Immature Teratoma with Growing Teratoma Syndrome and Gliomatosis Peritonei Coexistence

Huseyin Salih Semiz[1], Hilal Sahin[2], Sinan Unal[3], Semra Demirli Atici[4], Mehmet Faruk Kose[5] and Emine Handan Zeren[6]

[1]*Department of Medical Oncology, University of Health Sciences Tepecik Training and Research Hospital, İzmir, Turkey*
[2]*Department of Radıology, University of Health Sciences Tepecik Training and Research Hospital, İzmir, Turkey*
[3]*Department of Medical Oncology, Dokuz Eylul University Faculty of Medicine, İzmir, Turkey*
[4]*Department of General Surgery, University of Health Sciences Tepecik Training and Research Hospital, İzmir, Turkey*
[5]*Department of Gynaecologic Oncology, Acibadem University Faculty of Medicine, Istanbul, Turkey*
[6]*Department of Pathology, Acıbadem University Faculty of Medicine, Istanbul, Turkey*

ABSTRACT

Ovarian germ cell tumours constitute 5% of all ovarian cancers. During the natural course and treatment of these tumours , there may be more unusual cases. One of them is gliomatosis peritonei, which is characterised by the spread of glial cells on the peritoneal surfaces, while the other one is growing teratoma syndrome characterised by the rapid growth of benign component and loss or shrinkage of the malignant component in response to systemic chemotherapy during the treatment of germ cell tumours. Herein, we present a case of coexistence of gliomatosis peritonei and growing teratoma syndrome during the treatment of a 29-year female with immature ovarian teratoma.

Key Words: *Germ cell tumours , Growing teratoma syndrome, Gliomatosis peritonei, Ovaries*.

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INTRODUCTION

Ovarian germ cell tumours (GCTs) constitute 5% of all ovarian cancers.[1] Gliomatosis peritonei (GP) is a rare clinical condition which occurs with the spread of neuroectodermal plate originated glial cells to the peritoneal surfaces. This entity may be confused with peritoneal carcinomatosis or peritoneal tuberculosis. It is associated with mature teratoma (MT) or immature teratoma (IT) of the ovary and surgery is accepted as the primary treatment.[2] Growing teratoma syndrome (GTS) is another rare condition, characterised by the rapid growth of the benign teratoma component in contrast to the regression of the malignant component during treatment of GCTs. GTS and GP coexistence is reported to be even rarer in the literature.[3] Herein, we share a case of GTS and accompanying GP that developed during systemic chemotherapy in a 29-year female diagnosed with IT at 16 weeks of pregnancy.

Correspondence to: Dr. Semra Demirli Atici, Department of General Surgery, University of Health Sciences Tepecik Training and Research Hospital, İzmir, Turkey
E-mail: hsalihsemiz@hotmail.com

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CASE REPORT

A 29-year woman without any history of malignancy, family history, gestational history or co-morbidity presented to a gynaecologist with abdominal distension at 16th week of pregnancy. Magnetic resonance imaging (MRI) revealed a 10 cm mass originating from the left ovary with solid and cystic components, which was confirmed at laparoscopic examination (Figure 1). Left oophorectomy with aspiration of cystic component was performed during laparoscopy which demonstrated IT in frozen sections. After an uneventful pregnancy period, a left adnexal recurrent mass was found during the cesarean section which proved to be MT on biopsy. Two months after her delivery, she was referred to our medical oncology clinic with increasing abdominal distension. Abdominopelvic MRI revealed a 19.5 cm heterogenous mass in the pelvis with accompanying ascites (Figure 2a). Thoracic contrast-enhanced computed tomography was normal. In laboratory examination, alpha-fetoprotein (AFP), beta human chorionic gonadotropin (hCG) and lactate dehydrogenase (LDH) levels were; 537 ng/mL (normal: 10-20 ng/mL), 0.6 mIU/ml (normal: 0-10 mIU/ml) and 164 U/L (normal: 140-280 U/L). A high AFP level was thought to be related to the yolk sac component accompanying MT. Therefore, systemic chemotherapy was planned. In the pulmonary function tests, FEV1/FVC was found to be 74% and obstructive pulmonary disease was suspected. Considering the toxic effects of bleomycin, chemotherapy was initiated with cisplatin and etoposide. On the 10th day of the first course of

chemotherapy, the patient developed grade 3 hyponatremia, grade 3 nausea, and vomiting. The second course of chemotherapy was given with 15% dose reduction. The patient complained of abdominal distension, abdominal pain, and shortness of breath when she presented for the third course of chemotherapy. On physical examination, severe distention and diffuse dullness were detected in the abdomen. Abdominal MRI showed an increase in the size of mass up to 27 cm and new peritoneal implants (Figure 2b). The patient, whose AFP, beta-hCG, and LDH values were within normal range, was operated on considering GTS.

