MAXIMAL FERTILITY SPARING SURGERY IN A PATIENT WITH A RARE MIXED GERM CELL TUMOR

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Abstract- Ovarian mixed germ cell tumors consisting of endodermal sinus tumor and immature teratoma are very rare and have been reported only in a few case reports. Here we report a rare mixed germ cell tumor of ovary which was consisted of endodermal sinus tumor and immature teratoma components with an unusual intraabdominal location. Patient was a 21 years old girl with the chief compliant of abdominal pain. Ultrasound and CT scan showed a lobulated cystic mass. Laparotomy was performed and due to unusual localization, in which tumor was localized as a tumoral bridge between two ovaries, we performed maximal fertility sparing surgery by preserving ovaries, tubes and uterus. After surgery, 4 courses of chemotherapy with bleomycin, etoposide, and cis-platinum were done. Alfa feto-protein became negative after 3 courses of chemotherapy and she was under observation and in good health during follow up period.

Acta Medica Iranica, 42(5): 393-396; 2004

Key words: Germ cell tumor, endodermal sinus tumor, teratoma, yolk sac tumor, tumor marker, fertility sparing surgery

INTRODUCTION

Ovarian germ cell tumors represent a relatively small proportion of all ovarian tumors (20%). Teratomas, the most common germ call tumor, account for approximately 15% of all ovarian tumors, of which 95% are mature cystic teratoma (dermoid cyst).

The most common forms of malignant germ cell tumors (%5 of all ovarian germ cell tumors) are dysgerminoma, endodermal sinus tumor (EST) and immature teratoma, in decreasing order of frequency. Mixed germ cell tumors, which contain at least two components of malignant germ cell tumors, are rare (1).

Received: 11 Jan. 2003, Revised: 23 May 2004, Accepted: 26 May 2004

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In a case series study, the most common components of such tumors were reported as dysgerminoma (80%), EST (70%), immature teratoma (53%), choriocarcinoma (20%), and embryonal carcinoma (16%). The most common combination was dysgerminoma and EST (2). Ovarian mixed germ cell tumors consisting of endodermal sinus tumor and immature teratoma are very rare and have been reported only in a few case reports (3). Abdominal pain with or without pelvic pain is the most common presenting symptom in ovarian germ cell tumors. These symptoms could be seen in up to 75% of cases, but abdominal or pelvic masses without any symptom could be detected in just 10% of cases.

Most of EST tumors secrete alfa feto-protein (AFP) (4) and can arise almost in any part of the body, from the pineal to the coccygeal region (5,6). Mixed lesions may secrete AFP, human chorionic gonadotropin (hCG), both or neither, depending on the components of tumor.

CASE REPORT

A female patient, 21 years old and virgin, was referred to Vali-e-Asr Hospital in August 2003 with the chief complaints of flank pain and abdominal mass. On physical examination, a huge mass which its upper limit was in epigastric region (xyphoid process) could be palpated. Five days prior to hospitalization, an ultrasound reported cystic mass containing internal septums (lobulated mass) that occupied the whole abdominal cavity. CT scan showed a cystic mass with thick septums extended from pelvis to lower borders of the liver.

Prior to surgery, tumor markers were reported as CA-125 = 250 IU/ml, ß-hCG < 8.2 mIU/ml, LDH = 478 IU/ml and AFP = 1410 ηg/ml.

After four days of hospitalization and obtaining the necessary tests, surgery was performed. Because of hugeness of tumor, the operation was performed through a midline incision from xyphoid to pubis. After drainage of about 500 ml of serosal ascites, we explored the abdomen and a huge mass (8 kg) was found that bridged between two apparently normal ovaries (Fig. 1).

Tumor was attached to the right ovary (pediculated) and was bonded to the left ovary by adhesive bands (Figures 2 and 3).

