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An unusual testicular mass: myeloid sarcoma as a rare extramedullary manifestation of acute myeloid leukemia

R. Padma Priya Dharshini[1*], S. Babu Peter[1], G. Chengalvarayan[1] and S. Kalpana[1]

Abstract

Background We report an unusual testicular mass with small bowel and retroperitoneal lymph node deposit proven to be myeloid sarcoma after complete histopathological and hematological workup. Myeloid sarcoma (MS) usually involves lymph nodes and head and neck regions. Uncommon sites like testis and ovary are rarely involved and pose a diagnostic challenge. Extramedullary myeloid sarcoma is most commonly associated with hematological malignancies like acute myeloid leukemia and myelodysplastic syndromes. It can precede or co-occur with AML. Considering it as a differential diagnosis in atypical presentation of testicular tumor helps in early treatment.

Case presentation We present a case of TMS with small bowel and retroperitoneal deposits presenting initially as intussusception and a vague scrotal pain. The patient underwent unilateral left radical inguinal orchidectomy. Surgical pathology revealed myeloid sarcoma of the testicle. And later jejunojejunostomy was done for small bowel deposit causing obstruction with retroperitoneal lymph node biopsy taken which revealed granulocytic sarcoma deposit. He developed peripheral blood involvement 4 weeks postoperatively, and bone marrow biopsy showed acute myeloid leukemia.

Conclusions With very short median survival period of 7.5 months, high index of suspicion is required where multifocal lesions are observed in various sites as in our case.

Keywords Testicular myeloid sarcoma, Granulocytic sarcoma, Acute myeloid leukemia, MRI scrotum

Background

Testicular MS is one of the rare types of MS, and our literature search shows that there is a paucity of data on TMS [1]. TMS with concomitant jejunal and retroperitoneal deposit as in our case has not been reported so far. Overall, MS has been classified into four types: (i) primary MS, (ii) MS as an extramedullary manifestation of AML, (iii) MS as isolated recurrence of AML and (iv) MS with coexisting bone marrow relapse of AML [2]. Histopathological examination with immunohistochemical testing remains the gold standard for diagnosis [3].

Case presentation

A 28-year-old gentleman presented with complaints of abdominal pain and constipation since 1 month with dull dragging type of scrotal pain. Ultrasound scrotum showed left extratesticular hyperechoic mass extending intraabdominally along the spermatic cord with bilateral chronic hydrocele (Fig. 1). Initial CT abdomen revealed left testicular vein thrombosis and retroperitoneal lymphadenopathy causing obstructive grade 2 left hydroureteronephrosis (Fig. 2a–c) with transient jejunojejunal intussusception (Fig. 3a, b). Blood workup showed elevated LDH (356). Patient was then subjected

*Correspondence:
R. Padma Priya Dharshini
padmaravichandran92@gmail.com
[1] Barnard Institute of Radiology, Madras Medical College, Chennai 01, India

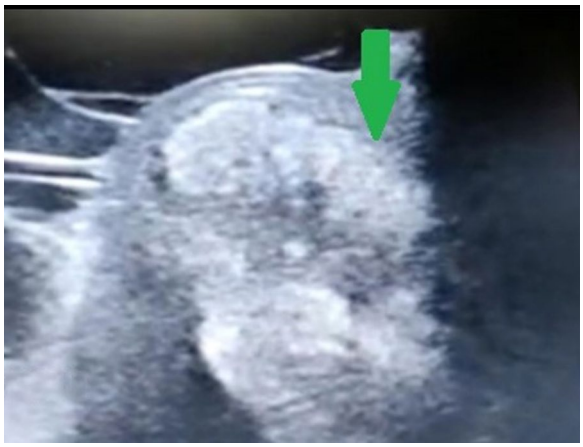

Fig. 1 High frequency ultrasound scrotum showing ill-defined extra testicular mass (arrow) with chronic hydrocele

to high-resolution MRI scrotum and abdomen in 3 Tesla scanner (Skyra, Siemens, Healthineers, Erlangen, Germany) which showed enlarged left testis with altered signal intensity, diffusion restriction and a left extratesticular mass (Fig. 4a, b). Left spermatic cord was thickened (Fig. 4c), with large retroperitoneal nodal mass abutting the gonadal vessels and compressing the ureter (Fig. 5a, b). A diagnosis of testicular lymphoma with retroperitoneal and small bowel deposit was made, and left high inguinal orchidectomy was done with left ureteric DJ stenting (Fig. 6a). IHC was positive for CD99, CD34, CD45, MPO, Ki 67–90%. Pathological diagnosis of granulocytic sarcoma was made (Fig. 6b).

