Growing Teratoma Syndrome

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ABSTRACT

Growing teratoma syndrome is seen in young women following surgery for immature teratoma of the ovary. It needs management with adjuvant chemotherapy following debulking procedures for residual disease.

Keywords: Growing teratoma syndrome, Immature teratoma, Malignant germ cell tumor.

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INTRODUCTION

Growing teratoma syndrome is a rare metastatic complication of malignant germ cell tumor. It was first described in non-seminomatous germ cell tumor (NSGCT) of testes. While the incidence of NSGCT of the testes treated with chemotherapy is between 1.9% and 7.5%, it is rarely seen in the ovary. We are reporting this case for its rarity.

CASE REPORT

A 22-year-old Mrs. S, G2P1L1, LCB 5 years, with LMP on October 6, 2014, and expected date of confinement on July 13, 2015, at 38 weeks of gestation admitted for safe confinement. Her antenatal period was uneventful and she was carrying a healthy fetus.

History

• *First pregnancy:* At 9 weeks of gestation, her ultrasonogram (USG) showed a live intrauterine pregnancy

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with corresponding growth and a heteroechoic right ovarian cyst of size 54×35 mm with cystic and solid components (dermoid). A repeat USG at 16 weeks showed an increase in the size of the cyst to 94×104 mm. Hence, on 20.09.2010, a laparotomy with right ovariotomy was done. Left ovary normal. *HPR-macroscopy*: Brownish nodular mass measuring

• *HPR-macroscopy:* Brownish nodular mass measuring 12 × 13 × 6 cm. Capsule appears ruptured. The cut section shows solid and cystic spaces. Solid areas whitish glistening and cystic spaces filled with mucinous material, cartilage, hair, and fat.

Microscopy shows ovarian cyst lined by squamous epithelium section from solid areas shows glial tissue, choroid plexus, lobules of fat, cartilage, bony tissue, and skin appendages.

Diagnosis: Mature teratoma (benign). She had term normal delivery one week before the expected date of confinement/3 kg male baby in February 2011. No other antepartum/intrapartum or postpartum complications.

Two months later, she had complaints of mass in the abdomen for which she consulted a doctor and ultrasonogram was done which showed a multicystic heteroechoic mass measuring 18×10.4 cm seen in the pelvis extending to the right side of the abdomen. Another mass of size 18.1×8.4 cm seen in the supra-umbilical region.

Contrast-enhanced computed tomography (CECT): Well defined heterogenous solid and cystic mass measuring 16×13 cm showing scattered areas of fat, calcification, and soft tissue component is seen in the pelvis (midline and the right side). Lesion shows minimal post contrast enhancement suggestive of immature teratoma. The lesion is seen to displace the bowel loop to the left side. Superiorly, it is reaching the level of the umbilicus. Stranding of fat seen adjacent to the lesion infiltration. Another lesion measuring 16×10 cm with similar appearance seen in the right suprarenal region. The lesion is seen abutting the posterior surface of the right lobe of the liver. The fat plane between the lesion and liver is not clearly defined. Heterogeneous attenuation lesion is also seen in the right perihepatic region and subdiaphragmatic location suggestive of immature teratoma with the capsular breach and peritoneal extension. Ascites seen; right adrenal not visualized separately, both ovaries not well visualized. The remote possibility of a capsular breach and granulomatous peritonitis also need to be considered.



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• *Blood investigations:* WNL,tumour markers-β hCG : <0.1 IU/mL, AFP-280.3 ng/mL.

Laparotomy was done on 02/05/2011, and intraoperative findings were: Uterus shows small granules, right ovary not visible, left ovary normal, a large cystic mass with solid components about 20×18 cm arising from the omentum. Ascites was present, Bowel also showed granular deposits, a large mass 18×15 cm in a subhepatic area with yellowish fluid and solid content, mass removed from the omentum and mass under the liver debulked (whole mass could not be removed) and sent for HPE.

• *HPR:* Tumor mass from omentum—immature teratoma grade 2, tumor mass under liver—peritoneal gliomatosis, omentum—teratomatous nodule with peritoneal gliomatosis and ascitic fluid cytology—occasional clusters of atypical cells and mononuclear cells.

Postoperative adjuvant chemotherapy with bleomycin, etoposide and paclitaxel regimen 5 cycles completed. The patient was on regular follow up. Repeat USG taken 2 months postoperatively showed an ill-defined mixed echogenic complex solid and cystic mass lesion in right suprarenal/subhepatic area of size 12 × 8 cm. The lesion is compressing/intending under the surface of the liver and extending peri hepatically. Omental deposits are likely.

She underwent repeat laparotomy done on 02/09/2011.

Intraoperative findings: Large retroperitoneal tumor 13×10 cm, solid and cystic, predominantly solid. Densely adherent to the diaphragm, under the surface of the liver. No ascites, no visceral disease. Tumor excised in toto enbloc with part of the diaphragm.

Patient developed haemoperitoneum in the immediate postoperative re-exploration was done. 1.5 L of blood. Bleeding from the undersurface of liver parenchyma, no major bleed; perihepatic packing is done.

Re-exploration done on 05/09/2011—bleeding from a right hepatic vein near the IVC—right hepatectomy done. Hemostasis maintained final diagnosis-growing teratoma syndrome (retroperitoneum).

The patient was on regular follow-up. No further abdominal masses detected that she conceived for the second time in October 2014 and she delivered normally on July 12, 2015 without any complications.

DISCUSSION

Growing teratoma syndrome (GTS) is defined as an enlarging mature teratoma that arises during or following chemotherapy for a malignant germ cell tumor.¹ Two possible explanations for the occurrence include selective elimination of the malignant cells by chemotherapeutic agents or differentiation of malignant cells into mature teratoma components following exposure to chemotherapeutic agents. Other explanation for the appearance of growing teratoma syndrome may be as a result of micrometastases of the remaining immature teratoma cells within the peritoneal cavity. This may be as a result of intra-abdominal dissemination despite intact capsule which may occur spontaneously preoperatively.

The presenting symptoms may be abdominal distension or abdominal discomfort as in our case and the initial histopathology may be an immature teratoma. As the patients are young they may undergo unilateral salpingoovariotomy due to fertility concerns. The development of GTS had been reported as early as 3 months and in some cases, delayed until 8 years. Our patient developed GTS after 7 months. Monitoring of response to chemotherapy is by serum tumor markers till low or normal levels.²⁻¹² However, despite normalization of serum tumor markers during chemotherapy, the metastatic tumor grows and is usually identified on radiological imaging or ultrasonography, and complete surgical excision is the treatment of choice with retroperitoneal para-aortic and pelvic lymph node dissection. Malignant transformation has been reported in 3% of the cases.

The overall prognosis for GTS is good with few reported deaths. The 5-year survival rate is 89% for patients who have undergone surgery following GTS. However close follow-up is essential as GTS has developed 10 years later on follow-up.

CONCLUSION

Our patient had two successful pregnancy outcome following GTS and is on follow-up.

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