International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Sumathi N et al. Int J Reprod Contracept Obstet Gynecol. 2023 Jan;12(1):281-283
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20223513

Case Report

A rare case of extraovarian dysgerminoma in pregnancy

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Received: 16 November 2022
Revised: 06 December 2022
Accepted: 07 December 2022

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ABSTRACT

Extraovarian germ cell tumors are very rare and their occurrence in pregnancy is extremely rare. We presented a case of extraovarian dysgerminoma on the anterior abdominal wall in a G2P1L1, 34 weeks. It is challenging to manage such tumors, obstacles in the way namely are diagnosis as the presentation can be different from classical ovarian GCTs, limitations in the investigatory modalities in pregnancy, and management involving three independent entities-tumor, mother, fetus.

Keywords: Extraovarian GCT, Anterior abdominal wall, G2P1L1

INTRODUCTION

Cancer during pregnancy is a rare event, occurring approximately once per 1,000 pregnancies annually, corresponding to 0.07% to 0.1% of all malignant tumors.[1] Dysgerminoma is the most common germ cell tumor constituting about 1-2% of all malignant ovarian tumors. Most cases occur in adolescence and at a relatively early stage. Primary extraovarian EGCTs are thought to originate from malignant transformation of the residual primordial germ cells. The objective of this case report was to share a rare case of extraovarian dysgerminoma in pregnancy and how it was managed by multi disciplinary team approach involving specialists in OBGYN and medical oncology at our tertiary care centre.

CASE REPORT

A 28 years old G2P1L1, previous cesarean delivery, last child birth 2 years, of gestational age 34+3 weeks presented with the complaints of lower abdominal ulcerated mass and inguinal swelling.

Patient was apparently normal 4 months ago, when she noticed a swelling in the cesarean scar site. The swelling progressively increased in size over 3 months, later got ulcerated associated with pain.

The patient did not have any co-morbidities, no family history of similar illness. Her last menstrual period was 12 June 2021, estimated delivery date was 19 March 2022.

On examination, patient was moderately built, moderately nourished, vitals were within normal limits. Systemic examination was normal. Abdominal examination-uterus 34 weeks size, no contraction, presenting part mobile, fetal heart rate good. An ulcerative mass of size 15×10 cm in the lower abdomen, multiple inguinal nodes palpable.

Her investigations included basic blood evaluation, antenatal USG which were within normal limits.

MRI abdomen and pelvis

A 12.8×10.4×9.2 cm irregular proliferative mass lesion in the right side of the lower abdominal wall with restricted

diffusion and T2/T1 hetero intense areas suggesting internal hemorrhage and necrosis.

Multiple enlarged, conglomerate lobulated lymphnodes noted in right external iliac, common iliac, bilateral internal iliac, para aortic, inter aortocaval and renal hilar locations seen displacing the gravid uterus anteriorly.

Uterus was gravid GA-36-38 weeks, placenta fundal, posterior, liquor just adequate, cervical length-5.1 cm.

Right ovary-4.1×1.9 cm, Left ovary-3.9×1.8 cm.

Hepatomegaly, splenomegaly was present.

Microscopy and immunohistochemistry

Section studied shows sheets of tumor cells with vesicular nuclei, prominent nucleoli and scant cytoplasm separated by fibrous septae infiltrated by lymphocytes, SALL 4, OCT 3/4, CD 117-positive.

EMA-negative, pan cytokeratin-negative, CD3-negative, CD20-negative, cytokeratin-negative, desmin-negative, PR-negative, CD10-focally, impression- dysgerminoma. Tumour markers included AFP-159.2, beta-HCG-681, LDH-511. A final diagnosis of Dysgerminoma was made and she was admitted for further management and safe confinement.

She was transfused with 1 packed cell and was put on injection cisplatin 20 mg IV for 5 days, three more packed cells transfused.

A planned repeat lower uterine segment cesarean under general anaesthsia was performed at 36+2 weeks after getting cardiac and anaesthetic fitness. Intraoperatively bilateral Fallopian tubes normal, bilateral ovaries normal, posterior surface of uterus normal.

