Pleural Fluid Myeloid Sarcoma: A Case of Rare Extramedullary Involvement

Diane Carr, DO, CT(ASCP)1,2; Alexandra Mease, MD 1; Mohammad Hussaini, MD 1,2; Evita Henderson-Jackson, MD 1,2
1. Department of Pathology and Cell Biology, University of South Florida, Tampa, FL; 2. Department of Anatomic Pathology, Moffitt Cancer Center, Tampa, FL

Introduction

- Myeloid sarcoma, also known as granulocytic sarcoma, is a rare extramedullary manifestation of acute myeloid leukemia (AML) or other myeloproliferative disorders.
- The tumor consists of immature myeloid cells that form solid masses in an extramedullary site, often the skin, gastrointestinal tract, lymph nodes, or bone.
- The presence of myeloid sarcoma in pleural fluid is exceptionally rare and can pose a diagnostic challenge due to its atypical presentation and possible resemblance to other malignancies or chronic inflammatory changes.
- Accurate diagnosis can be critical for guiding treatment and improving patient outcomes.
- This case highlights the unusual presentation of a myeloid sarcoma in pleural fluid cytology and underscores the importance of thorough immunohistochemical evaluation in diagnosing extramedullary myeloid neoplasms.

Case Presentation

- A 77-year-old male with mild anemia, severe thrombocytopenia, and previous diagnosis of MDS/AML with TP53 mutation presents for inpatient hospitalization due to hypoxia and pain.
- CT pulmonary angiography shows an infiltrative mass involving the left anterior chest wall, ribs, and pleura.
- A large left pleural effusion with pleural thickening and near complete atelectasis of the left lower lobe consistent with pleural tumor involvement.
- Likely neoplastic lymphadenopathy in the left supraclavicular, axillary, subpectoral, and internal mammary regions is noted.
- Thoracentesis was performed removing 1 L of fluid with specimens submitted to pathology for cytologic evaluation and flow cytometric analysis.

Results

- Cytologic evaluation revealed a malignant effusion composed of an atypical mononuclear infiltrate with medium to large cells exhibiting immature chromatin, variable prominent nucleoli, moderate to ample cytoplasm, and occasional anaplastic cells.
- Immunohistochemical stains were performed on the cell block. The neoplastic cells were positive for CD4, CD33, CD43, CD123(weak), myeloperoxidase (weak), and negative for CD34, CD117, calretinin, TTF1, CK AE1/AE3, lysozyme, CD56, CD99, and TdT.
- Additionally, the infiltrate was positive for CD68, but negative for ALK 1, CD30, and CD163.
- The morphologic and phenotypic profile was compatible with involvement by acute myeloid leukemia (AML).
- Flow cytometry further confirmed extramedullary disease with 73% of the cells expressing AML markers.
- Shortly after diagnosis the patient requested discharge home, with supportive hospice care, and subsequently expired.

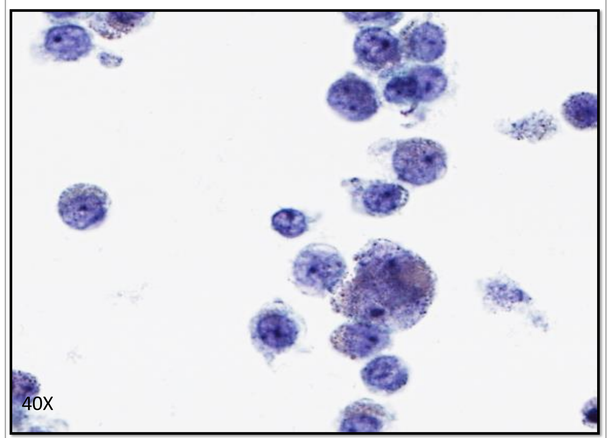

Fig 1. ThinPrep showing atypical mononuclear infiltrate comprising of medium-to-large cells with immature chromatin, variably prominent nucleoli, moderate to ample cytoplasm, and occasional anaplastic cells.

Images

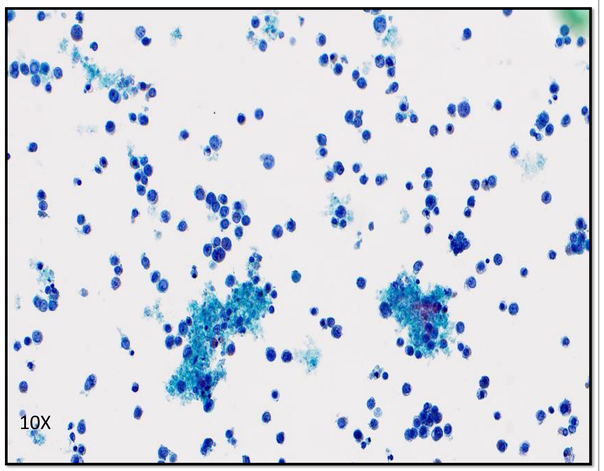

Fig 2. ThinPrep showing a highly cellular sample with abundant neoplastic appearing cells demonstrating atypical myeloid/histiocytic differentiation.

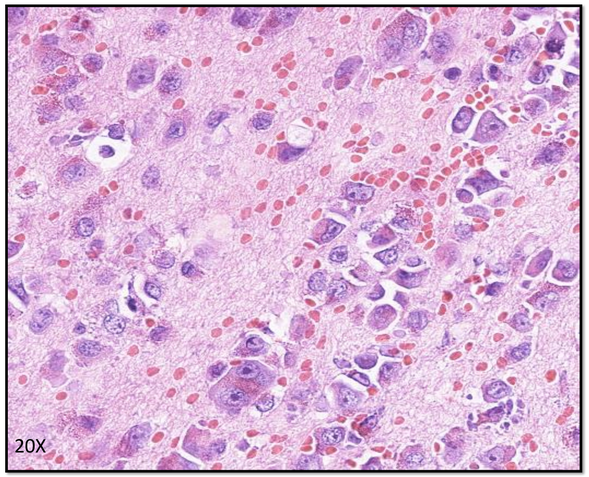

Fig 3. H&E stained cell block showing atypical, pleomorphic, immature mononuclear cells with a high nuclear-to-cytoplasmic ratio and prominent nucleoli in a background of inflammatory cells and red blood cells.

Conclusions

- Myeloid sarcoma represents a rare extramedullary manifestation of acute myeloid leukemia (AML) and related myeloid neoplasms.
- Detection in pleural fluid, as in this case is exceedingly rare and can present unique diagnostic challenges.
- Both cytology and flow cytometry are essential for identifying myeloid sarcoma in fluid specimens.
- The cellular morphology as well as the immunohistochemical staining supported the diagnosis in this case.
- Myeloid sarcoma in serous effusions can indicate advanced or relapsed AML and portend to a poor prognosis, often suggesting aggressive disease behavior and treatment resistance.
- The patient's history of progression from MDS to AML with TP53 mutation, and refractory disease despite multiple therapeutic regimens, highlights the complex clinical trajectory common in high-risk AML cases.
- Myeloid sarcoma can be challenging to diagnose due to the lack of typical blast markers like CD34 and CD117, as in this case, requiring a thorough work up to exclude other entities.
- This case underscores the value of a comprehensive diagnostic approach including review of clinical history, cytomorphology, immunohistochemistry, and flow cytometry in detecting rare presentations of AML.

References

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