1 month later during postoperative follow-up, patient presented with acute abdomen pain. CT abdomen done with oral contrast revealed circumferential wall thickening

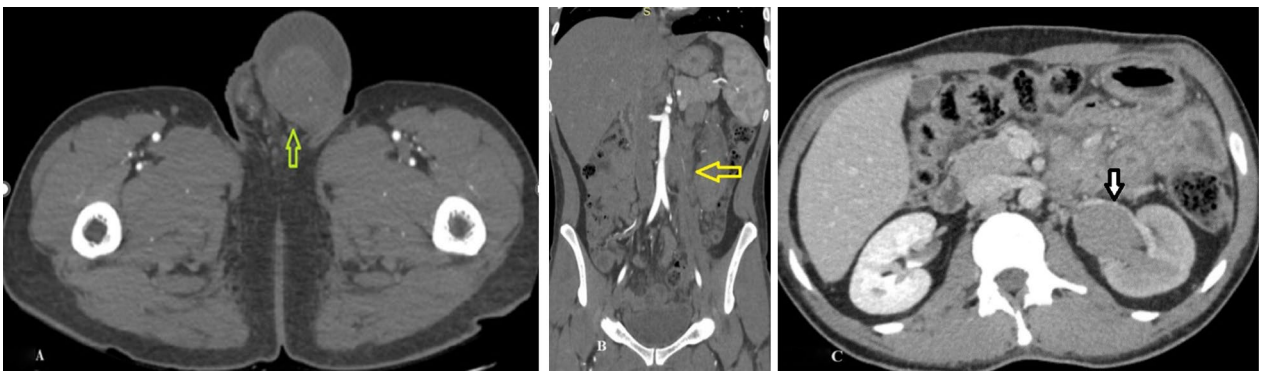

Fig. 2 **a**–**c** Intravenous contrast CT pelvis axial and coronal sections of abdomen and pelvis showing heterogeneously enhancing left testis (green arrow) with retroperitoneal nodal deposit abutting the gonadal vessel and compressing the ureter (yellow arrow) with delayed nephrogram (white arrow)

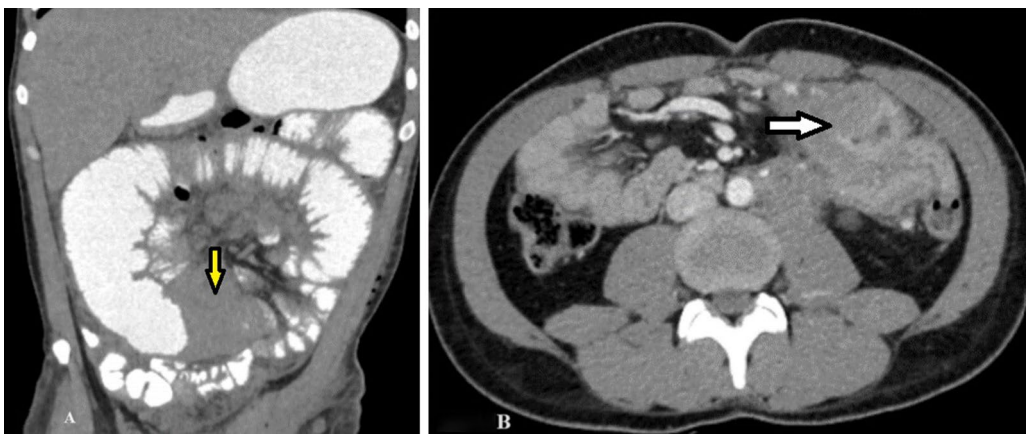

Fig. 3 **a**, **b** CT abdomen with oral contrast showing circumferential wall thickening in jejunum causing luminal obstruction (yellow arrow) was initially presented as jejunojejunal intussusception (white arrow) with circumferential mass as a lead point

