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Case Report

A Rare Presentation of Myeloid Sarcoma as Supraclavicular Mass: A Case Report in Chronic Myeloid Leukemia

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Abstract

Myeloblasts or immature myeloid cells form myeloid sarcoma, an extramedullary myeloid tumor which commonly affects sites like skin, soft tissues, bone, peritoneum, and lymph nodes. Myeloid sarcoma typically occurs during accelerated and blast phase of chronic myelogenous leukemia. It is a very uncommon presentation of CML in its early medullary chronic phase. This is a rare case of CML that presented with an ulcerative lesion on the left clavicular area and a soft swelling in the anterior cervical region above the clavicle, which are aggressive and rare extramedullary manifestations of chronic myeloid leukemia. Notable finding include TLC of 418.92 X 10E9/L, peripheral blood film showed anisocytosis, poikilocytosis, polychromasia, macrocytosis and nucleated RBC, karyotyping for Philadelphia Chromosome was positive. Overall findings are consistent with myeloproliferative neoplasm (chronic myeloid leukemia in chronic phase). The patient started on TKI therapy and showed improvement initially. Despite improvement, patient's relapse suggests potential resistance, emphasizing the need for vigilant follow-up and consideration of alternative treatment strategies.

Keywords: *Chronic myeloid leukemia, Myeloid Sarcoma, Philadelphia chromosome*

Introduction

Myeloblasts or immature myeloid cells form myeloid sarcoma, an extramedullary myeloid tumor. It is also called Chloromas and granulocytic sarcomas. The most frequently affected sites are the skin, soft tissues, bone, peritoneum, and lymph nodes or any organs [1]. Myeloid sarcoma seldom develops in the Chronic Phase. Still, it typically does in bone marrow and peripheral blood during the accelerated phase or blast phase when chronic myelogenous leukemia (CML) is present. Also, myeloid sarcoma is a very uncommon presentation of CML in its early medullary chronic phase [2].

Myeloid sarcoma involving the skin is frequently linked to trisomy 8 in chronic myelogenous leukemia (CML). There seems to be a higher frequency of skin lesions in individuals with trisomy 8, yet the significance of this observation in cutaneous myeloid sarcoma is still unknown.

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Although they are uncommon, BCR-ABL1 positive myeloid sarcomas have been seen without systemic disease, and myeloid sarcomas in CML can initiate the blast phase of the disease. Given that chromosomal gains, like those involving chromosome 8, are frequently seen in both bone marrow and extramedullary sites, these lesions may be a reflection of clonal evolution [3].

Here we have described a rare case of CML that presented with an ulcerative lesion on the left clavicular area and a soft swelling in the anterior cervical region above the clavicle, which are aggressive and rare extramedullary manifestations of chronic myeloid leukemia (CML). It is identified to be myeloid sarcoma, showing that the disease progresses to the blast crisis stage. This case highlights the significance of identifying extramedullary lesion in advanced CML by describing this unusual appearance.

Case Presentation

A 40 y/o male with no comorbidities presented with c/o undocumented weight loss for 3 months and general weakness for one month. General physical examination and systemic examination were clinically insignificant.The complete blood picture (CBC) revealed marked leukocytosis with TLC of 418.92 X 10E9/L. The rest of the CBC was unremarkable. Due to the high suspicion of a WBC neoplasm, a peripheral blood film was ordered which showed anisocytosis, poikilocytosis, polychromasia, macrocytosis and nucleated RBCs. Promyelocytes, myelocytes and metamyelocytes were also observed. Overall findings are consistent with myelopro-liferative neoplasm (chronic myeloid leukemia in chronic phase). To confirm the findings of the CBC, bone marrow (BM) aspiration and trephined section conducted by the medical team along with results of reticulin stain, as well as the karyotyping forPhiladelphia Chromosome which came out positive (figure 1).

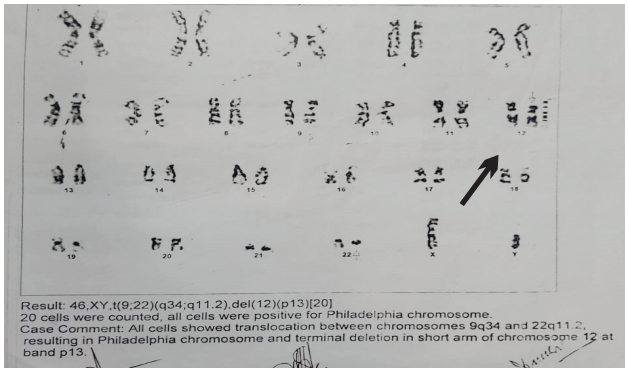

Figure 1: Karyotyping showing positive for Philadelphia Chromosome

These results confirmed the dx. Following the first presentation and receiving the dx, the patient was started on TKI therapy and the agent selected was Imatinib, 100mg 2 BD PO for 1 month. As a result, the patient showed improvement, and his TLC got normalized. The Doctors suggested continuing the Rx until advised otherwise. After that, the patient lost follow-up and reported a few months later w/c/o a left supraclavicular mass (figure 2).

