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Case Report

Radiologists Hornswoggled by Machiavellian Malignant Neoplasms-Primary Ovarian Granulocytic Sarcoma: A Rare Case Report and Literature Review

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Abstract

Introduction: Myeloid sarcoma, also known as granulocytic sarcoma or chloroma, is a rare tumor composed of myeloid precursor cells occurring outside the bone marrow. These tumors are considered a unique manifestation of acute myeloid leukemia (AML) and are distinct from leukemic infiltrates. Here, we present a particularly rare case of extramedullary ovarian granulocytic sarcoma encountered at Sri Ramachandra Institute of Higher Education and Research, Chennai, Tamil Nadu, India, which is only the third reported case globally of primary ovarian granulocytic sarcoma without underlying AML.

Case Presentation: A 32-year-old woman presented with abdominal pain and was found to have a left ovarian mass, initially suspected to be torsion. She was 9 weeks pregnant and underwent management for miscarriage and cystectomy. Imaging revealed a large heterogeneous lesion in the left adnexa. Laboratory investigations showed elevated LDH levels. MRI confirmed a predominantly hypointense mass, suggestive of ovarian origin. Histopathological examination of the biopsied lesion confirmed the diagnosis of granulocytic sarcoma, with subsequent normal bone marrow investigations.

Conclusion: This case highlights the diagnostic challenge and rarity of primary ovarian granulocytic sarcoma, particularly in the absence of concurrent AML. Early recognition and diagnosis are crucial for appropriate management and follow-up to monitor for potential progression to AML. This report adds to the limited literature on this rare entity and underscores the importance of considering granulocytic sarcoma in the differential diagnosis of ovarian masses, especially in young females.

Key words: Acute myeloid leukaemia, Chloromas, Extramedullary ovarian granulocytic sarcoma/chloroma, Granulocytic sarcomas, Hematopoietic and lymphoid tissues, Leukemia; Myelogenous; Chronic; BCR-ABL Positive, Leukemia; Myeloid; Acute, Leukemia; Promyelocytic; Acute, Myelodysplastic Syndromes, Myeloid precursor cells, Myeloid sarcoma (granulocytic sarcoma or chloroma), Ovarian granulocytic sarcoma, Primary ovarian granulocytic sarcoma, Sarcoma; Myeloid

INTRODUCTION

Myeloid sarcoma (granulocytic sarcoma or chloroma) is a tumor formed by myeloid precursor cells in any localization other than the bone marrow.[1]

Chloroma/myeloid sarcomas, also called granulocytic sarcomas, chloromas are rare extramedullary masses comprised myeloid precursor cells. These types of tumors represent a unique presentation of acute myeloid leukemia, whether in isolation to, preceding, or simultaneous with bone marrow and peripheral blood disease.[2]

Different from leukemic infiltrates, myeloid sarcomas characteristically efface the involved tissue architecture and are recognized as a distinct entity in the WHO classification of tumors of hematopoietic and lymphoid tissues.

Here I'm presenting you a very rare case of extramedullary ovarian granulocytic sarcoma/chloroma that I encountered in my hospital Sri Ramachandra Institute of Higher Education and Research Porur, Chennai, Tamil Nadu-Pin code-600116, India.

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Only two cases have been recorded so far of a primary ovarian granulocytic sarcoma without underlying acute myeloid leukemia throughout the world.

Only 20 cases of granulocytic sarcoma with acute myeloid leukemia have been recorded so far worldwide.[1,2,4,5,12]

CASE PRESENTATION

A 32-year-old woman came to my institute with the following history and laboratory findings.

History

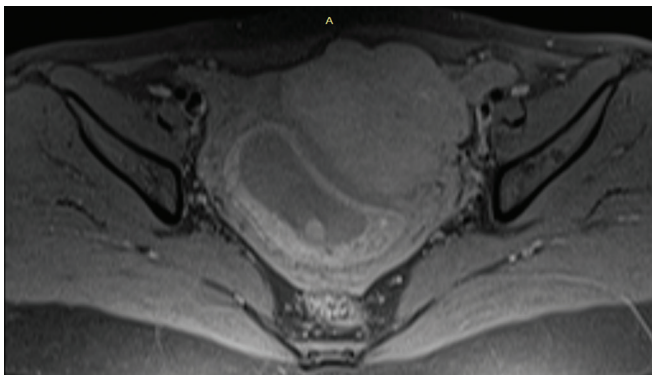
- G3P2L2A1 – 9 weeks gestation
- c/o pain in the abdomen × 1 month
- left ovarian mass – Torsion of the ovary was suspected
- patient admitted for MTP and cystectomy
- No comorbidities
- Previous h/o 2 LSCS
- PA tense abdomen, palpable mass measuring 10 × 6 centimeters.
- Left fornix full
- Outside USG scan showed left ovarian mass, large heterogenous lesion predominantly hypoechoic lesion of size 10 × 5 centimeters noted in the left adnexa.
- Screening ultrasound revealed maintained arterial and venous flow to the lesion.