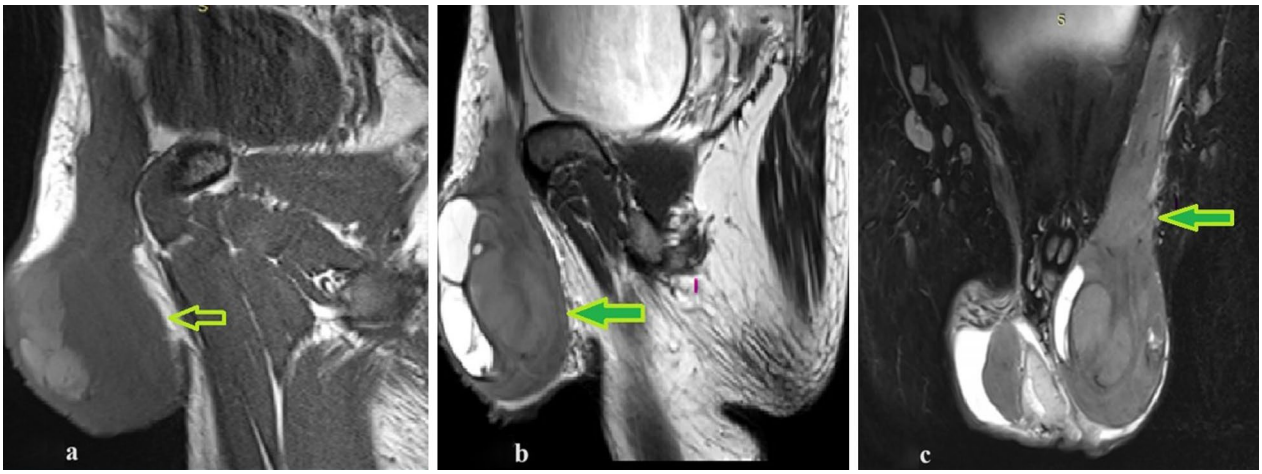

Fig. 4 **a**–**c** High-resolution MRI scrotum (T1, T2 sagittal and STIR coronal) showing T2 hyperintense bilateral hydrocele with T1 hypo- and T2 hyperintense extratesticular mass with thickened spermatic cord extending intraperitoneally

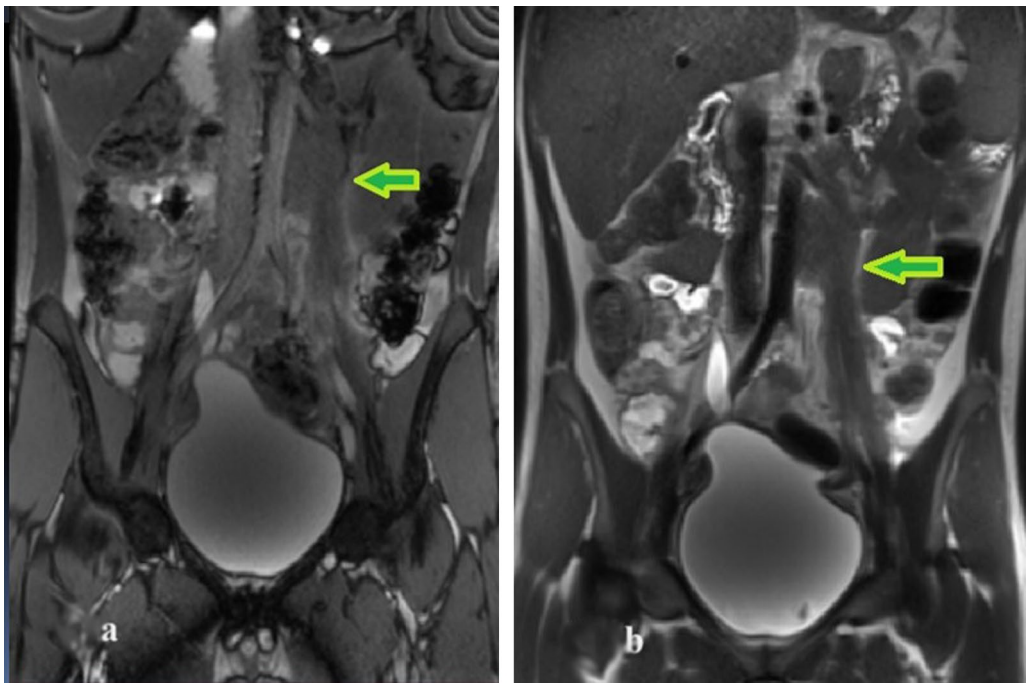

Fig. 5 **a**, **b** TRUFI, HASTE abdomen coronal showing multiple enlarged para-aortic lymph nodes abutting left gonadal vessels

in jejunum causing complete small bowel obstruction. Conservative jejunojejunostomy was done (Fig. 6c) with retroperitoneal nodal mass biopsy which showed myeloid sarcoma deposits (MPO positive). Though blood counts were normal initially, a hematological workup was made to rule out AML which commonly precedes GS. Peripheral smear showed 9% blast cells and 12% atypical cells and thrombocytopenia. Bone marrow IHC markers came positive for CD34, 45, 13, 33, 117 and MPO strongly positive, and hence, the diagnosis of AML was made.