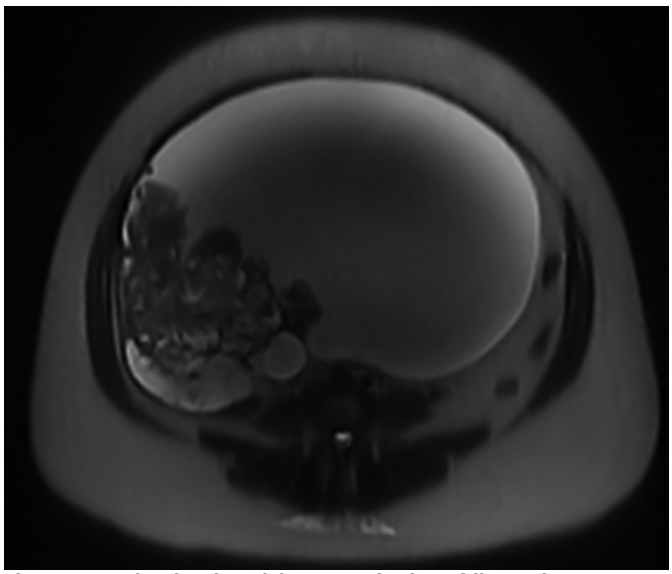

Figure 1: MRI showing the pelvic mass at the time of diagnosis.

The patient underwent pelvic mass excision, omentectomy, retroperitoneal lymph node dissection, cholecystectomy (with cholelithiasis), appendicectomy, and peritonectomy, with the resultant R0 resection. Histopathology revealed MT in the primary recurrent pelvic mass, whereas GP was detected in pelvic lymph nodes. GP was detected in the omentum, anterior abdominal wall, and bladder peritoneum. The patient was planned to be given four more courses of chemotherapy postoperatively. However, the treatment was discontinued after three courses due to grade 2 nausea and vomiting. Follow-up of the patient at the 32nd month after cessation of chemotherapy revealed no signs of recurrence or metastasis.

DISCUSSION

Both GP and GTS are rare conditions, and it is even more rare to see both of them together. In a retrospective study of 196 cases with IT, the incidence of GTS was found to be 19%, and GP to be 5%.[4] The diagnosis of GTS is usually based on three criteria: normalised tumour markers (AFP, beta-hCG, LDH) that are initially elevated, growth of masses during GCT treatment, and the absence of evidence of malignancy in histopathological examinations.[5] This patient met all three criteria. As in this patient, in case of suspicion of this entity, the primary treatment is surgery. The immediate decision of surgery is of paramount importance. Otherwise, many deleterious clinical scenarios from pressure symptoms to death may occur.

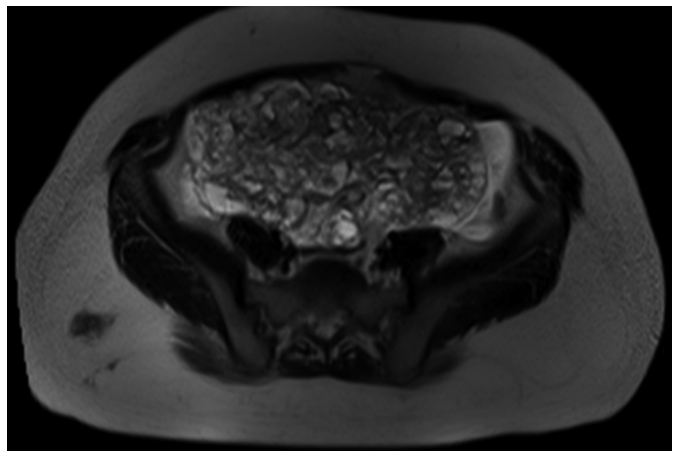

Figure 2a: MRI showing the pelvic mass before initiation of chemotherapy.

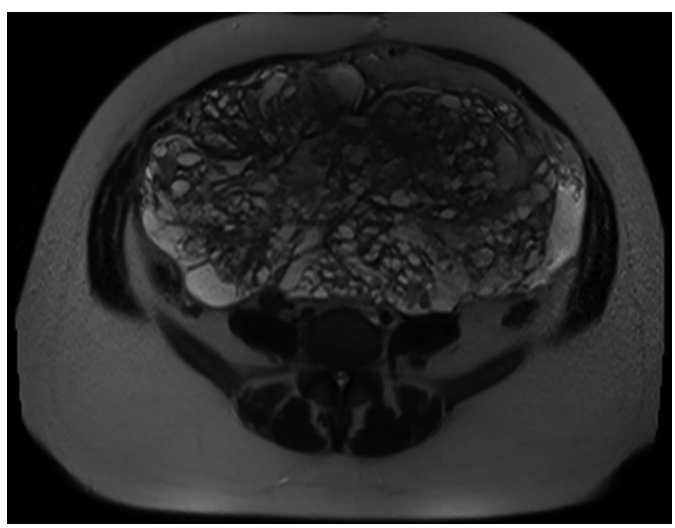

Figure 2b: MRI showing rapid progression of the mass and new peritoneal implants during the chemotherapy.

