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Original Research

# A Myeloproliferative Syndrome Revealed by An Eosinophilic Lung

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**Summary:**

Hyper eosinophilic syndrome (HES) is defined by eosinophilia greater than 1.5 G/L persisting for more than six months and associated with specific visceral damage (cardiac, neurological, pulmonary, etc.), after elimination of the "classic" causes of hyper eosinophilia.

We report the case of a 25-year-old man who presented to the emergency room for acute-onset febrile dyspnea, transferred to the pulmonology department for suspected Covid, an FNS performed as part of a routine check-up. reveals a leukocytosis of more than 65 G/L including 80% eosinophils, an etiological search launched, first eliminated the secondary causes and firstly the allergies, led to a cytogenetic analysis which came back positive in favor of an eosinophilic myeloproliferative syndrome with the demonstration of a FLIPI-PDGFRA transcript, he benefited from targeted therapy with a total regression of the symptoms followed by recurrent cytogenetics.

**Key words**: Myeloproliferative Syndrome, Hyper Eosinophilia, Dyspnea, Cytogenetics.



## Introduction:

Hyper eosinophilic syndrome (HES) is defined by eosinophilia greater than 1.5 G/L persisting for more than six months and associated with specific visceral involvement (pulmonary, neurological, cardiac, etc.), after elimination of the "classic" causes of hyper eosinophilia[1]. A myeloproliferative variant of HES is associated with the fusion of the FIP1L1 and PGDFRα genes, leading to the constitutive activation of a receptor with tyrosine kinase activity. We report a case, revealed unexpectedly by pulmonary involvement (febrile dyspnea)[2]

## Observation:

A 25-year-old man with no particular history, no allergies and no cardiovascular risk factors was hospitalized in the pulmonology department after referral to medical emergencies for sudden onset of febrile dyspnea associated with a fever.

General examination: general condition preserved, asthenic, nocturnal fever, dyspnea, precordialgia, splenomegaly stage IV The ECG is unremarkable, a thoracoabdominopelvic CT showed homogeneous splenomegaly, with pulmonary parenchymal involvement in centrilobular ground glass, predominantly apical, with areas of

geographical somatic, vertebral, dorsolumbar and pelvic bone condensation (Fig 1).

The echocardiogram: finds a thickened mitral valve with cord rupture, a moderate mitral leakgiving moderate grade II mitral regurgitation (Fig 3)

