Effective Treatment of Peritoneal Myeloid Sarcoma with Decitabine and Venetoclax

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Background

- Myeloid Sarcomas are rare presentations of myeloid progenitor cells in extramedullary anatomic sites, occurring with or without bone marrow involvement.
- This can occur without antecedent AML, concurrently with AML, or following initial AML diagnosis.
- Peritoneal Myeloid Sarcoma is **extremely rare**, with **no established guidelines for systemic therapy** in elderly patients above the age of 65. [1]
- There is limited data on the incidence of FLT3-ITD in myeloid sarcoma, but in an analysis by Ansari-Lari et al., the mutation was present in tissue samples in three out of twenty patients. [2]

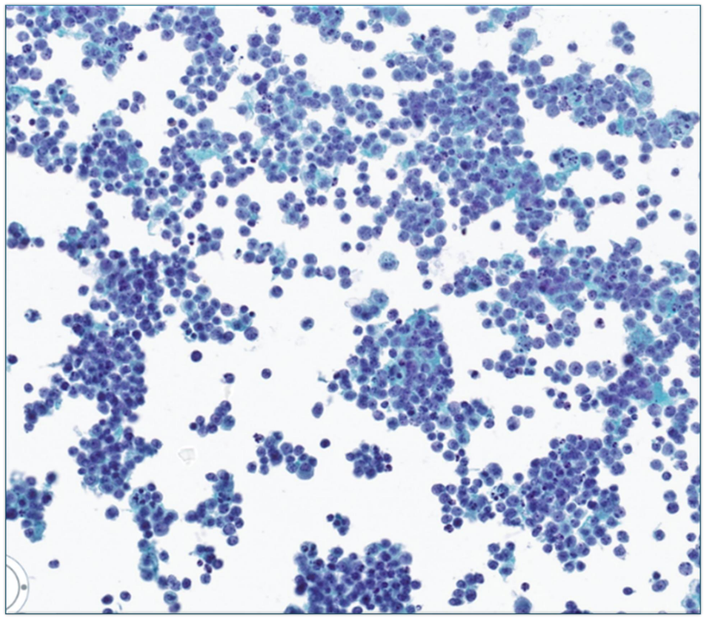


Figure 1 Cytology smear of the peritoneal fluid showing numerous immature discohesive cells with high nucleocytoplasmic ratio, immature nuclear chromatin, and prominent nucleoli (Pap stain, x200).

Clinical Presentation and Course

- 78-year-old female, former smoker with a history of depression, emphysema, ventricular bigeminy, hypertension, hyperlipidemia, and osteoporosis, presented with abdominal pain, vomiting, obstipation, and diarrhea for 6 months.
- Initial CT Abdomen and Pelvis (CTAP) revealed asymmetric jejunal/ileal wall thickening and mesenteric adenopathy, initially attributed to gastroenteritis.
- Symptoms recurred after one month with CTAP notable for progressive bowel wall thickening, ascites, and new splenomegaly.
- Paracentesis (Fig. 1) was performed with flow cytometry, revealing 60% myeloblasts (CD34 +, HLADR +, and CD33 +). Bone marrow was morphologically uninvolved, but Next Generation Sequencing (NGS) of the aspirate was positive for *FLT3-ITD*, *ASXL1*, and *TET2* mutations, consistent with **peritoneal myeloid sarcoma with minimal medullary involvement**
- Initiated on **decitabine and venetoclax**. Post-cycle 1 imaging showed interval reduction in mesenteric edema and resolution of adenopathy.
- PET/CT after cycle 5 revealed **no evidence of FDG-avid metabolic disease**, with minimal stranding in the right lower quadrant suggestive of low volume or treated disease.
- Progressive ascites after C8 treatment delay with paracentesis demonstrating monocytic cells (CD33+), with FLT-3 and NPM1 no longer detectable, but mutant *ASXL1* and p. L840Xfs (c.2518delC) mutant *TET2* remained positive. Decitabine and venetoclax were resumed.
- Most recent imaging demonstrated **resolution of bowel wall thickening and ascites.**
- Decitabine and venetoclax are continued every 4 weeks. The patient is transfusion independent and maintains an excellent PS.

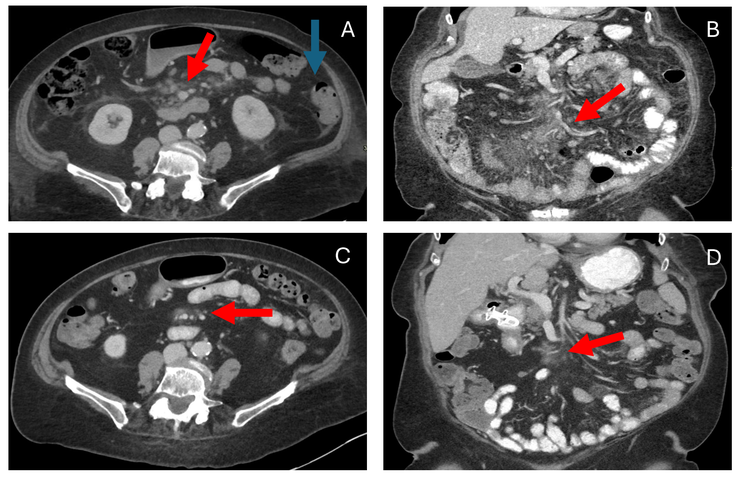


Figure 2: (A) Contrast-enhanced axial CT of the abdomen and pelvis demonstrates increased central mesenteric attenuation ("misty mesentery") and associated mesenteric lymphadenopathy (red arrow) in addition to mesenteric soft tissue edema/infiltrate (blue arrow). The central mesentery involvement can be best appreciated on the coronal reformat (B, red arrow). After initiation of therapy, there is substantial decrease in the degree of mesenteric involvement with notable decrease in size and conspicuity of mesenteric lymph nodes (C, red arrow). The overall extent of mesenteric improvement is best appreciated on the coronal plane (D, red arrow).

Discussion

- Older patients have medical contraindications, side effects, and higher risk molecular features leading to poor response to induction chemotherapy.[3]
- The VIALE-A trial demonstrated improved survival with venetoclax and hypomethylating agents in older AML patients, although its effect on extramedullary disease was not specifically studied.[4]
- **This case demonstrated the first reported instance of peritoneal myeloid sarcoma treated with decitabine and venetoclax, with the patient's clinical course showing marked clinical response and outstanding tolerability.**

References

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