Twisted mixed germ cell tumor of the ovary in a child

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ABSTRACT

We report an 8-year-old female patient with a mixed germ cell stromal tumor (MGCT) of the left ovary. Exploration revealed that the tumor had been twisted around its pedicle. These ovarian tumors are classified among interesting type of tumors due to the variability of neoplastic tissues in the same tumor mass, in the twisted ovarian tumor. Four histological types of tumor tissues were found: well-differentiated adenocarcinoma, dysgerminoma, immature teratoma, and focus of papillary serous cyst adenoma as well. During our review of the literature, this reported case of MGCT of the ovary was among the extremely rare cases, which had been reported. Mixed germ cell tumor is well documented in the literature since 1950.

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The germ cells first appears in the embryo in the wall of the yolk sac, from there they migrate to genital ridge on the posterior abdominal wall and incorporated into the developing gonads which later descends into the pelvis or the scrotum.[4,6] The primitive gonad is neither ovary nor testicle, but may develop into either; the origin of the tumor has long been a matter of dispute.[3,4,6] Two very rare ovarian tumors are composed of a mixture of germ cells and sex cord-stromal elements. These are the gonadoblastoma, and the mixed germ cells sex cord-stromal tumors (MGCT).[5,7,8] These 2 tumors are included in the classification of ovarian tumors as a separate category. The youngest patient ovarian tumors in general are divided histologically into epithelial, germ cell, and stromal malignancies.[9] In ordinary ovarian tumors the median age at diagnosis of ovarian tumors is 61% and the over all 5-year survival rate is 37%, while in mixed germ-cell-sex-cord is usually occurs among infants and young adults, on record was 6 weeks and the oldest 38 years,[3,6,9] with excellent prognosis after surgical intervention. Although the etiology of ovarian cancer is uncertain, approximately 5% of patients with ovarian tumors come from families where one or more first degree relatives also have the disease.[4,7] Early lesions are largely asymptomatic, and advanced tumors may produce only nonspecific symptoms such as abdominal distention, and vague gastrointestinal pains, others presented by their complications such as twisting, rupture, with acute abdominal presentations.[6,8,10]

Our aim in reporting this case due to its extreme rarity and the variability of tissues in the same tumor mass, more than one type of germ cell tissues could be detected like dysgerminomas, immature teratoma, serous-cyst adenoma, and adenocarcinoma.

Case Report. An 8-year-old female student patient, transferred to Emergency Hospital, Erbil province on April 2001, with repeated episodes

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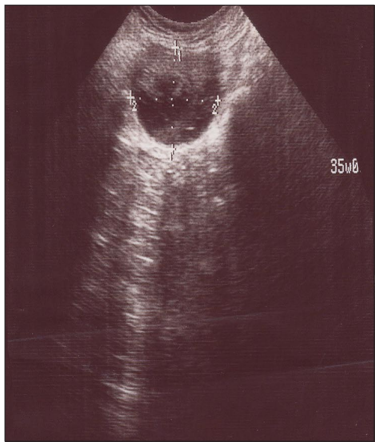

Figure 1 - Ultrasonographic picture with mixed echogenicity of the pelvic mass.

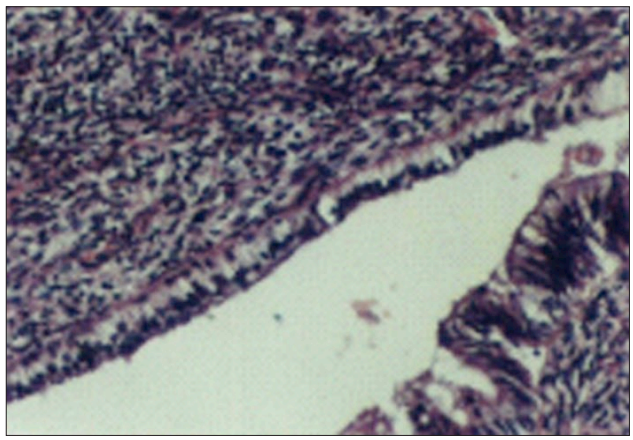

Figure 3 - Well-differentiated adenocarcinoma (X40).

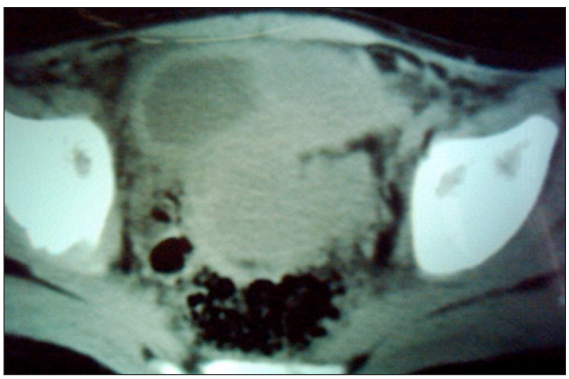

Figure 2 - Computed tomography scanning of the pelvic mass showing the pelvic mass between the rectum and urinary bladder.