GP is generally considered to be a benign entity; however, there is no clear evidence of its origin yet. Although the presence of GP increases the risk of recurrence of peritoneal disease, it is believed that it does not adversely affect overall survival.[6] Nevertheless, on a rare occasion, it is reported to demonstrate the malignant transformation in some case reports.[7] Histological confirmation of malignancy is important if relapse is considered. Although surgery is the first-line treatment of choice in ovarian GCTs, it is also accepted as an excellent option after neoadjuvant systemic chemotherapy aiming tumour reduction in cases with bulky disease. Since our patient had bulky masses, we considered neoadjuvant therapy rather than surgery as the first-line treatment. Although there is no sufficient evidence regarding the benefit of surgery after neoadjuvant chemotherapy in those patients, there are small series of cases in the literature with 10-year survival up to 87% in FIGO stage 3 and 4 patients who were operated after neoadjuvant chemotherapy.[8] Moreover, this approach is also feasible due to the fact that ovarian GCTs are highly chemosensitive tumours.

In conclusion, this case emphasises the fact that rare develop-

ments can occur in the course of ovarian GCTs treatment and rapid intervention is life-saving. In this case, the importance of multidisciplinary management for rare clinical conditions has also been demonstrated.

PATIENT'S CONSENT:
Informed consent was obtained from the patient for the publication of this case report

COMPETING INTEREST:
The authors declared no competing interest.

AUTHORS' CONTRIBUTION:
HSS: Designed the study and drafted the manuscript.
HS: Participated in the preparation of radiological images.
HSS, SU: Interpreted the data participated in coordination and designed the experiments.
HSS, SDA: Collected the patients' data.
HSS, SU: Participated in the medical treatment of the patient.
MFK, EHZ: Supervised and conceived the study and participated in coordination.
EHZ: Participated in the examination of histopathological images.
All the authors have approved the final version of the manuscript to be published.

REFERENCES

1. Tewari K, Cappuccini F, Disaia PJ. Malignant germ cell tumours of the ovary. *Obstet Gynecol* 2000; **95(1)**:128-33. doi:10.1016/s0029-7844(99)00470-6.
2. Liang L, Zhang Y, Malpica A. Gliomatosis peritonei: A clinicopathologic and immunohistochemical study of 21 cases. *Mod Pathol* 2015; **28(12)**:1613-20. doi: 10.1038/modpathol.2015.116.
3. Takashi U, Tsutomu T, Koji T, Koichi Y, Norimasa S. Growing teratoma syndrome as an unusual cause of gliomatosis peritonei: A case report. *Gynecologic Oncology* 2005; **99(3)**: 761-3. doi: 10.1155/2011/123527.
4. Mrabti H, El Ghissassi I, Sbitti Y, Amrani M, Hachi H, Errihani H. Growing teratoma syndrome and peritoneal gliomatosis. *Case Rep Med* 2011; **2011**:123527. doi:10.1155/2011/1235274.
5. Yoon NR, Lee JW, Kim BG, Bae DS, Sohn I, Sung CO, *et al*. Gliomatosis peritonei is associated with frequent recurrence, but does not affect overall survival in patients with ovarian immature teratoma. *Virchows Arch* 2012; **461(3)**:299-304. doi:10.1007/s00428-012-1285-0
6. Dadmanesh F, Miller DM, Swenerton KD, Clement PB. Gliomatosis peritonei with malignant transformation. *Mod Pathol* 1997; **10(6)**:597-601.
7. Brown J, Friedlander M, Backes FJ, Harter P, O'Connor DM, de la Motte Rouge T, *et al*. Gynecologic cancer ıntergroup (GCIG) consensus review for ovarian germ cell tumours . *Int J Gynecol Cancer* 2014; **24(9 Suppl 3)**:S48-S54. doi:10.1097/IGC.0000000000000223.
8. Simone CG, Markham MJ, Dizon DS. Chemotherapy in ovarian germ cell tumours: A systematic review. *Gynecologic Oncology* 2016; **141(3)**:602-7. doi: 10.1016/j.ygyno.2016.02.007.