A mass of size 7×6 cm in the subhepatic region-nodes.

No peritoneal breech was found.

Baby details-alive late preterm girl baby. Birth weight- 2 kg. Apgar-7/10 and 8/10. Baby was admitted in NICU in view of preterm/LBW/maternal dysgerminoma on chemotherapy/D1RD and later was discharged.

Patient was reviewed by medical oncologist. She was started on BEP regimen on post operative day 7. A total of 4 cycles of chemotherapy had been given. PET CT after 4 cycles showed low grade metabolically active ulcerated soft tissue mass noted in the right inguinal region, pubic region and right lower abdominal wall.

Low grade metabolically active right sided pelvic, retroperitoneal and left supraclavicular nodes.

Low grade metabolically active peritoneal nodule and minimal ascites.

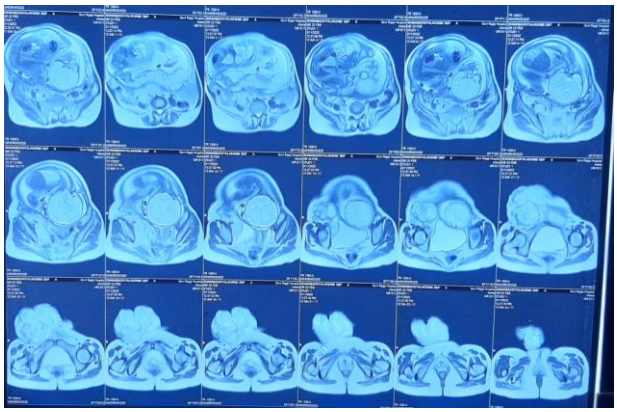

Figure 1: MRI abdomen and pelvis showing irregular proliferative mass lesion in the right side of the lower abdominal wall with restricted diffusion and T2/T1 heterointense areas suggesting internal hemorrhage and necrosis.

DISCUSSION

Cancer occurrence during pregnancy is uncommon but not so rare. Incidence is one in 1000 pregnancies annually.[1] Among gynaecological tumors, the association of dysgerminoma with pregnancy is extremely rare; the incidence is about 0.2-1 per 100,000 pregnancies.[2] Ovarian cancer represents the second most common gynaecological tumor occurring in pregnancy, dysgerminoma being the commonest ovarian neoplasm observed in this setting.[4] The pathogenesis of extraovarian dysgerminoma is unknown. Jahromi et al3 suggested primary extragonadal GCTs are thought to originate from malignant transformation of the residual primordial germ cells.[3] The differential diagnosis in this case included stromal cell sarcoma/stromal endometriosis. IHC markers SALL 4-OCT 3/4 helped clinch the diagnosis. The microscopic features of dysgerminoma are varying amounts large vesicular cells with clear cytoplasm, well differentiated cell boundaries and centrally located nuclei. The tumor cells grow in sheets or cords separated by scant fibrous stroma, which is infiltrated by lymphocytes and may contain non caseating granulomas. All ovarian and extraovarian dysgerminoma are very sensitive to platinum based chemotherapy. Yet the five year survival rate depends on lymph node status, being significantly worse in patients with positive nodes (98% vs. 90.9%).[3]

Relapse following treatment if occurs, is most commonly encountered in the first 2 years and the survival following chemotherapy of relapse is still over 90%.[5]

CONCLUSION

Cancer in pregnancy is on the raise. Although, it would be a rare encounter during one's practice, we ought to be vigilant in investigating any unusual symptomatology

during pregnancy, as it is challenging in identification during pregnancy. Surgery can be carried out in any trimester. Radiotherapy is contraindicated. With newer and safer chemotherapeutic agent we can expect modestly good prognosis in these pregnant mothers.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Sumathi N, Rajasekaran PN, Valarmathy S, Divya V, Deivasikamani D, Jayashree. A rare case of extraovarian dysgerminoma in pregnancy. Int J Reprod Contracept Obstet Gynecol 2023;12:281-3.