Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/apme

Case Report

Growing teratoma syndrome – Case report and review of literature

Ruquaya Mir [a], *Sandeep Kaul* [b,*], *V.P. Singh* [a]

[a] Sr. Consultant, Surgical Oncology Indraprastha Apollo Hospital, India
[b] Surgical Oncology Indraprastha Apollo Hospital, Sarita Vihar, New Delhi, India

ARTICLE INFO

Article history:
Received 11 July 2014
Accepted 31 October 2014
Available online 24 November 2014

Keywords:
Growing teratoma syndrome
Non-seminatous germ cell tumors
Immature teratoma

ABSTRACT

Germ cell tumors (GCT) constitute less than 3% of all ovarian cancers. These tumors occur predominantly in children and women under 30 years of age. The immature ovarian teratoma is the third commonest of the germ cell tumors following dysgerminoma and endodermal sinus tumor. The growing teratoma syndrome (GTS) is an extremely rare metastatic complication of a malignant germ cell tumor. The finding of a growing solid mass (or masses) during or after chemotherapy treatment for GCT of the ovary should raise the possibility of the growing teratoma syndrome. These lesions should be resected to confirm the diagnosis, to exclude malignancy, to relieve a possible pressure on adjacent organs and to prevent a possibility of malignant transformation in future.

1. Case history

A 16/yr female resident of Uzbekistan presented to our hospital in 2013 with a diagnosis of immature teratoma for which she had underwent a trans-abdominal hysterectomy with bilateral salphingo-ophorectomy in 2010 in Uzbekistan followed by 3 cycles of standard BEP chemotherapy. Presently she had progressive abdominal distension which had started while patient was still receiving chemotherapy. She had occasional complaints of abdominal pain, vomiting and constipation but had good appetite and maintained good health. On general physical examination, her vitals were stable but pallor was present. There was a vertical midline scar extending from the xiphoid to pubic symphisis. Per abdominal examination revealed a large lobulated palpable non-tender mobile mass filling whole of the abdomen with overlying skin stretched, visible veins were present with flow from below upwards upon eliciting Harvey's sign. Lab investigations revealed Hb-10.6 g/dl, TLC 12,800 cumm^3, DLC N86L9M5, ESR-36 mm/1st hr, PCV 32.1%, Platelet Count-196,000, PTI 13.5 s, aPTT 30.6 s, INR 1.3, S-Urea-19 mg/dl, S-Creatinine-1.3 mg/dl,LDH-332 IU/L, αFP-0.66 IU/ml, HCG <2 IU/ml. Contrast enhanced computerized tomogram of the chest and abdomen revealed a large complex solid/cystic mass in the abdomen and pelvis 40 × 21 × 25 cm, occupying most of the right side of the abdominal cavity with specks of calcification, displacing small bowel and pancreas. The right lobe of the liver was distorted and scalloped by the mass, bilateral ureters revealed hydroureters. The right hemi-diaphragm was elevated with right lower lobe and rest of lung fields were clear. After informed consent patient underwent an

* *Corresponding author.*
E-mail addresses: drsandeepkaul@yahoo.com, drsandeepkaul@gmail.com (S. Kaul).
http://dx.doi.org/10.1016/j.apme.2014.10.004

exploratory laparotomy. The intra-operative findings revealed minimal ascites, extensive peritoneal disease, a large multilobulated tumor in the pelvis, abdomen and right subdiaphragmatic region closely abutting segment V, VI and VII of right lobe of liver.

Multiple well encapsulated lesions were present. The right lobe of the liver was adherent to the upper abdominal mass without any parenchymal infiltaration. Optimal cytoreduction with peritonectomy was done. The significant intra-operative events were intra-operative hypotension/hypovolemia, on tumor mobilization and IVC compression, iatrogenic diaphragmatic tear which was repaired primarily with 1.0 prolene. Intra-operatively 4 units of PRC and 2 units of FFP were transfused. The patient had an uneventful post-op recovery and was discharged on 7th post-operative day. Though the patient has not come for follow-up but relatives have informed us that she is doing well and is asymptomatic.

2. Histopathology report

The tissue show orderly arrangement and are represented by cystic spaces lined variably by gastrointestinal type epithelium, respiratory epithelium, and keratinized stratified squamous epithelium cuffed by thick layer of smooth muscle; lobules of mature adipose tissue; plates of hyaline cartilage and mature lamellar bone; lobules of seromucinous glands and gastric fundic gland; cells; blood vessels and skin appendages. Islands of primitive neuroepithelium or immature tissues are not identified in multiple sections examined.

