

SHORT COMMUNICATIONS

Myeloid sarcoma – A Case Report

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INTRODUCTION

The diagnosis of myeloid sarcoma is challenging due to its rarity and heterogeneity of presentation. It can affect any organ and present at any age, although one of the more commonly involved sites is the skin. We present a unique case of isolated myeloid sarcoma that presented as generalized erythematous nodules involving the face, neck, chest, abdomen, back, and extremities.

CASE REPORT

A 76-year-old male with a four-year history of idiopathic thrombocytopenia on no medications presented with acute onset generalized erythematous nodules worsening over 7-weeks. Review of symptoms was remarkable for myalgias (shoulders/thighs), unintentional 9lbs weight loss, and lower lip numbness. Physical examination revealed innumerable plum-colored 5-20mm nodules involving the face, neck, chest, abdomen, back, and extremities (Figure 1). No palpable lymphadenopathy was appreciated. A skin biopsy was

performed from a 1cm nodule on the left arm (Figure 2A).

Histologic sections revealed a dense dermal infiltrate of medium-sized mononuclear cells with finely dispersed chromatin, small nucleoli, increased nuclear:cytoplasmic ratio, and irregular nuclear contours (Figure 2A). Staining was positive for CD43 (Figure 2B), lysozyme (muramidase) (Figure 2C), CD33, CD56, and CD45. Staining was negative for CD3, CD4, CD20, CD34, CD68 (PG-M1), CD117, CD 123, CD163, myeloperoxidase, TdT, and PAX-5.

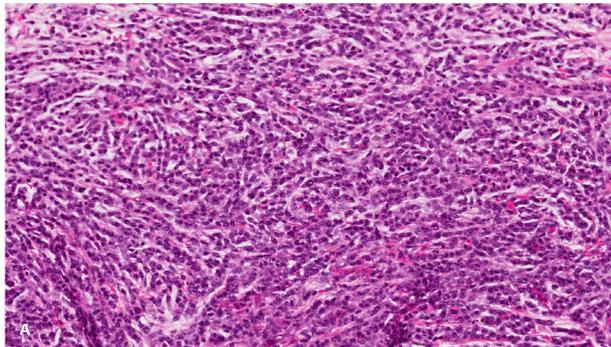
Bone marrow biopsy demonstrated hypercellular bone marrow with granulocytic/megakaryocytic hyperplasia, and dysgranulopoiesis. Peripheral blood smear

Figure 1: 76 year old male presenting with an eruption of innumerable plum-colored nodules.



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Figure 2: Histologic sections of skin biopsy taken from a 1cm nodule on the left arm: A, stained with hematoxylin and eosin. B, stained positive for CD43. C, stained positive for lysozyme.



showed left-shifted neutrophilia, with no morphological evidence of acute leukemia. Flow cytometry on peripheral blood was unremarkable. qRT-PCR studies performed on bone marrow aspirate ruled out BCR-ABL positive chronic myelogenous leukemia (CML). Testicular ultrasound revealed leukemic infiltrates.

The histopathologic and bone marrow biopsy findings were consistent with a diagnosis of myeloid sarcoma (MS).

DISCUSSION

The differential diagnosis for cutaneous MS includes B-cell lymphoma, acute myeloid leukemia (AML), cutaneous T-cell lymphoma, Ewing sarcoma, melanoma, round blue cell tumors, and poorly differentiated carcinoma.

MS is an extramedullary tumor of myeloblasts most often associated with AML (incidence 2.5%-9.1%).¹ Common sites of involvement include the skin and lymph nodes, followed by testes, bone, peritoneum, and gastrointestinal tract.^{1, 2} There is no age predilection (range 1 – 81 years). Clinically, the tumors vary in size (1-20 mm) and symptoms are typically due to compression (pain or bleeding).³

The diagnosis of MS is established through immunophenotyping. Common positive immunohistochemical markers are CD43, CD68, lysozyme, myeloperoxidase, and CD117.^{2,4,5} Peripheral smear, flow cytometry, and bone marrow biopsy (BMB) are needed to categorize the disease and determine the extent of involvement. In our patient, peripheral blood smear, flow cytometry, and BMB were essential for excluding AML and CML, as well as other leukemias and lymphomas.

The literature on MS is limited to case reports and small retrospective studies, preventing the development of optimal therapeutic approaches. In the studies available, MS is generally treated with AML chemotherapy regimens and for select patients allogeneic hematopoietic stem cell transplant (HSCT).³ However, in older patients such as our patient, HSCT and intensive chemotherapy are poorly tolerated, necessitating different strategies, such as treatment with 5-azacytidine.¹

CONCLUSION

The diagnosis of myeloid sarcoma is challenging due to its rarity and heterogeneity of presentation, it can affect any organ and present at any age. MS is a histological diagnosis necessitating immunophenotyping. The tumor most commonly stains positive for CD43, CD68, lysozyme, and myeloperoxidase. While optimal treatment regimens for MS are not known, systemic AML chemotherapy regimens have been shown to prolong disease free survival and improve quality of life for patients.

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