Follow-up: Patient started on chemotherapy as a palliative treatment.

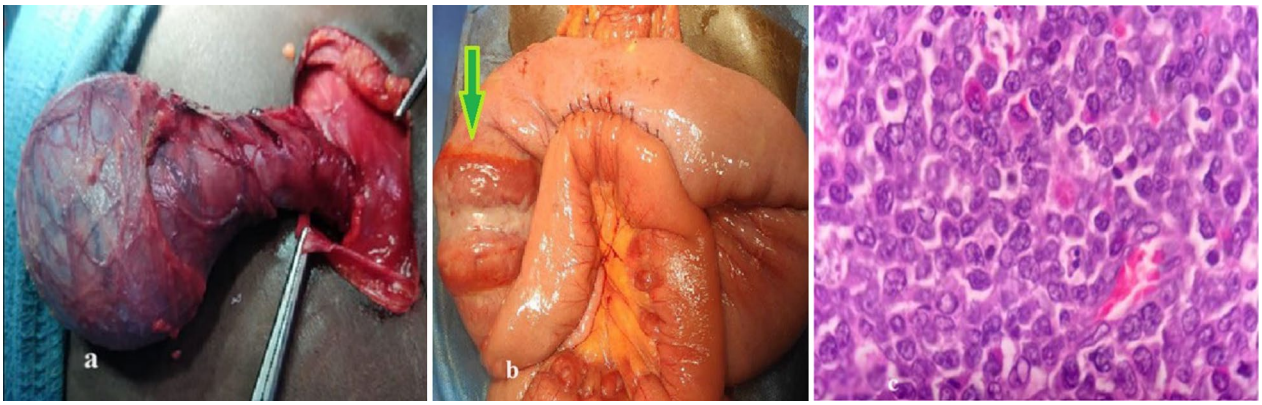

Fig. 6 **a**–**c** Intraoperative and histopathological image of left testis and small bowel deposit showing neoplastic cells which are positive for CD99, CD34, CD45, MPO, Ki 67–90%

Discussion

Granulocytic sarcoma is a rare extramedullary tumor composed of immature granulocytic precursors commonly associated with AML, CML, polycythemia vera, myelofibrosis. Rarely, it may manifest as a primary presentation before the onset of systemic disease in acute myeloid leukemia [4, 5]. We should offer them as a differential for timely workup and early diagnosis.

Granulocytic sarcoma lesions will be multiple and solid, recurring at different sites. The central nervous system, subcutaneous tissues and genitourinary system are the most common sites of disease.

At immunohistochemical staining of MS, CD68, lysozyme and CD43 are the most widely expressed markers with variable expression of a wide range of other antigens, including myeloperoxidase, CD33, CD34, CD117 (c-Kit), etc. MRI imaging features of soft-tissue granulocytic sarcoma masses are largely isointense and mildly hyperintense appearing on T1- and T2-weighted MR imaging, respectively, with variable degrees of enhancement which is difficult to differentiate from carcinoma and lymphoma. Another differential will be abscess in which the mass will be peripherally enhancing with central T1 hypo- and T2 hyperintensity with diffusion restriction. Signs strongly suggestive of granulocytic sarcoma are multiple, enhancing, solid masses occurring at different sites and time points during the course of disease in a patient with either acute myeloid leukemia or myeloproliferative or myelodysplastic disorders. Median survival period after the diagnosis of MS is 7.5 months (range 1–41 months) [6].

Conclusions

Myeloid sarcoma of testis can mimic other testicular tumors like lymphoma and metastasis. A high index of suspicion may be warranted in instances where multifocal lesions are observed in various sites as in our case to suggest this rare diagnosis and may be offered as a differential diagnosis to facilitate further workup.

Abbreviations

MS	Myeloid sarcoma
TMS	Testicular myeloid sarcoma
AML	Acute myeloid leukemia
CML	Chronic myeloid leukemia
LDH	Lactate dehydrogenase
MPO	Myeloid peroxidase
IHC	Immunohistochemistry

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Author contributions

RP performed the study and was a major contributor in writing the manuscript. SB and SK analyzed and interpreted the patient data regarding the testicular myeloid sarcoma. GC did surgery and helped in analyzing intraoperative correlation of mass. All the authors had read and confirmed the manuscript.

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Availability of data and materials

The datasets used or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Consent to participate was obtained from the patient and her attenders. Verbal consent was obtained from the patient.

Consent for publication

Consent to publication was obtained. Informed verbal consent was obtained from the study participant.

Competing interests

The authors declare that they have no competing interests.

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