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Case Report

Misdiagnosis of a Case of Abdominal Granulosa Cell Tumor: A Case Report and Review of the Literature

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Abstract

Granulosa cell tumors (GCT) are the most common malignant sex cord–stromal tumors of the ovary. Extraovarian GCT is a very rare tumor, with symptom similar to GCT. It is assumed to arise from the ectopic gonadal tissue along the embryonal route of the genital ridge and is usually seen in children and adolescents. We present a case of 68-year-old female with abdominal granulosa cell tumor. Pre-operative sonography and magnetic resonance image (MRI) revealed an 8.9m*9.1cm*8.2cm lesion occupying the right pelvic. Exploratory laparotomy showed a mass arising from the right rectus abdominis protruding to the pelvic cavity, without breaking through the peritoneal. Surgical excision and the immuno-histopathology of excised masses showed features of adult-type granulosa cell tumor (GCT). The patient was followed up for 5 years and there has been no recurrence so far. Although ultrasonography and computed tomography help to attain a preoperative diagnosis, determining the exact location of intra-abdominal masses can be a difficult task at times. The case is presented for its rarity and preoperative misdiagnosis.

Keywords: Extraovarian granulosa cell tumor; Rectus abdominis; Misdiagnosis

Introduction

The ovarian granulosa cell tumor (GCT) comprises 2-5% of all ovarian tumors and over 70% of germ cell tumors originating from sex cord-stromal cells [1]. Histopathological examination can divide GCT into two subtypes: an adult type GCT that is typically found in older women, and a juvenile type GCT that is recognized primarily in children and young adults. GCT that is developing at extraovarian is called extraovarian granulosa cell tumor (EGCT), which is even rarer than the primary ovarian type. Until now, only a few of cases had been reported. The localization is including broad ligament, retroperitoneum, fallopian tube, adrenal gland, preperitoneal and mesenteric [1-14]. As far as we're able to discern, this case we present is the first case of rectus abdominis original extraovarian GCT.

Case Presentation

A 68-year-old woman, G2P2, presented with postmenopausal bleeding over last 3 months. She underwent a right oophorocystectomy 15 years ago, and the result of pathology was benign ovarian tumor from an undetails information. The pelvic examination revealed a normal size and shape of the uterus and the left ovary, while the right pelvic mass was found, which had poor activity. Transvaginal and transabdominal sonography showed an endometrial thickness measuring 1.4cm, with slightly enhanced echo and a number of small non echo area, which had a clear boundary with the myometrium. A complex mass was found arising from the right pelvic, and the bilateral ovary were unclear. MRI showed an 8.9m*9.1cm*8.2 cm sized solid-cystic mass adjacent

to the right adnexa and having a unclear boundary with bladder which was pressed by the mass and moved backward. Besides, endometrium was thick (Figure 1).

The results of tumor markers including CA125, CA19-9, AFP and CEA were normal. Cervical cytology was within normal limits. Endometrial curettage revealed proliferative endometrium with a small number of free exudates.

The patient underwent pelvic and peritoneal exploration for a pelvic malignancy. In the operation, we found a large solid-cystic tumor protruding from right lower quadrant abdominal wall into the pelvic cavity, without breaking through the peritoneal, which felt like a musculus rectus abdominis origin tumor. No pathological findings related to the uterus or both ovaries were observed. So, an abdominal wall tumor resection was performed.

Externally, the mass tumor had multiple nodules and wasn't encapsulated, being covered with some fat and muscle. The cut surface was composed of pale yellowish grey soft tissue and was cystic with necrosis.

Macroscopic examination revealed that the histological piece was consisted of sheets, and different densities of tumor cells with small, distinctive gland-like structures, enlarged nucleus, nucleolus and partial nuclear grooves were visible. Call-Exner bodies were not noted, while hemorrhage and cystic degeneration were observed (Figure 2). The tumor cells were immunoreactive for Vimentin, SMA, PR and partially reactive for α-inhibin. The tumor cells were negative for AE1/AE3, CD10, Desmin, ER, CK7, EAM stanings, and Ki-67 index was about 20%. Combined with pathological and immunohistochemical results, we consider it adult-type GCT. The patient was followed up for 5 years without chemotherapy and there has been no recurrence so far.

