Original Research Paper

Volume - 12 | Issue - 07 | July - 2022 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Obstetrics & Gynaecology

TEEN WITH TERATOMA- A RARE CASE REPORT OF ADOLESCENT IMMATURE TERATOMA WITH MULTIPLE COMPONENTS

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ABSTRACT Immature teratoma is the second most common germ cell tumour. This has a rare incidence of less than 1% of ovarian teratomas. We report a 19 years old, unmarried girl with abdominal pain and abdominal distension since 1 month. Abdominal examination showed an abdominopelvic mass of size 20cm x 10cm seen arising from right iliac fossa extending up to umbilicus in the midline. Tumour markers were checked. CA-125 was 723 units/ml. CA 19-9 level was 13,444 units/ml. MRI of the abdomen and pelvis showed a hetero-dense abdominopelvic lesion measuring 17cm x 14cm x 10 cm arising from right ovary. It was thick walled, having cystic components with nodular soft tissue density lesion with fat and calcification. There was moderate ascites and mild right pleural effusion. She underwent staging laparotomy with frozen section which included right salpingo-oophorectomy, omentectomy, appendicectomy, right pelvic and para-aortic lymphadenectomy. Histopathological report of the specimen showed FIGO Stage 1A, Grade 2 immature teratoma. The tumour had a rare presentation of 16 immature primitive elements. She underwent adjuvant chemotherapy with etoposide and cisplatin regimen for 4 cycles. She also had ovarian suppression with GnRH analogue every 3 months. She was monitored with CA-125 and CA 19-9 levels for therapeutic response.

KEYWORDS : Immature teratoma, Germ cell tumour

INTRODUCTION

The word teratoma is derived from the Greek word "teraton" meaning monster.[1]It was initially used by Virchow. Immature teratoma is the second most common germ cell tumour. Incidence is fewer than 1% of all ovarian cancers[2]. It accounts for 10-20% of ovarian malignancies seen in the adolescent age group[2]. It is frequently seen in the first and second decades of life[3]. It can rarely occur in postmenopausal women[2].

CASE REPORT

Miss X, 19 years old, unmarried, came with complaints of abdominal pain and abdominal distension since 1 month. There is no history of any menstrual disturbances, loss of appetite, loss of weight or excess facial hair growth. There was also no bowel and bladder disturbances or breathlessness. Vitals were stable. Her body mass index was 29kg/m2. Abdominal examination showed an abdominopelvic mass of size 20cm x 10cm seen arising from right iliac fossa extending up to umbilicus in the midline. It was freely mobile, smooth in surface, regular margins, not warm or tender, not lobulated with lower border not palpable. The mass was dull on percussion. Rectal examination revealed that pouch of douglas was free.

She underwent a series of investigations. Tumour marks were checked. CA-125 was 723 units/ml and CA 19-9 level was 13,444 units/ml. They were notably elevated. Others markers like carcino-embryonic antigen (CEA), alpha feto-protein (AFP), lactate dehydrogenase (LDH) and beta hCG were normal. MRI of the abdomen and pelvis showed a hetero-dense abdominopelvic lesion measuring 17cm x 14cm x 10 cm arising from right ovary, as seen in the sagittal section in Figure 1. Figure 2 shows a coronal section of the mass. It was thick walled, having cystic components with nodular soft tissue density lesion with fat and calcification. There was moderate ascites and mild right pleural effusion.

A multi-disciplinary team of doctors involving the gynaecoloist, oncologist, pathologist and radiologist proposed the management of right salphingo-ovariotomy with frozen section. Intra-operatively, a mass of size 24cm x 20cm x 10cm was noted arising from the right ovary, extending up to the umbilical region with solid component on one side, cystic area on the other side and minimal ovarian tissue seen at the periphery, as shown in Figures 3 and 4. Left ovary appeared polycystic. The uterus, left fallopian tube, omentum, peritoneum and intestines appeared normal. Peritoneal washings were taken for cytology. The pathologist reported frozen section as a possible immature teratoma. Hence, the decision was made to proceed with a staging laparotomy that involves omentectomy, appendicectomy, right pelvic and para-aortic lymphadenectomy. Post-operative period was uneventful.

Histopathological report of the specimen showed FIGO Stage 1A, Grade 2 ovarian tumour. The tumour was limited to the right ovary with capsule intact, no tumour on the ovarian surface. Figure 5 depicts the cut section of the tumour with solid areas showing hemorrhage, calcification and cystic changes. Peritoneal washings were negative for malignancy. Right pelvic and para-aortic nodes were free of tumour.

Overall, this immature teratoma had a rare microscopic composition of 16 components, namely, skin with keratinised material, appendageal structures, pigment epithelium, smooth and skeletal muscle fibres, adipose tissue, cartilage, bone, gastric and intestinal mucinous epithelium, ciliated pseudostratified columnar epithelium, sero-mucinous salivary gland tissue, glial tissue, cerebellar tissue, choroid plexus and foci of immature neuroectodermal elements. Figure 6 shows a microscopic image of the tumour showing immature neuroglial tissue, neuro-epithelial rosette seen in a background of cellular stroma. Figure 7 shows the presence of cartilage, bone and salivary glands in the tumour tissue. Figure 8 elicits the presence of stratified squamous epithelium, ciliated columnar epithelium, sebaceous glands and hair follicles.

During the post-operative follow up, she underwent adjuvant chemotherapy, namely etoposide and cisplatin regimen for 4 cycles. She also underwent ovarian suppression with GnRH analogue, Inj Leuprolide 11.25mg intramuscular every 3 months. She was monitored with CA-125 and CA 19-9 levels for therapeutic response.