Laboratory Investigations

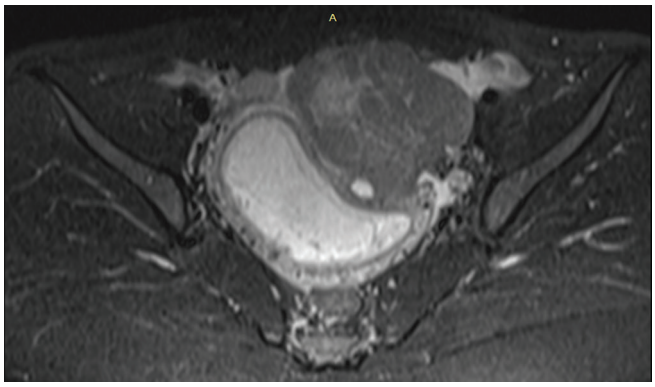
- Elevated LDH-398 (208–378).

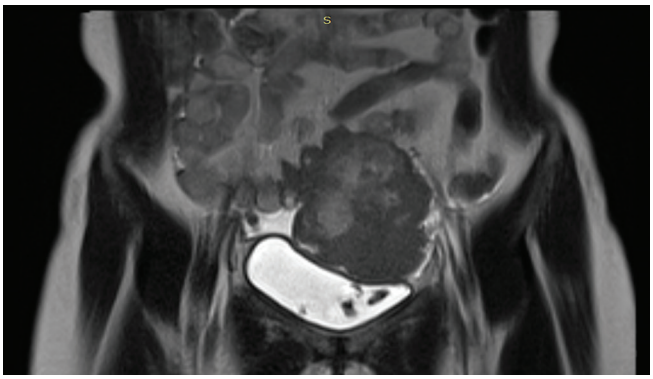
MRI Sequences

- T1 TSE TRA FS-Gravid uterus seen with single intrauterine gestation.

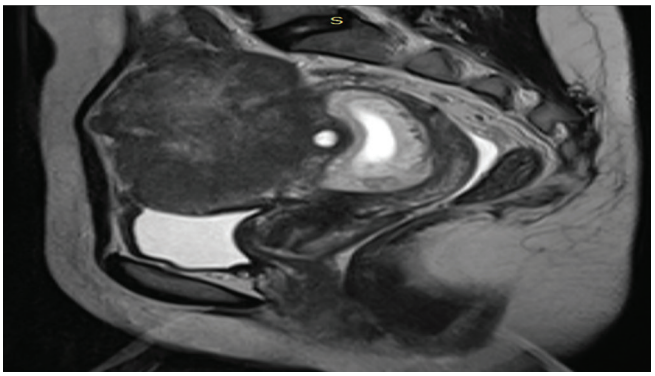


- T1 TRIM TRA-Gravid uterus seen with single intrauterine gestation.
- T2 HASTE COR A predominantly T2 hypointense left adnexal mass is seen measuring 9.2 × 6.1 × 7.7 cENTIMETERS (CC × AP × Trans).
- A few small subcentimetric follicles are seen at the periphery of the lesion, indicating that it is ovarian in origin.





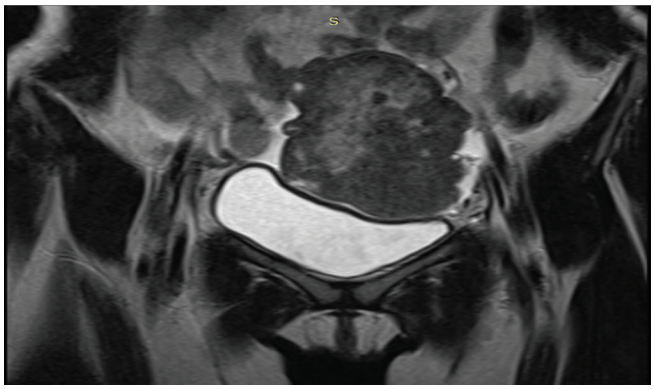
- T2 TSE SAG. A predominantly T2 hypointense left adnexal mass is seen measuring 9.2 × 6.1 × 7.7 cms (CC × AP × Trans). A few small subcentimetric follicles are seen at the periphery of the lesion, indicating that it is ovarian in origin. Mild free fluid seen in the pelvis.