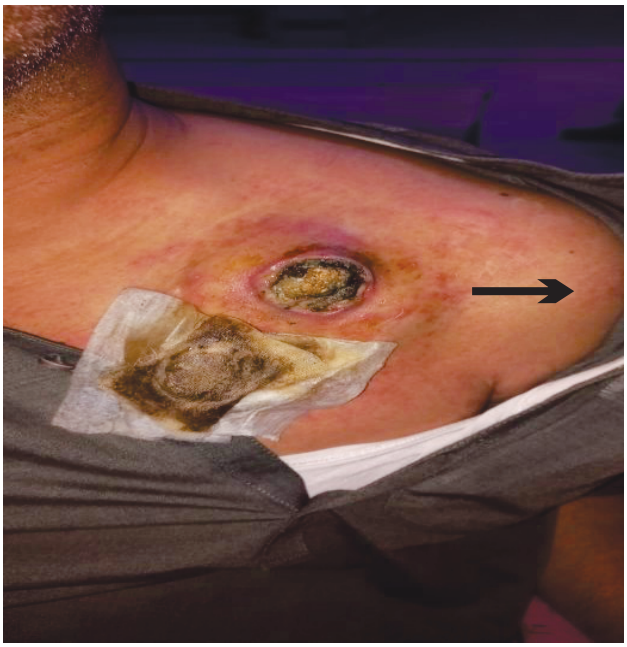

Figure 2: Supraclavicular mass above left clavicle

An Ultrasound of the mass was performed, which suggested enlarged lymph nodes with fatty hilum. An FNAC was performed that led to the findings suggestive of Non-Hodgkin Lymphoma (NHL). The incisional biopsy concluded with malignancy as well. To get to the root of it, immunohistochemistry was performed, which was positive for LCA, CD99, CD117, and MPA while TDT, CD20, CD3 were found to be negative. This confirmed the dx of Myeloid Sarcoma.

Bone marrow and trephine section biopsy were carried out once again which revealed cellularity 50%, and the rest 50% is replaced by atypical/blast cells. Immunohistochemical stains of the bone marrow were positive for MPO, weak positive for CD117, while negative for CD10. This was consistent with the dx of Acute Myeloid Leukemia. Overall findings favor Acute Myeloid Leukemia (AML). Having lost the follow up for months, and reporting again for the second time, he was again put on glynib (Imatinib).

Discussion

Myeloid Sarcoma (MS), also referred to as granulocytic sarcoma, is an infrequent extramedullary malignancy characterized by the proliferation of immature myeloid cells. It is often associated with hematologic conditions such as chronic myeloid leukemia (CML) or acute myeloid leukemia (AML) [4]. Myeloid sarcoma (MS) can present across a wide age range, with median ages varying depending on the study population. For instance, a study focusing on head and neck region MS reported a median age of 61 years (range: 17–85 years) [5]. Our patient, a 40-year-old, aligns with this age distribution. However, the clinical variability of MS frequently leads to misdiagnosis[5]. Historical reviews indicate that MS is often mistaken for conditions such as non-Hodgkin lymphoma, Ewing sarcoma, and undifferentiated carcinoma [6-7]. The definitive diagnosis of MS relies on histopathological examination and immunohistochemical analysis. Common markers, including MPO, CD99, CD117, and leukocyte common antigen (LCA), were all positive in this case. Notably, CD43, while typically associated with T-cell neoplasms, suggests a myeloid origin in the absence of CD3, as observed in our patient[8]. Imaging studies such as MRI and CT scans play crucial roles, but PET scans are particularly advantageous for early detection and tumor burden evaluation [9].

This report highlights a unique case involving a known CML patient with a 2.5-month history of supraclavicular swelling. To our knowledge, this is the first such presentation documented among the 51 cases previously indexed on PubMed. Common anatomical sites for MS after being diagnosed with CML include lymph nodes, soft tissues, and skin [10]. Unlike the systemic blast phase typically accompanying MS in CML, our case demonstrated isolated extramedullary progression without systemic involvement. This atypical presentation highlights a potentially distinct clinical subset of MS [10].

The proliferative index, as indicated by Ki67, is a critical diagnostic and prognostic marker. A high Ki67 index (>30%) is generally associated with aggressive disease. In our case, the Ki67 index ranged between 40–50%, consistent with aggressive features and supporting the diagnosis of MS [11-12].

Treatment Modalities

The management of MS requires a multimodal approach tailored to individual patient factors, including age, comorbidities, and disease burden. Despite receiving imatinib for two years, our patient developed MS, raising concerns about emerging resistance. In such scenarios, transitioning to second- or third-generation tyrosine kinase inhibitors (TKIs), potentially in combination with systemic chemotherapy, is a common strategy [10].

Additionally, radiotherapy has been explored as an adjunctive treatment in certain cases, as documented in recent studies [13]. MS associated with CML generally portends a poor prognosis due to its aggressive nature and systemic progression. However, the absence of systemic involvement in this patient underscores a rare clinical presentation, suggesting the need for further studies to delineate the prognostic and therapeutic implications of such cases.

Conclusion

This case underscores the aggressive nature of myeloid sarcoma as a rare extramedullary manifestation of CML, leading to transformation into AML. The diagnostic challenge posed by its mimicry of lymphoma highlights the importance of histopathological and immunohistochemical confirmation. Despite the initial response to TKI therapy, the patient's relapse suggests potential resistance, emphasizing the need for vigilant follow-up and consideration of alternative treatment strategies.

Patient Consent: Informed consent was obtained

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Conflict of interest: None

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