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Hematologists 2019- Chronic Neutrophilic Leukemia: A Rare and Difficult Diagnosis of Exclusion- James Ziai- Yale University School of Medicine

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Abstract

Chronic neutrophilic leukemia (CNL) is very rare myeloproliferative disorder that presents diagnostic challenges for both pathologists and treating clinicians. Because this disease entity is extremely rare, and because it's typically a diagnosis of exclusion, its important for pathologists and hematologists to be accustomed to CNL when approaching the patient with a myeloproliferative clinical picture. Thus, the objectives of this report are: 1) to detail the clinical case of a 59 year old male veteran with initial presentation of hyperleukocytosis, 2) to review the differential diagnosis of a granulocytic myeloproliferative presentation and demonstrate the laboratory and clinical criteria utilized to establish a diagnosis of CNL during this case, and 3) to briefly review the current literature on the diagnosis and treatment of CNL

Keywords: Chronic neutrophilic leukemia; A typical chronic myelogenous leukemia; Myeloproliferative diseases

Introduction:

Chronic neutrophilic leukemia (CNL) could be a rare myeloproliferative neoplasm characterized primarily by leukocytosis, but often lacking distinct clinical, laboratory, and molecular features. Assessing the patient with an atypical myeloproliferative picture and correctly making the diagnosis of CNL can be challenging for pathologists and clinicians alike. The aims of this report are to detail the clinical case of a 59-year-old male veteran with initial presentation of hyperleukocytosis so as to demonstrate the laboratory and clinical criteria utilized to establish a diagnosis of CNL. We also briefly review the current literature on the diagnosis and treatment of CNL.

Case Report:

A 59-year-old male veteran presented to our facility (VA Connecticut Healthcare System, West Haven, CT, USA) with chief complaints of shortness of breath (SOB) and fatigue of approximately two weeks duration. The patient had been diagnosed with hypertension in the past and was taking amlodipine, labetalol, and aspirin. He had no relevant case history and was a non-smoker. On review of systems the patient denied wheezing, cough, orthopnea, syncope, chest pain, fever or any particular exacerbating or remitting factors for his current condition. Pertinent physical exam findings included a lethargic appearance, mild splenomegaly, and a 2x2x1cm fixed painless, hard mass on the right anterior mandible.

Imaging studies were also performed to investigate for the possibility of other malignant or reactive causes for the patient's leukocytosis. Chest x-rays, performed on admission and on several days during the course of the patient's hospital stay, showed some evidence of pulmonary congestion consistent with mild congestive heart failure, but without evidence of an infectious infiltrate or malignant process. A CT scan of the thorax performed on day 4 of admission showed mild, "non-specific" mediastinal adenopathy, without other significant findings. Retroperitoneal ultrasound demonstrated echogenic kidneys and a 4 cm hepatic cyst cluster, but no other significant abnormal findings. In the absence of any evidence for a reactive cause of the leukocytosis, a bone marrow aspirate and biopsy were performed. The bone marrow core biopsy was markedly hypercellular for age with a cellularity estimated at 90% (Figure 1a). Myeloid maturation was sequential with a predominance of mature granulocytes, mainly localized in interstitial areas (Figure 1b and 1c). proliferation but otherwise appeared normal and megakaryocytes were mildly increased with normal morphology .There was no relative increase in myeloblasts, eosinophils, basophils, or mast cells. Moderate focal reticulin fibrosis was noted in paratrabecular regions, but didnt reach to cortical areas. The bone marrow aspirate reiterated the core biopsy findings. There was toxic granulation noted on neutrophils and a few hypersegmentation noted on peripheral smear.

Discussion:

Myeloproliferative neoplasms are clonal hematopoietic stem cell disorders characterized by proliferation of one or more elements of the neoplasms associated with abnormalities of PDGFRα and PDGFRβ were also essentially ruled out by negative molecular testing. Second, neutrophil precursors make up less than 10% of circulating leukocytes. Finally, the patient's chronic, relatively indolent clinical course is consistent with most reports of CNL as opposed to atypical CML which often follows a rapid and aggressive downward course. In summary, this is a case of a 59-year-old man presenting with SOB and hyperleukocytosis ultimately diagnosed as CNL. This case demonstrates the issue in firmly establishing a diagnosis of CNL and the way clinical and laboratory criteria may be utilized to discriminate between similar, but distinct, disease entities. In this case, a diagnosis of CNL not only influenced the initial treatment choice, but also provided important prognostic implications. A lack of reproducible molecular abnormalities remains a major hurdle

in clearly identifying CNL cases. Future work in characterizing the molecular profiles of this and other rare myeloproliferative neoplasms would aid not only in their classification and diagnosis, but could also yield novel treatment options.

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