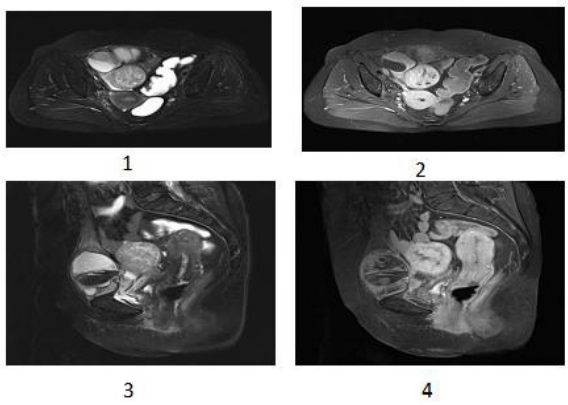

Figure 1: 1 is T2W1 axial view and 3 T2WI sagittal view. They show a huge mixed mass in right front of uterine, of which round solid part having slightly high signal, the cystic part having mixed signal.
The mass has a closely relationship with abdominal rectus abdominis, and a clear boundary with uterine.
2 is enhanced axial view and 4 enhanced sagittal view. They show that the solid part of mass has marked enhancement, the wall or separation of cystic part has inhomogeneous enhancement.

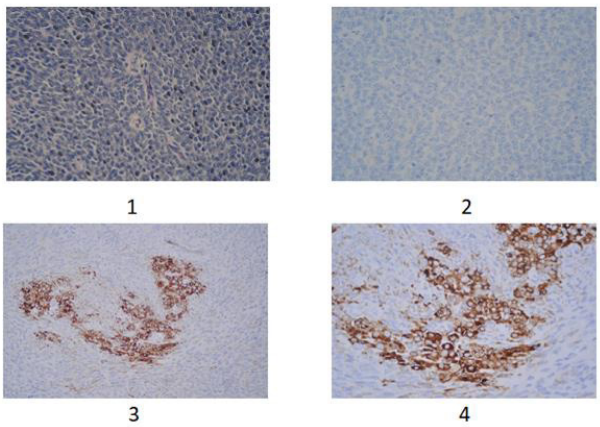

Figure 2:
1: Small, distinctive, gland- like structures filled with an acidophilic material recalling immature follicles (Call- Exner bodies)×200 HE
2: Characteristic coffee bean cells as seen, with microfollicular pattern showing pale oval nuclei and nuclear grooves×200 HE
3: Microphotograph showing a positive reaction to α-inhibin immunostain (×200)
4: Microphotograph showing a positive reaction to α-inhibin immunostain (×400)

Discussion

GCT is an uncommon sex-cord stromal tumor. Extraovarian GCT is even rarer, which was first reported in 1938 by Voigt WW [6]. Up to now, only 15 cases have been publicly reported in the English journals. Among them, 5 cases arose in the broad ligament [1-5], 3 in the retroperitoneum [6-8], 2 in the fallopian tube [9,10], 2 in the adrenal gland [11,12], 2 in the preperitoneam [13,14], and 1 was in the mesenteric [15]. The age of patients ranged from 22 to 67 years old (mean 47.8). Clinically, 7 of them were associated with estrogen-stimulating symptoms, such as postmenopausal bleeding, irregular menstruation, and endometrial hyperplasia [2,3,7,11-14]. Some of them presented with abdominal pain, the others did not have any symptoms and just found the tumor accidently. Torsion occurred in one case origin from fallopian tube [9]. Hemoperitoneam occurred due to the rupture of tumor origin from the mesenteric in another case [15]. According to the symptom of postmenopausal bleeding and the thickened endometrium, we believed that increased estrogen level does exist in this patient, although we did not detect the level of estrogen regretfully. There is no unified treatment for extraovarian GCT. The cases reported so far adopted surgery treatment without exception. The operation mode is different according to the location of the lesion and the age of the patient. Postoperative recurrence occurred only in 1 case [4].

The pathogenesis of ovarian GCT is not clear yet. Supernumerary ovaries have provided evidence of formation of ovarian sex-cord stroma at extragonadal sites [16-20]. So extraovarian GCT could theoretically occur at any site as the primordial germ cell can migrate anywhere during the embryonic life. And patients with history of oophorectomy have the potential to suffer from extraovarian GCT [13]. This patient we presented had accepted right oophorocystectomy with a negative histology 15 years ago. It suggested that the mass is not a metastatic granular cell tumor but an extraovarian GCT. So far, there has been no rectus abdominis origin extraovarian GCT reported, it is the first case. We presumed that the mass was a malignant tumor of ovary before the operation, considering that MRI and sonography showed the mass convex to the pelvic cavity and seems contacting with the ovary closely. The preoperative diagnosis was inconsistent with the results of surgical exploration. The patient`s history of oophorocystectomy proved the tumor may be derived from ectopic ovarian tissue indirectly.

In future, we should pay attention to preoperative evaluation of the extraovarian GCT. When the mass fixed in the adnexal with poor activity, we should consider it may be a tumor of the abdominal wall. When the patient present with endometrial thickening, postmenopausal bleeding or irregular menstruation, we should also prospect of the tumor secreting estrogen such as GCT, in addition to exclude endometrial cancer. For there are only few of case reported, the treatment and prognosis of extraovarian GCT need to be further investigated.

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Conflict of Interest

Authors declare no conflict of interest.

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