ANSWER Q2-2017

Diagnosis: Myeloid neoplasm with eosinophilia and PDGFRA rearrangement

Myeloid and lymphoid neoplasms with PDGFRA rearrangement are a rare disease. The most common genetic abnormality is *FIP1L1-PDGFRA* resulted from a cryptic deletion at 4q12. It usually presents as chronic eosinophilic leukemia. Less common presentations include acute myeloid leukemia, T-lymphoblastic leukemia (T-LBL), or AML & T-LBL simultaneously. Pre-existing eosinophilia was documented for majority of the cases. The most serious clinical findings relate to endomyocardial fibrosis.

Since the majority of cases do not have an increase of blasts or any abnormality on conventional cytogenetics, the detection of the *FIP1L1-PDGFRA* fusion is usually needed for a definitive diagnosis.

FIP1L1-PDGFRA-associated CEL is very responsive to imatinib, 100-fold more sensitive than CML. Majority of the patients could achieve molecular remission by imatinib. Prognosis appears favorable if cardiac damage has not already occurred. Resistance to imatinib is extremely rare.