Evidence of somatic malignancy arising within this tumor not seen. Histology picture with normal tumor markers confirms diagnosis of GTS (Fig. 1, Fig. 2, Fig. 3).

3. Discussion

The growing teratoma syndrome (GTS) is an extremely rare metastatic complication of a malignant germ cell tumor (GCT) described for the first time by Logothetis et al.[1] The syndrome is defined as a detection of an enlarged mass during or after chemotherapy treatment for GCT. The histology of the lesion is of mature teratoma with no malignant elements. The syndrome appears in 1.9–7.6% of the patients after treatment for testicular non-seminomatous germ cell tumor (NSGCT).[2] Logothetis coined this phenomenon the "growing teratoma syndrome" (GTS). According to his original description, three criteria are required for the definition of the GTS. First, normalization of previously elevated tumor markers (αFP or βHCG). Second, enlargement of the primary tumor & third, only mature teratoma elements in pathologic examination. All these criteria should be met after or during chemotherapy treatment of NSGCT.

GTS is much less common after ovarian GCT. Kattan et al[3] were the first to use the term "growing teratoma syndrome" in woman. The patient had a recurrent mature teratoma implants during chemotherapy treatment for metastatic malignant teratoma of the ovary.

One large series from a single institution, MD Anderson Cancer Center,[4] and the two large series from the Gynecologic Oncology Group[5,6] reported the presence of Mature teratoma (MT) from the second – look operation for GCT in 13/52 patients, 10/76 patients, and 21/108 patients, respectively, with an overall incidence of 17.8%.

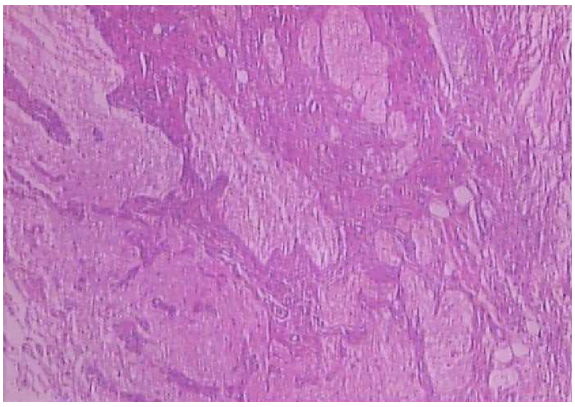

Fig. 1 – 10X nodules of mature glial tissue separated by fibrous septae.

Teratoma is a tumor whose components are derived from more than one germ cell layer. Mature teratoma (MT) is composed of well-differentiated elements, whereas immature teratoma is only partially differentiated and is in this respect akin to foetal tissue. These are distinct entities in the WHO classification. MT is classified as a benign entity and is resistant to primary chemotherapy. Surgical resection is the standard treatment for these patients. MT is the only component found during GTS. As immature teratoma is considered by some authors to be a malignant NSGCT, its presence in growing masses should not be equated with GTS.

The pathogenesis of the GTS is still a subject of controversy. Two basically distinct mechanisms are currently under consideration: malignant cell differentiation into MT or selective chemotherapy-induced destruction of components other than MT. The demonstration that malignant germ cells can differentiate into MT could support the first hypothesis. However, several arguments support the model according to which chemotherapy kills malignant cells during concomitant enlargement of MT.

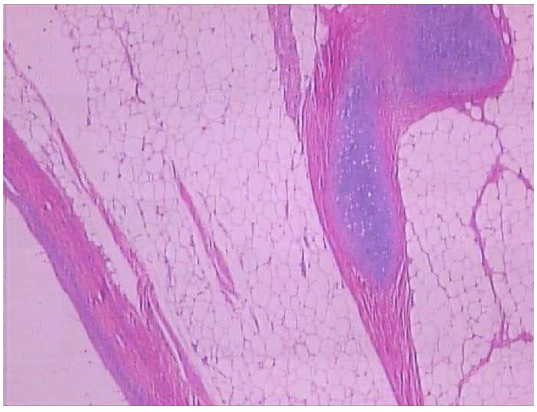

Fig. 2 – 10X admixture of plates of hyaline cartilage mature adipose tissue and stratified squamous epithelium.