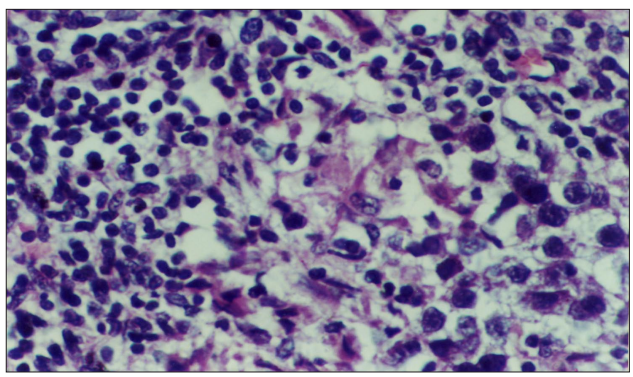

Figure 4 - Dysgerminoma among the contents of the mass. (X40).

of colicky abdominal pain, nausea, vomiting and abdominal distention, of 48 hours duration. On examination; she was extremely ill, markedly dehydration, with low blood pressure, generalized abdominal tenderness and distention, with a palpable large tender, firm, and lobulated mass on the left side of the abdomen. Her blood pressure was 65/40 mm Hg, pulse rate 120/min, hemoglobin 9.3 gm/d, white blood cell 12500/cmm, erythrocyte sedimentation rate 90 mm/h, x-ray of the abdomen showed marked gaseous distention with calcified shadows on the left side of the abdomen. Ultra-sonography and computed tomography scanning of the abdomen revealed a mixed echogenic mass (15 x 13 x 10 cm) (**Figures 1 & 2**), arising from the left adnexa with areas of cystic changes inside the mass. Resuscitation had been started immediately, however, despite good rehydration and other supportive measures, her condition reminds unstable, after 4 hours from her admission; a surgical exploration of the abdomen was performed. Operative finding showed severe distention of the small bowel loops, moderate amount of blood stained ascites, with a large unilateral, lobulated tumor mass, firm, well encapsulated, twisted around its pedicle, not ruptured arising from the left ovary. Left salpingo-oophorectomy including the tumor was performed; the cut surface of the tumor showed areas of cystic degeneration, uniform in color, from gray white to light tan. The ascetic fluid analysis was negative for malignant cells (the presence of ascites does not affect staging unless malignant cells are present).[9,10] Her satisfactory smooth recovery was complete on the 5th postoperative day and remained well, discharged on the 7th postoperative day in good condition.

Microscopic features of the tumor consisted of: 1) trabeculae, columns of cells separated by connective tissue varying from loose and edematous and hyalinized, 2) solid "tubules" with no lumen surrounded by fine connective tissues, and 3) sheets of intermixed germ cells and sex cord derivatives

showing no particular arrangement. All these patterns were mixed in the tumor; the tumor tissues were invaded with wide areas of dysgerminomas in addition to foci of immature teratoma and well-differentiated adenocarcinoma (**Figure 3**), the teratomatous foci were also invaded by sheets of dysgerminoma (**Figure 4**). The tumor had also a focus of papillary serous cyst adenoma. The diagnosis of mixed germ cell tumor of the ovary was made on the bases of these histological findings.

Discussion. Mixed germ cell sex cord stromal tumors are seen in phenotypically and genotypically normal females, mainly infants and young adults. The most common clinical presentation is an abdominal mass; iso-sexual precocious puberty has been reported.[11,12] Mixed germ cell sex cord stromal tumors can be differentiated usually from gonadoblastoma by their gross appearance, while mixed germ cell tumors are large, unilateral and lobulated, with firm consistency. However, the gonadoblastoma are small tumors, rarely exceeding 4 cm in diameter and so are seldom palpable, spicules of calcification may be obvious and more than one tumor swelling present in one side. The germ cell tumor are characterized by a diagnostic problem and controversies, their common origin from embryonic remnants and in some instance marked by sex hormone disturbances, usually originated from the primitive mesenchyme of the ovary.[2,3] The germ cells as mentioned above appears in the embryo in the wall of the yolk sac, from there they migrate to genital ridge on the posterior abdominal wall and incorporated into the developing gonads, which later descends into the pelvis or the scrotum.[5] These tumors usually arises in children and young adolescent, but may occur in adults, occasionally occurs in women in the first 3 decades of life, as a rule the patient is normal sexually, the tumor grows rapidly, producing symptoms of distention and abdominal fullness.[1-3] Torsion around its pedicle may occur, like in our reported case producing clinical presentations of an acute abdomen, most of these tumors are unilateral.[5,6] Among the important tissue contents of these types of tumors is dysgerminoma, associated with[2,3] elevated levels of human chorio-gonadotrophic hormone or lactate dehydrogenase.[4,6,12] The presence of ascites does not affect staging unless malignant cells are present in the staging of these types of tumors.[9,10] The microscopic appearance in these tumors is confusingly varied, and as different parts of the tumor may differ in structures, it is important to cut a number of blocks.[2,3] Treatment is by removal of the tumor or unilateral salpingo-oophorectomy and the prognosis is good, even in the very rare examples that have been associated with multi original malignancies, as it already happened in our case.[7,8,9] According to the staging of MJCT by The Federation Internationale de Gynecologie et d, Obstetrique and the American Joint Committee on Cancer, the tumor was stage IA because it was limited to I ovary; the capsule was intact, and no tumor was present on the ovary surface, and no malignant cells were present in the peritoneal washing.[5,6,9] Standard treatment options in the management and follow-up of the patient was according to the following protocol.[8] For patients with stage I germ cell tumors, unilateral salpingo-oophorectomy should be performed when fertility is to be preserved. For all tumors other than pure dysgerminoma and low grade (grade I) immature teratoma, chemotherapy is usually given postoperatively, reserving chemotherapy for cases in which post-surgery recurrence is documented.[5,8,10] There is considerable experience with VAC, a combination of vincristine, dactinomycin, and cyclophosphamide given in an adjuvant setting, however, combination containing cisplatin, etoposide, and bleomycin (BEP) are now preferred due to a lower relapse rate and shorter treatment time.[3,6,9] While a prospective comparison of VAC versus BEP has not been performed, it should be noted that in well-staged patients with completely respected tumors, relapse is essentially unheard-of following platinum-based chemotherapy. However, the disease will recur in approximately 25% of well-staged patients treated with 6 months of VAC.[9,10] Follow up of the patient for 2.5 years was excellent with no evidence of local recurrence or distant metastasis.

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