Case Report

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A Rare Cause of Roth Spots and Pyoderma Gangrenosum: Chronic Myeloid Leukemia

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1. Abstract

Chronic myeloid leukemia is a rare malignant hematopathy that primarily affects young adults between the ages of 30 and 50, with a preference for males. Typically, its onset is insidious, often discovered incidentally during routine clinical examinations or through blood tests. Occasionally, it presents with nonspecific symptoms or left upper abdominal discomfort, leading to the identification of splenomegaly. Rarely, it may manifest as hemorrhagic complications or hyperuricemia-related symptoms. In sub-Saharan Africa, diagnosis often occurs late due to pronounced symptoms. We report the case of a 21-year-old woman in whom chronic myeloid leukemia was unexpectedly discovered based on atypical clinical manifestations.

2. Keywords:
Chronic Myeloid Leukemia, Roth Spots, Pyoderma Gangrenosum

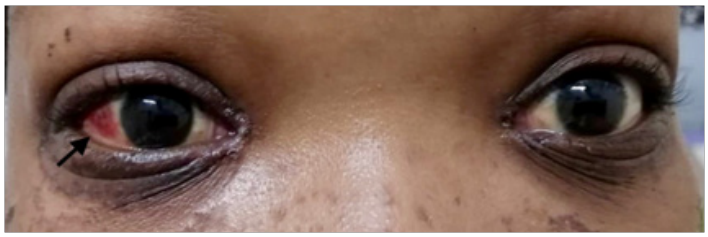
3. Introduction

Chronic myeloid leukemia (CML) is characterized by the proliferation of the granulocytic lineage and the presence of a clonal-acquired cytogenetic abnormality: the Philadelphia chromosome (Ph). This chromosome results from a reciprocal translocation between chromosomes 9 and 22, leading to the formation of a fusion oncogenic protein called Bcr-Abl, which exhibits dysregulated tyrosine kinase activity essential for leukemogenesis [1]. One of the peculiarities of this disease in sub-Saharan Africa is its aggressive clinical presentation at the time of diagnosis, largely due to delayed diagnosis [2-3]. Against this backdrop, we present the case of a patient whose discovery of CML occurred unexpectedly, with atypical ocular and cutaneous manifestations.

4. Observation

Madame A. B., 21 years old, nulligravida, with no significant medical history, was admitted to the ophthalmology department due to a sudden bilateral decrease in visual acuity (BAV). This was accompanied by diffuse subconjunctival hemorrhage (Image 1)and neurosensory symptoms such as headaches, ear buzzing, and dizziness. Her visual acuity was estimated at 3/10 in the right eye and 4/10 in the left eye. The anterior segment examination was normal.

Image 1: Subconjunctiva lhemorrhage



Upon fundus examination, bilateral stage 2 papillary edema, diffuse retinal hemorrhages centered around Roth spots, and severe retinal vasculitis characterized by venous dilation and tortuosity were observed. Additionally, there were scattered areas of dry exudates throughout the retina. These findings were confirmed by retinal angiography. Orbital-cerebral MRI results were normal. Furthermore, the clinical examination revealed an ulcerated, budding, inflammatory, painful lesion with a purulent and bleeding base, and irregular borders. This lesion was located between the occipital region and the nape of the neck. Initially considered an abscess, it rapidly evolved despite nonspecific antibiotic therapy and debridement (Image 2a & b).Swabbing of the wound yielded numerous neutrophils and isolated Staphylococcus aureus. The appearance strongly suggested an ulcerated, superinfected form of pyoderma gangrenosum (PG). The otherwise clinical examination revealed Hackett grade IV splenomegaly without hepatomegaly or lymphadenopathy. The patient's general condition was altered (WHO performance status 2). Abdominal ultrasound and computed tomography confirmed the splenomegaly

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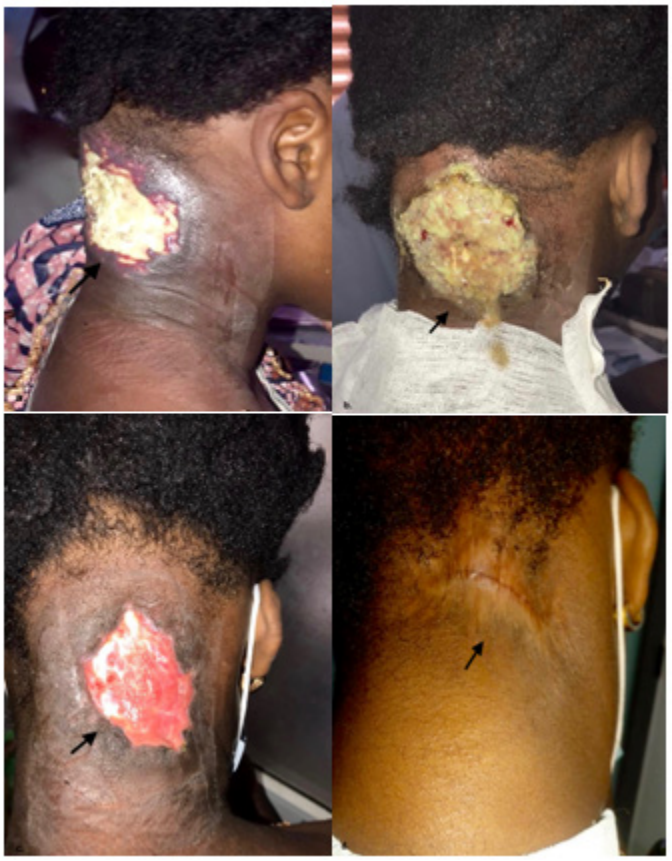
without signs of portal hypertension.