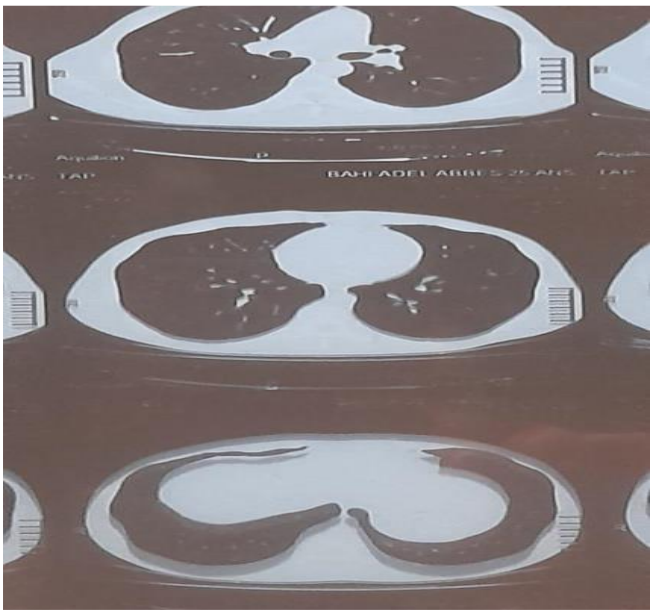

**Figure 1: pulmonary parenchymal involvement in centrilobular ground glass, predominantly apical**

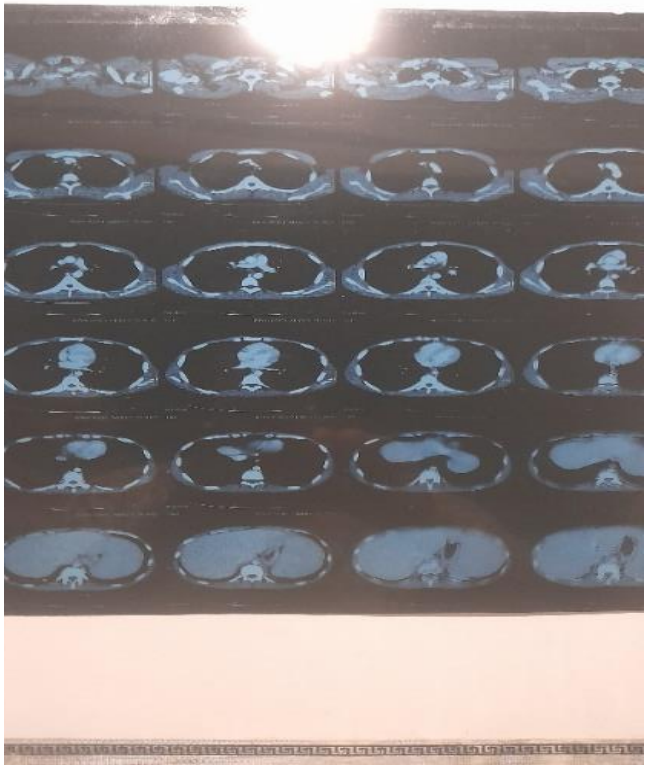

**Fig 2 presence of a homogeneous splenomegaly**

Given the pandemic context, a serology as well as a Covid PCR performed came back negative, 3 blood cultures taken at the time of the fever peak came back sterile.

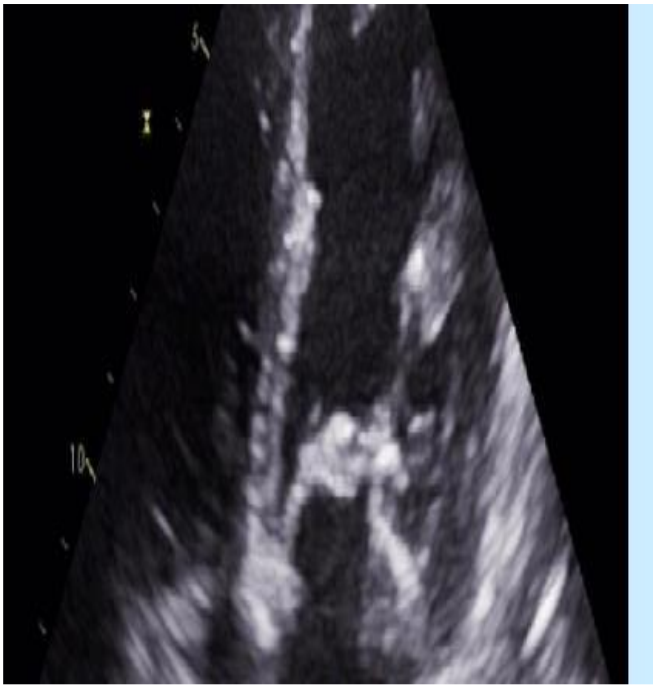

**Figure 3: Thickened mitral valve with chordae rupture, moderate mitral regurgitation grade II**

An FNS finds moderate anemia at 9 g/dl normochromic normocytic, aregenerative, moderate thrombocytopenia at 124 G/L, leukocytosis at 65,000/mm3 including 81% polynuclear eosinophils (52,650 elements/mm3), without myelemia (Fig 4)

