OncoExpert
INTERNATIONAL JOURNAL OF INTEGRATED ONCOLOGY

Received on 14th November, 2018; Received in revised form 14th December, 2018; Accepted, 18th December, 2018

MIXED GERM CELL TUMOR OF OVARY AND MAYER ROKITANSKY KUSTNER HAUSER SYNDROME: AN UNUSUAL COMBINATION!

Rajiv Gandhi K, Surendra Kumar Saini*, Jitesh Sarvaiya, Mitanshu Sharma, Dhara Kacha
Department of Radiotherapy & Oncology, M P Shah Medical College & Associated G G Govt. Hospital, Jamnagar, Gujarat (INDIA)

Keywords:
Mayer Rokitansky Kustner Hauser, Mixed Germ Cell, Mullerian Agenesis, Ovarian Tumor, Primary Amenorrhea, Syndrome

Corresponding author:

Surendra Kumar Saini,
Assistant Professor,
Department of Radiotherapy & Oncology,
M P Shah Medical College & Associated G G Govt. Hospital, Jamnagar, Gujarat (INDIA)
Email drsurensaini@gmail.com

Abstract: Germ cell tumor of ovary in young female is most common ovarian neoplasm but represent less than three percent of all ovarian malignancies in females. Mixed germ cell tumor is a variant of germ cell tumors and its presentation with Mayer Rokitansky Kustner Hauser Syndrome, which represents the most common type of utero-vaginal agenesis, is very unusual. However, for patients with primary amenorrhea, Mayer Rokitansky Kustner Hauser syndrome is the second common cause. We describe an interesting, uncommon and unexpected case in a young Indian female patient with primary amenorrhea due to this syndrome coexisting with mixed germ cell tumor.

Introduction

Mayer Rokitansky Kustner Hauser (MRKH) Syndrome is a rare syndrome characterized by congenital aplasia of uterus and upper part of vagina to a variable degree. Women show normal development of secondary sexual characteristics and a normal 46, XX karyotype. Isolated cases of epithelial ovarian tumors have been described rarely with MRKH syndrome in the literature. We describe here an atypical case of MRKH syndrome who presented with abdominal mass and on evaluation was diagnosed as mixed germ cell tumor of the ovary.

Case report

An 18 years, unmarried, Indian female presented in the department of gynecology of G G Government Hospital affiliated with M P Shah Government Medical College, Jamnagar with complaints of lower abdominal pain for 8 months, abdominal distension for 3 months and constipation for 15 days. Other than primary amenorrhea, she had no other significant history.

Per abdomen examination revealed a side to side mobile, solid mass arising from pelvic cavity, occupying all quadrants of abdomen and reaching superiorly up to epigastric region. External genitalia and external urethral meatus were normal. She had appropriate development of secondary sexual characteristics for her age (B/L Breast -Tanner scale II, Pubic hair- Tanner scale II).

On evaluation for abdominal distension, she was found to have absent uterus and rudimentary vagina in ultrasound scan. She did not have any hearing, cardiovascular, respiratory, or skeletal anomalies.

On imaging with contrast enhanced CT scan, a multi-loculated, solid cystic mass of size 9.8 x 16.7 x 21 cm^3 arising from pelvic cavity was seen, with bilateral ovaries not being separately visualized from the mass lesion. Uterus was not visualized in pelvic cavity. Mild ascites was noted. There was no peritoneal or omental deposit. Bilateral kidneys, liver and all other solid and hollow organs were normal. Her chromosomal study showed modal chromosome karyotype, 46XX (normal female karyotype in all metaphases).

Based on clinical, radiological and karyotype study, she was diagnosed as having MRKH Syndrome. Preoperative serum alpha fetoprotein (AFP) was more than 1000 IU/ml (normal value 0-15 IU/ml) while serum LDH was marginally elevated to 353 U/L (normal value 140-280 U/L). Beta-HCG (Human chorionic gonadotropin) and CA-125 were within normal limits.

She underwent laparotomy and right ovarian tumor removal with omental biopsy, peritoneal biopsy and peritoneal wash. Per-operative findings revealed a multi-loculated, predominantly cystic, 20 x 15 x 9 cm^3 mass with irregular surface involving right ovary. Histopathology report showed it to be a mixed germ cell tumor, with components of yolk sac tumor as well as embryonal carcinoma, associated with various differentiations like hepatoid and intestinal [Fig 1 and 2]. Omental and peritoneal biopsy specimens and peritoneal fluids were negative for malignancy. Post operative recovery was uneventful.