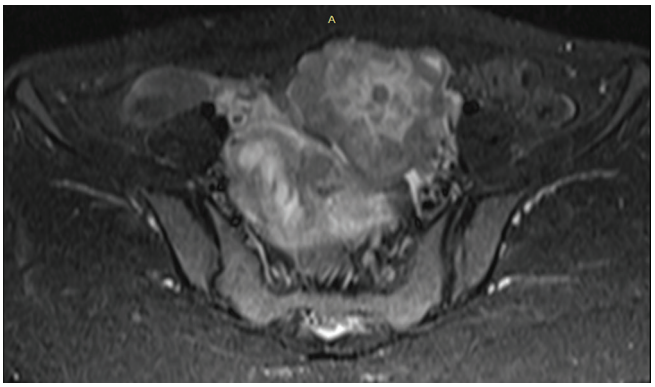


- T2 TSE COR- Normal appearing left ovary is not seen separately from this lesion which is seen anteriorly in between the left side of the uterus and the anterior abdominal wall.
- T1 TRIM TRA-1 Mildly thickened left pedicle is seen.

Histopathological Diagnosis-Granulocytic Sarcoma

The lesion was subsequently biopsied and a diagnosis of primary ovarian granulocytic sarcoma/chloroma was given after bone marrow investigations came out normal.





The patient is being followed up regularly to see if she develops acute myeloid leukemia in the future.

Through this publication, the world will know the incidence of this case.[6-8]

DISCUSSION

Rarity and Significance of the Case

- The case of primary ovarian granulocytic sarcoma (also known as chloroma) reported in this article is exceptionally rare. As noted, there are only two documented cases of primary ovarian granulocytic sarcoma without underlying acute myeloid leukemia (AML) globally. This underscores the uniqueness of the presented case, highlighting the challenge of diagnosing such rare conditions.
- Most documented cases of granulocytic sarcoma are associated with acute myeloid leukemia. The rarity of primary ovarian granulocytic sarcoma without AML makes this case particularly significant, contributing valuable data to the existing body of literature.

Clinical and Radiological Features

- The clinical presentation and radiological findings in this case align with descriptions found in the literature for granulocytic sarcoma, though they are uncommon for ovarian masses. The predominantly hypoechoic lesion and the absence of venous flow could be misleading in terms of differential diagnosis.
- The imaging characteristics—such as the large, heterogenous lesion and the specific MRI findings—are consistent with those reported for granulocytic sarcoma but are rare in the ovarian context. The case highlights the importance of considering granulocytic sarcoma in differential diagnoses when encountering unusual ovarian masses.

Histopathological Diagnosis

- Histopathological examination confirmed the diagnosis of granulocytic sarcoma. This reaffirms that, despite the rarity, proper histological evaluation is crucial for accurate diagnosis, especially when dealing with atypical presentations.
- The bone marrow investigations that came out normal further supported the diagnosis of a primary ovarian granulocytic sarcoma, illustrating the need for comprehensive diagnostic workup to rule out concurrent or preceding leukemic involvement.

Implications for Management and Follow-Up

- Given the rarity of the case, management strategies are not well-established. This case emphasizes the need for ongoing follow-up to monitor for potential development of acute myeloid leukemia, as the risk remains due to the nature of granulocytic sarcoma.
- Regular monitoring and a multidisciplinary approach involving radiologists, oncologists, and pathologists are essential to manage and potentially preempt complications or progression.

Literature Review and Comparison

- Comparing this case with existing literature, such as studies by Njoh RF, Akyol S, and others, provides context and validates the rarity of the condition. Previous literature often focuses on cases with associated AML or other hematological malignancies, which makes this primary ovarian case even more distinctive.
- The literature review also highlights that while granulocytic sarcoma is well-documented in other anatomical sites (such as the orbit or mediastinum), its occurrence as a primary ovarian tumor is extraordinarily rare and not widely reported.

Contributions to Knowledge

- This case contributes to the understanding of granulocytic sarcoma as an extramedullary tumor and provides insights into its rare presentation in the ovary. The study underscores the importance of considering rare malignancies in differential diagnoses and contributes to the broader oncological and radiological knowledge base.[1,4,11,12]

DISCUSSION AND CONCLUSION

The discussion reinforces the rarity and clinical significance of the presented case of primary ovarian granulocytic sarcoma. It highlights the need for heightened awareness among radiologists and oncologists about such rare entities and the importance of comprehensive diagnostic and follow-up strategies. This case not only adds to the limited literature on primary ovarian granulocytic sarcoma but also emphasizes the role of interdisciplinary collaboration in managing rare malignancies.

The conclusion of the article emphasizes the rarity and significance of the case of primary ovarian granulocytic sarcoma presented. The case highlights the unique presentation of granulocytic sarcoma as an extramedullary tumor, with this particular instance being exceptionally rare as it occurred without accompanying acute myeloid leukemia. The study serves as a valuable contribution to the literature on this rare condition and underscores the importance of awareness and ongoing monitoring for potential progression to leukemia.[1,3,9,10]

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DATA AVAILABILITY

This case report study was done at Sri Ramachandra Institute of Higher Education and Research, Porur, Chennai-600116, Tamil Nadu, India.

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