryocytes. Cytogenetics on the marrow showed a normal karyotype, and myeloid-directed next generation sequencing on the blood was negative for mutations.

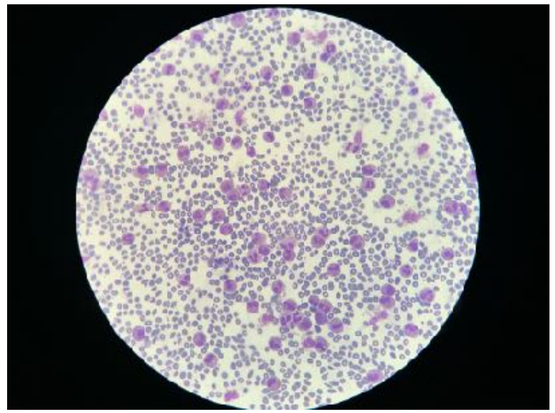
Lung biopsy showed increased eosinophils within vessels, pleural fibrosis and no interstitial fibrosis or vasculitis.

Interferon alfa was given 3 times weekly with no response. The 4 drugs were well tolerated.

He was followed over 9 months, requiring antibiotics, anticoagulation, and oxygen support. Eosinophilia remained high. The patient expired from respiratory failure.

Conclusions

HES may be life-threatening in the pediatric population. Diagnosis and therapy can be challenging.



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