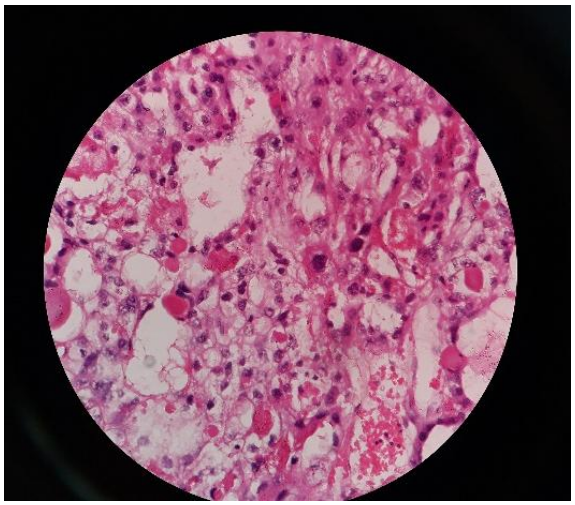

Fig 1: Microcystic pattern loose meshwork lined by cuboidal cells, round to polygonal tumor cells

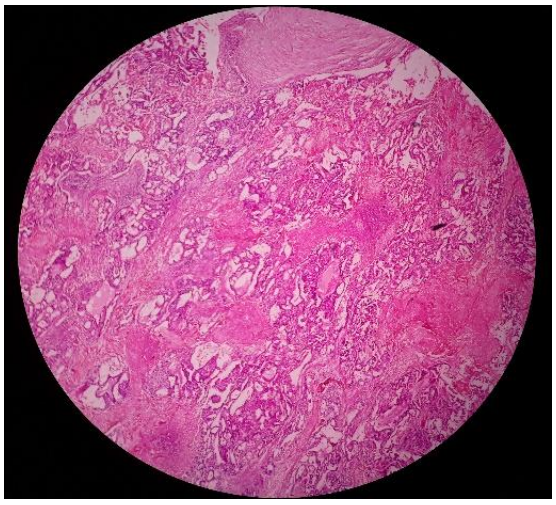

Fig 2: Occasional Schiller Duval bodies (central vessel surrounded by tumor cells)

Two weeks following surgery, serum markers study showed AFP level to more than 1000 IU/ml but other markers within normal ranges. She was planned for adjuvant combination chemotherapy with Bleomycin, Etoposide and Cisplatin (BEP regimen). After two cycles of BEP regimen, her pulmonary function test exhibited restrictive pattern, so in subsequent two cycles only Etoposide and Cisplatin were given. Further chemotherapy was stopped due to intolerance. On first month of her follow up after chemotherapy, serum AFP level decreased to 44.20 IU/ml and serum beta-HCG and LDH levels were within normal range. On second monthly follow up, her serum AFP level further decreased and came to 14.10 IU/ml (within normal limits).

Discussion

The association of a mixed germ cell tumor with MRKH syndrome is extremely rare. Patients with MRKH syndrome are not known to have increased risk for germ cell tumors. Therefore, the coexistence in this case could simply be incidental. To the best of our knowledge this case report represents the fifth reported case of coexistent malignant germ cell tumor with Mayer-Rokitansky-Kustner-Hauser syndrome and first case of mixed germ cell tumor with Mayer-Rokitansky-Kustner-Hauser syndrome[1-4.] Among the four reported cases so far; two were of yolk sac histology, one was immature teratoma and one recent report from Ethiopia of non specific germ cell tumor histology.

Normal breast development and an absent uterus are known to have testicular feminization (46 XY), Mayer Rokitansky Kustner Hauser syndrome (46XX), or some variation of both. Although testicular feminization may have up to 20% incidence of coexistent ovarian germ cell malignancies but no such pattern has been observed with MRKH. Serum marker study usually diagnose germ cell tumor preoperatively. It is advisable to get the chromosomal analysis done to differentiate these two conditions and variants to decide appropriate treatment, planning of genital organ repair and follow up.

References

1. Koonings PP, al-Marayati L, Schlaerth JB, Lobo RA. Mayer-Rokitansky-Kustner-Hauser syndrome associated with endodermal sinus tumor of the ovary. Fertil Steril. 1991 Sep;56(3):577-8.
2. Tsaur GT, Lee MH, Su SL, Wu MJ, Huang TW. Mayer Rokitansky-Kustner-Hauser syndrome with immature teratoma of the ovary at age 4 years. Gynecol Oncol. 1995;56: 456-9.
3. Takeuchi, K., Oomori, S., Oda, N., Maeda, K., Kaji, Y. and Maruo, T. Coexistence of Mayer Rokitansky Kustner Hauser syndrome and yolk sac tumor of the ovary in a prepubertal girl. Acta Obstetricia et Gynecologica Scandinavica. 2006;85:245-7.
4. Tesfaye G, Fekad B, Worku A. A Child with Malignant Ovarian Tumor and Mullerian Anomally (Mayer Rokitansky Kustner Hauser Syndrome). J Cancer Diagn. 2017;2(1): 109.

How to cite this article:

Gandhi KR, Saini SK, Sarvaiya J, Sharma M, Kacha D: Mixed germ cell tumor of ovary and Mayer Rokitansky Kustner Hauser syndrome: an unusual combination! OncoExpert 2018:4(1); 11-13