DISCUSSION

Teratoma is a common germ cell tumour with an incidence of 25% of

all ovarian tumours.[4] It is derived from the 3 basic germ cell layers.[4] It is usually divided into mature and immature teratomas. Mature teratomas are always benign with typical composition of various tissues and organs. Immature teratomas comprises of immature primitive components. It is commonly found in the first and second decades of life.[4] This has a rare incidence of less than 1% of ovarian teratomas. There are a few ways of classifying teratoms. They can be classified using the Gonzales-Crussi grading system[5]: 0- mature, 1- immature, possibly benign, 2- immature, possibly malignant. Another method is based on the contents of the teratoma. They can be solid teratomas containing tissues from more complex structures, cystic teratomas containing pockets of fluid like cerebrospinal fluid, sebum or fat and mixed teratomas having both solid and cystic parts.

Immature teratomas commonly present in the younger age group. Our patient presented with symptoms at 19 years of age. Hence, management is better tailored in such a way that we keep fertility preservation in mind. In our patient, the uterus and left ovary were conserved, after considering the stage and grade of the teratoma. Close post-operative follow up in such young patients who desire future fertility is vital due to the possibility of recurrence. H.P. Gupta et al[6] has reported a case of a 18-year old, who was previously operated for unilateral mature ovarian teratoma, presenting with a recurrent but immature ovarian teratoma on the opposite side. Due to adhesion of recurrent tumour on uterine serosa and malignant look of the tumour, she had undergone hysterectomy with tumour removal. Lee et al[7] has also reported a rare retroconversion of immature to mature teratoma with multiple recurrences in a 20 year old post-chemotherapeutically, where the girl succumbed to the disease. A combined data analysis reviewing 81 adult ovarian immature teratomas revealed a relapse rate of 0, 3.7, and 20% in cases graded 1, 2, and 3, respectively.[8]

Our case shows the occurrence of immature teratoma of Grade 2 in an adolescent girl who underwent chemotherapy and currently recovering well with regular follow up. Similarly, Derveshi M et al[9] have reported an 18-year old with a high grade (grade 3) immature teratoma who unfortunately succumbed to the disease. Mature teratomas usually have an excellent prognosis. However, the outlook for patients with immature teratomas depends on the stage, amount of various components and grade of the tumour. The higher the grade, the poorer the prognosis.[10] In our patient, she has a rare presentation of 16 immature primitive components, as mentioned earlier, in the immature ovarian teratoma. She is currently under close follow-up.

To conclude, raising awareness among young girls on self-examination is important, despite masking factors like obesity, as ovarian tumours do not always cause pain abdomen or menstrual disturbances. Early recognition and prompt management by a multi-disciplinary team is vital for the best prognosis.

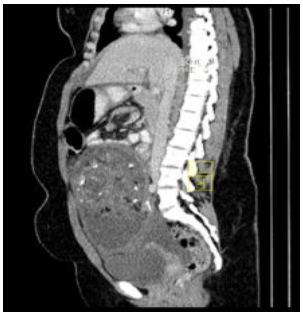

Figure 1

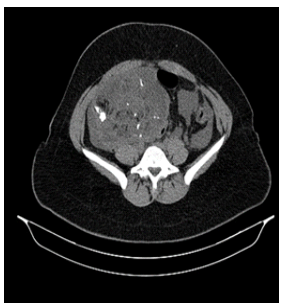

Figure 2

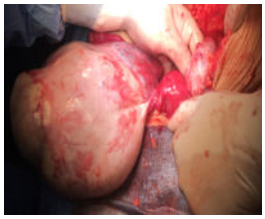

Figure 3

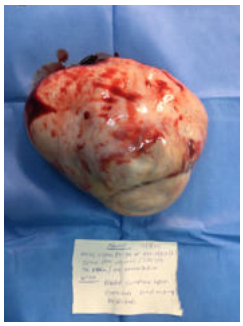

Figure 4

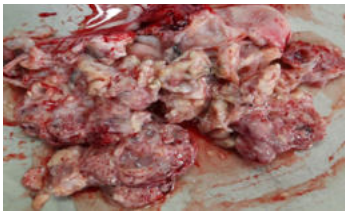

Figure 5

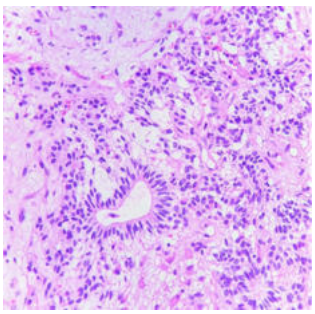

Figure 6

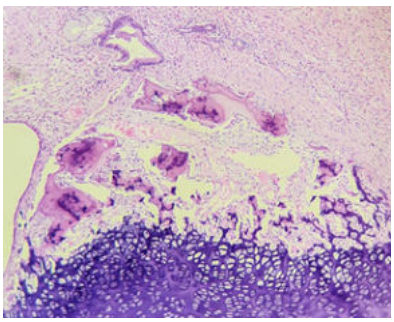

Figure 7

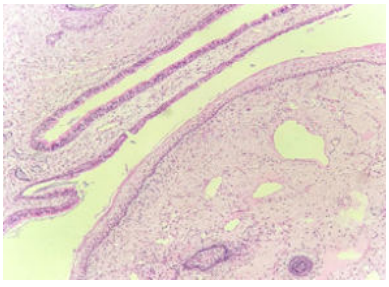

Figure 8

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