Tumoral mass was resected and sent for frozen section. Frozen section report was an 8 kg mass of mature cystic teratoma. During the period of frozen section reporting, we performed partial omentectomy and appendectomy but we conserved ovaries completely.

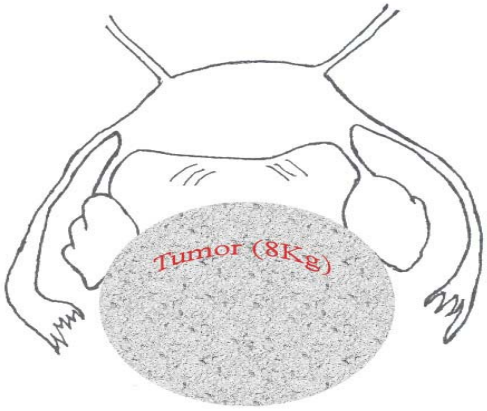

Fig. 1. Schematic view of the unusual localization of the tumor.

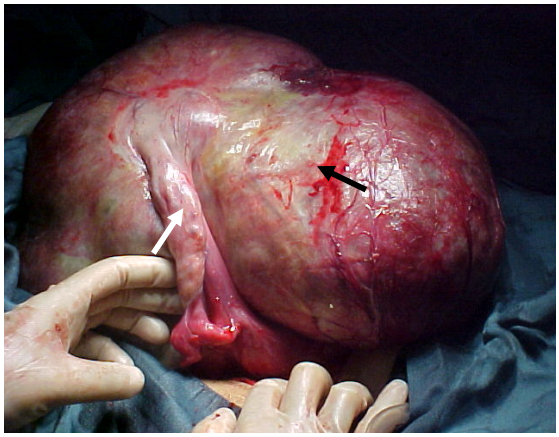

Fig. 2. Right ovary and the mass (white arrow, right ovary; black arrow, mass).

Cytology report of ascites was negative but mass was reported as a mixed germ cell tumor that consisted of immature teratoma (95%) and yolk sac tumor (5%) (Figures 4 and 5). After surgery tumor markers decreased, CA-125 to 229 IU/ml and AFP to 425 ηg/ml.

Because of pathology report of mixed germ cell tumor, patient received four courses of BEP (bleomycin, etoposide, and cis-platinum), once every three weeks. After 3 courses of chemotherapy, AFP and CA-125 reduced to levels less than 3.2 ηg/ml and 15 IU/ml, respectively. Last course of chemotherapy was received by the patient after AFP became negative and she was under observation and in good health during follow up period.

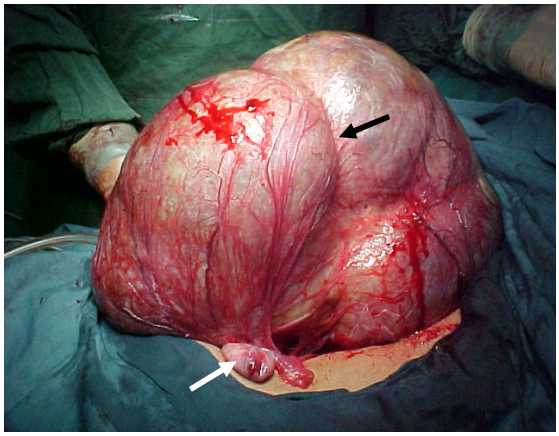

Fig. 3. Tumor adhesion to the left ovary (white arrow, left ovary; black arrow, mass).

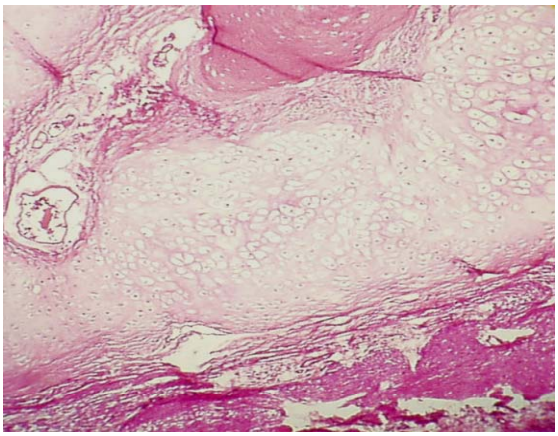

Fig. 4. Photomicrograph shows the teratomatous component composed of bone, cartilage and glial tissue (× 40).