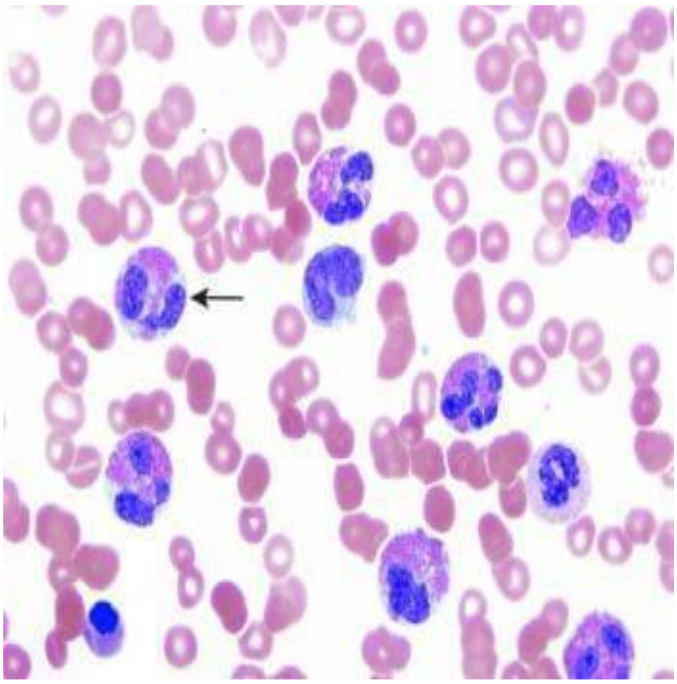

**Figure 4: in the peripheral blood smear: presence of numerous eosinophils within red blood cells.**

On myelogram: the smear is rich with eosinophilic hyperplasia at all stages of maturation.

A coproparasitology of the stools came back negative, and given the negativity of the corticosteroid test, a cytogenetic study was launched in search of the FLIP1 PDGFR mutation and which revealed the FLIP1 PDGFRA transcript in 88% of the cells with a rearrangement. from the 4q12 locus

It is therefore an eosinophilic myeloproliferative syndrome.

The patient was placed on the 1st generation tyrosine kinase inhibitor (TKI) Imatib at a dose of 100 mg/day with prophylactic anticoagulation.

The evolution was favorable with disappearance of the splenomegaly and normalization of the FNS after 20 days and above all a total disappearance of the cytogenetic anomaly after 6 months of treatment

**Discussion:**

The diagnosis of SHE must be considered in the face of any blood HE ≥ 1500/mm3 and/or tissue associated with organ dysfunction. The clinical manifestations are diverse but dermatological, respiratory and digestive disorders are the most common[3].

frequent. In fact, the clinical respiratory manifestations are cough but especially dyspnea which gradually worsens. Radiologically, these are often bilateral alveolar opacities, highlighted by chest CT in the form of a ground glass appearance with multifocal alveolar condensations [2 cardiac and thrombotic involvement are rarer but more serious[4]

-Any patient with an HE >1500/mm3 must benefit from an etiological assessment and the impact of the HE[1]. Although there are some preferential symptoms (examples: cardiac involvement during clonal SHE, angioedema and joint manifestations during lymphoid SHE), all clinical manifestations of SHE are possible, whatever the mechanism. pathophysiology underlying, HE.

-As the eosinophilic tissue infiltrate is an integral part of the definition of SHE, histological or cytological proof (bronchoalveolar washing for example) is always desirable[5]. However, if an eosinophilic tissue infiltrate cannot be detected (too invasive biopsy, corticosteroid therapy already initiated, etc.), the diagnosis of SHE can also be retained by default when all the following criteria are present: blood HE, organ damage compatible with HE (notably with the absence of differential diagnosis) and parallelism of evolution between organ damage and blood HE[6,7]

Summary of explorations in search of clonal eosinophilia

Hypereosinophilia > 1500/mm3

persistent ≥ 1 month and/or requiring treatment

Absence of allergic, parasitic or autoimmune cause

or obvious neoplastic

Criteria justifying the search for a clonal EO:

- Cortico-resistance
- Splenomegaly
- Tryptase ↑
- Vitamin B12 ↑
- Other unexplained abnormality(s) of the blood count
- Male sex (in case of unknown or non-evaluable cortico-sensitivity[8].

In our observation, the patient therefore had the criteria for clonal hyper eosinophilia in the face of splenomegaly, the absence of cause and especially resistance to corticosteroids.

Certain serious clinical manifestations, particularly cardiac (myocarditis, intracavitary thromboses with peripheral or cerebral embolic manifestations, coronary spasm), acute respiratory distress (severe acute asthma, hypoxemic pneumonia), or thrombotic (venous or arterial) require the implementation of an urgent treatment combining different classes of anti-eosinophilic treatment used sequentially, without waiting for the results of the etiological assessment of HE, which justifies the use of anticoagulants in our patient[9,10].

**Conclusion:**

FLIP1L1-PDGFRA chronic eosinophilic leukemia can be serious and fatal if there is a diagnostic delay

and the nature of the organ is damaged by the eosinophils. We must investigate patients presenting clinical manifestations related to hyper eosinophilic syndrome, looking for signs in favor of clonality of hematopoiesis, and a search for a T lymphocyte population for urgent therapeutic decisions.

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