soho2023
ELEVENTH ANNUAL MEETING

HYPER EOSINOPHILIA WITH FIP1L1-PDGFRA GEN FUSION PRESENTING AS A T CELL LYMPHOBLASTIC LYMPHOMA

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INTRODUCTION

Lymphoid neoplasms with PDGFRA rearrangements are a rare form of malignancies for which a standard therapeutic approach has not been published yet, making valuable to share the experience of different medical centers in terms of treatment..

AIM

To describe the clinical presentation, diagnosis, and use of Imatinib in a patient with hyper eosinophilia with PDGFRA rearrangement presenting as a T-Cell Lymphoblastic Lymphoma.

PATIENT

A 44-year-old previously healthy male, who presents with a two-month history of weight loss, fever, and painless lumps in the neck. His initial blood tests reveal a leukemoid reaction with severe eosinophilia (#19.4 x103/L). On examination, he had firm, fixed and painless bilateral lymph nodes in the neck of variable dimensions.

RESULTS

An excisional biopsy of a left inguinal node was performed, which was positive for a T cell lymphoblastic lymphoma with CD3+/CD5+ immunophenotype. Bone marrow biopsy reported a hypercellular marrow with eosinophilia, increased megakaryocytes with normal morphology, and increased mastocytes with fusiform morphology. C-Kit mutation is currently not tested in the country. The karyotype was normal (46XY), and a Fluorescence In situ Hybridization (FISH) showed FIP1LI/CHIC2/PDGFR with interstitial deletion detected in 100% of analyzed cells. The patient was diagnosed with a lymphoid neoplasm (T-Lymphoblastic Lymphoma) with PDGFRA rearrangement and was started on an intensive chemotherapy regimen based on CALBG 10403 protocol and a tyrosine kinase inhibitor (TKI) imatinib at a dose of 400mg once daily.

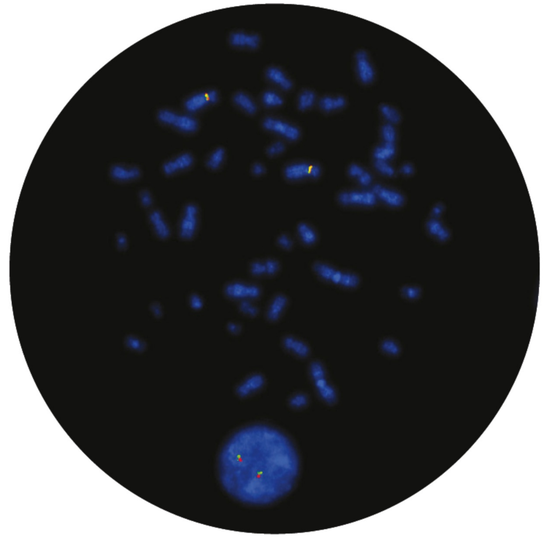


Image with illustrative purpose

CONCLUSIONS

The use of TKI monotherapy has been reported as safe in previous reports in these patients. However, follow up is needed to determine the risk of relapse and estimate the overall survival and progression free survival with the use of that strategy in comparison with TKI combined with traditional chemotherapy protocols.

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