Image 2 : Pyodermagangrenosum

a and b. Painful, ulcerated, budding lesion with a purulent, bleeding base, with clear, purplish, hypertrophic irregular borders, located between the occipital region and the nape of the neck. c. Ulcer with a red, non-budding base with a disinfiltrated irregular border.
d. Healed lesion



The blood count showed significant hyperleukocytosis at 673 G/L, with neutrophils at 313 G/L, eosinophilia, and basophilia at 13 G/L. Additionally, there was a normocytic normochromic anemia at 5.8 g/dL and a normal platelet count at 382 G/L. The blood smear revealed 43% myelocytes, including 3% blasts. The bone marrow aspirate demonstrated an overrepresented granulocytic lineage, well-balanced without maturation arrest, and 1% myeloblasts. The diagnosis of chronic myeloid leukemia (CML) was asserted by the presence of the Philadelphia chromosome (Ph) without additional cytogenetic abnormalities in the bone marrow karyotype and the MBCR-ABL transcript (31.65% ratio) on Real-time reverse transcriptase polymerase chain reaction (RQ-PCR). Overall, CML in its chronic form was established in this patient. No underlying conditions were found (negative serologies for hepatitis B, C, and HIV; negative antinuclear factors; no cardiovascular risk factors). Liver and kidney function, hemostasis, lipid levels, and blood glucose were all within normal limits. She received imatinib mesylate at a daily dose of 400 mg, along with preventive treatment for tumor lysis syndrome. Her progress was marked by significant improvement in visual acuity (10/10 in both eyes), resolution of vertigo, normalization of leukocyte count after one month of treatment, and complete healing of the lesion after three months of treatment (Image 2c & d).

5. Discussion

The prevalence of ocular involvement in leukemia can vary between 9% and 90% as per studies [4]. This involvement primarily occurs through two mechanisms: it can be primary, resulting from direct infiltration of neoplastic cells, or secondary, related to abnormalities in blood coagulation, treatments used, immunosuppression, and opportunistic infections [5]. The term “leukemic retinopathy” describes retinal manifestations associated with anemia, thrombocytopenia, and blood hyperviscosity, rather than direct leukemic infiltration. Some hemorrhages may appear as white-centered lesions containing leukemic cells, platelet aggregates, and fibrin or septic emboli, known as “Roth spots”. Blood hyperviscosity due to major leukocytosis can manifest as bilateral stasis retinopathy, characterized by dilation and tortuosity of all retinal veins, mimicking central vein occlusion without obvious blockage, also referred to as “paraproteinamucus fundus”. This condition is attributable to circulatory slowing, predominantly affecting the venous sector, and true venous occlusion may occur [6]. Our patient presented with severe leukocytosis at the time of diagnosis and no hemostatic disorders. Her retinal involvement is likely primary.

Pyoderma gangrenosum (PG) is a rare, non-infectious neutrophilic dermatosis that is often underrecognized. It typically presents as painful, rapidly evolving inflammatory skin ulcers, contrasting with the absence of inflammatory lymph nodes. Even minor local trauma can reveal or worsen the ulceration, a phenomenon known as the Koebner phenomenon. PG is frequently associated with neoplasms, inflammatory gastrointestinal, rheumatological, and/or hematological conditions. Diagnosis is often delayed after multiple therapeutic failures (antibiotics, surgical care). There are several forms of PG: ulcerative, pustular, bullous, and superficial or vegetating granulomatous forms [7-8]. The causes and mechanisms of pyoderma gangrenosum (PG) remain poorly understood, but they appear to be associated with immune system dysfunction and inflammation. PG, currently classified as a neutrophilic dermatosis, can sometimes be confused with or associated with other neutrophilic skin conditions such as Sweet syndrome (acute febrile neutrophilic dermatosis), subcorneal pustulosis, or erythema elevatum diutinium [9]. When considering differential diagnoses for ulcerated skin lesions, vasculitis, arterial or venous insufficiencies, infectious diseases, and malignant hematological conditions should be taken into account [10]. PG remains a clinical diagnosis of exclusion. Although histological features are often nonspecific, they help rule out other potential causes [11].

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6. Conclusion

Diagnosis delay and wandering often contribute to the clinical-biological complexity observed in patients with chronic myeloid leukemia (CML) in sub-Saharan Africa. The causes of this delay are multifaceted and include economic factors (limited access to healthcare, lack of universal coverage), logistical challenges (cost of diagnostic tests, inadequate technical facilities), and cultural factors, with traditional medicine playing a significant role. Reducing diagnostic delays and enhancing technical facilities would contribute to optimal patient management

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