

Unique combination of an ovarian dysgerminoma, streak ovary and uterine agenesis in a girl of 12

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SUMMARY

Dysgerminoma of the ovary is a rare malignancy. It is common in dysgenetic ovaries. The prognosis is excellent if treated properly. This is a case report of a girl with agenesis of the uterus, streak ovary and dysgerminoma. Her external genitalia seemed normal. Ultrasonography indicated uterine agenesis and showed a solid mass on right gonad. The left gonad was streak ovary. There was minimal ascites and cytology was positive for malignant cell. Tumor markers were normal. Bilateral gonadectomy with removal of the tumor was performed. She received chemotherapy and was disease free at 2-year follow up. The combination of an ovarian dysgerminoma with uterine agenesis and streak ovary is a rare finding. Ovarian dysgerminoma is chemosensitive and potentially curable even when in advanced stage.

Key words: Dysgerminoma; Ovary; Abnormalities, Multiple; Gonadal Dysgenesis; Gynecologic Surgical Procedures

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INTRODUCTION

Dysgerminoma, the most common malignant ovarian germ cell tumor, is the ovarian counterpart of testicular seminoma and is derived from undifferentiated primordial germ cells (1,2). Majority of dysgerminomas occur in patients in the third and fourth decades of life. They are bilateral in about 15% of cases (2). Some occur in patients with gonadal dysgenesis, including pseudohermaphroditism. Dysgerminoma can be associated with pure gonadal dysgenesis or Swyer syndrome, mixed gonadal dysgenesis and partial gonadal dysgenesis (3,4). Ovarian dysgerminoma has also been reported in Mayer-Rokitansky-Küster-Hauser syndrome (5).

CASE REPORT

A 12-year-old girl presented with a history of painless abdominal lump. The father had accidentally felt a mass in her lower abdomen, which prompted him to seek medical attention for her. She had been born at term, vaginally, after an uneventful pregnancy resulting from a non-consanguineous marriage. She had normal growth development and good school performance but had not attained menarche. Her abdomen was protuberant, with a large, firm and non-tender mass in the lower abdomen (Figure 1). The external genitalia seemed normal.



Figure 1. Dysgerminoma in the right ovary

Imaging studies revealed a large abdominopelvic solid mass (12 cm), which appeared to be arising from the right ovary and the uterus was not

visualized. Metastatic workup revealed no disease elsewhere. X ray chest was normal.

The left gonad was elongated and small streak ovary (Figure 2). There was no abdominopelvic lymphadenopathy. She had minimal ascites and the peritoneal cytology was positive for malignant cell. However, tumor markers (B-HCG, LDH, and AFP) were normal. Karyotyping could not be done because of its unavailability.



Figure 2. Left streak ovary with fallopian tube

Bilateral gonadectomy with removal of the tumor along with infracolic omentectomy was performed. Other abdominal organs appeared normal. Dysgerminoma in the right gonad was documented in the histopathology. She had dysgerminoma in the right ovary, left streak ovary and uterine agenesis. She received adjuvant chemotherapy (BEP x 6 cycles) and is now disease free with normal tumor markers and no evidence of tumor in clinical and radiological evaluation after 2 years of follow-up. She might be needing hormone replacement later on and is still in close follow up.

DISCUSSION

This girl has a unique combination of an ovarian dysgerminoma in right ovary with uterine agenesis and her left ovary was abnormal (streak ovary). There is a high risk of neoplasm in dysgenetic gonads.

Dysgerminomas tend to occur frequently in the adolescence period. Ovarian dysgerminoma has also been reported in Mayer-Rokitansky-Küster-Hauser

syndrome (5). Absence of the uterus, also known as Müllerian agenesis or Rokitansky syndrome, affects one in every 4,000 to 5,000 women. The Rokitansky Kuster Hauser syndrome that is characterized by developmental failure of the Müllerian duct structures often occurs with urinary tract abnormalities and skeletal abnormalities (6).

Although conservative surgical management is desirable for nulliparous women with unilateral dysgerminomas, the presence of 46, XY gonadal dysgenesis should be suspected in all premenarchal girls with ovarian germ cell malignancies. If karyotyping shows the presence of Y chromosome, bilateral gonadectomy is indicated because of the risk that another tumor may develop in the other ovary (7). Because these patients already are infertile, fertility preservation during the surgery is not an issue. Karyotyping was not done in this case, as this facility was not available in our center.

Dysgerminoma is a highly radiosensitive tumor and commonly treated with combined surgery and radiation. Combination chemotherapy is equally effective and is preferred than radiotherapy nowadays (8). As metastasis does not occur early in the course of disease, mostly via the lymphatic system, a five-year survival is 75%-90% (2,9).

CONCLUSION

The combination of uterine agenesis and streak ovary with ovarian dysgerminoma is a real and rare possibility. The prognosis is good if the tumor is detected early and treated judiciously. However, the patient cannot conceive owing to the absence of uterus and / or dysgenetic gonads. It is also mandatory to screen the siblings of a patient diagnosed with gonadal dysgenesis and malignancy for similar abnormalities.

Conflict of interest

We declare no conflicts of interest.

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