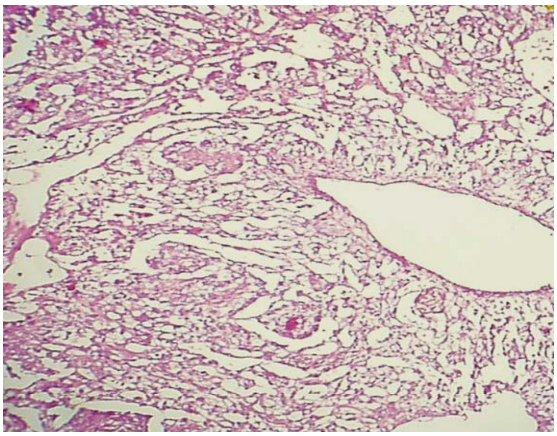

Fig. 5. The yolk sac component of the tumor with microcystic appearance and multiple Schiller Duval bodies (× 40).

DISCUSSION

Different variables make report of this case an important issue. First, it was a rare case of mixed germ cell tumor; second, its location was unusual and third, our patient was virgin and preserving fertility after surgery was an important aspect of treatment, which could not be achieved unless we preserved both ovaries and adnexa.

Most of mixed germ cell tumors consist of dysgerminoma accompanied by endodermal sinus tumor, choriocarcinoma or immature teratoma (1). Cases of mixed immature teratoma and endodermal sinus tumor are rare and reported just in a few case reports (3). In this case and in spite of just 5% of endodermal sinus tumor component of the mass, we detected high levels of AFP prior to surgery, which was due to high rate of secretion of this tumor marker by minimal amount of this component (7, 8).

Extra-ovarian germ cell tumors have been reported in all regions, from pineal body to coccygeal region (5, 6). Locations such as intracranial (9), intracardiac (10), intrapulmonary (11), retroperitoneal (12), vaginal (13) and sacrococcygeal (14) have been reported. This case had a rare localization (bridged between two ovaries). Description of unusual localization of germ cell tumors is displacement or arrest of migration of primary germ cells in their route to their proper state. During embryogenesis, primary cells of gonadal ridge extending from cranial cavity to external genitalia and remnants of these cells in this route of migration can be a place for future development of germ cell tumors (15).

Minimal surgery in ovarian germ cell tumor is unilateral oophorectomy or salpingo-oophorectomy and surgical staging. If conservation of fertility is an issue of concern, preservation of contra-lateral ovary and uterus must be considered as well. Even in the presence of metastatic disease, due to high sensitivity of these tumors to chemotherapy, preserving fertility would be recommended. Hysterectomy and bilateral salpingo-oophorectomy do not seem to change the outcome (16). In surgical stage I, FIGO stage I and grade I of immature teratoma or dysgerminoma, there will be no need for chemotherapy but in cases with higher stages, higher grades of immature teratoma or higher stages of dysgerminoma and in all patients with endodermal sinus tumors or mixed germ cell tumors, systemic chemotherapy after surgery is mandatory (17).

Recommended chemotherapy is cis-platinum containing combination chemotherapy such as BEP. Usually 3 to 4 courses of chemotherapy should be performed and in mixed tumors or endodermal ones another additional course, after negative result of tumor marker tests, should be performed (16).

In this particular case, patient underwent maximal fertility sparing surgery and ovaries, tubes and uterus were preserved for future fertility. After surgery both ovaries were of the same size.

In conclusion mixed germ cell tumors could have unusual localization and could be treated by conservative management.

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