Extra-Ovarian Granulosa Cell Tumor: A Case Report

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ABSTRACT

Extraovarian granulosa cell tumor is a very rare tumor, assumed to arise from mesenchymal tissue along the embryonal route of the genital ridge. One such rare case of extraovarian granulosa cell tumor was encountered in a 73-year-old female patient who presented with a large intraabdominal mass. Computerized tomography revealed a large retroperitoneal mass measuring 9 cm x 6.5 cm. Her past medical history was irrelevant. She underwent exploration laparotomy and the mass was excised. Histopathological examination of the excised mass showed features of adult-type granulosa cell tumor. Because metastatic epithelial tumors, particularly from the ovaries, may show identical morphology, immunostains for inhibin and Epithelial Membrane Antigen (EMA) were performed. The tumor showed positivity for inhibin while epithelial membrane antigen was negative thus confirming the diagnosis of granulosa cell tumor. A diagnosis of extraovarian granulosa cell tumor can only be done after excluding any previous history of granulosa cell tumor of the ovary. Immunostains help to differentiate granulosa cell tumors from other neoplasms.

Key words: Granulosa cell tumors, Retroperitoneal masses, Ovarian tumors

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Introduction

Granulosa cell tumor (GCT) of the ovary is a rare tumor representing 2-5% of all ovarian neoplasms.[1] It can recur or metastasize many years after initial treatment. Rarely, GCT can develop at an extraovarian site; we report one such rare case of extraovarian GCT in a 73-year-old female patient. Primary extraovarian GCT is an extremely rare tumor.[2] Until 2013, only eleven cases of such tumors have been reported in the English literature.[2-4] Ours is the twelfth case and the first case in Jordan.

Case report

A 73 year old female patient was admitted to Queen Alia Hospital, Amman, Jordan, with six a week history of a localized left upper quadrant abdominal pain, dull aching in nature and associated with occasional bouts of vomiting and a history of non documented weight loss. Her past medical history was irrelevant. Abdominal examination revealed a palpable and tender intra-abdominal mass in the left upper abdominal quadrant; it was about 9.0X8.0cm in its greatest diameter. The laboratory investigations including hematocrit, leukocytes, platelets, kidney function tests, electrolytes and liver function tests were within normal range.

Computerized tomography (CT) of the abdomen revealed a 9.0X 6.5 cm sized, large, well defined complex, irregular mass with enhanced wall, located at left paracolic gutter, bounded anteromedially by descending colon, laterally by abdominal wall and posteriorly by the left kidney as shown in Fig. 1. The patient underwent exploratory laparotomy with the presumptive

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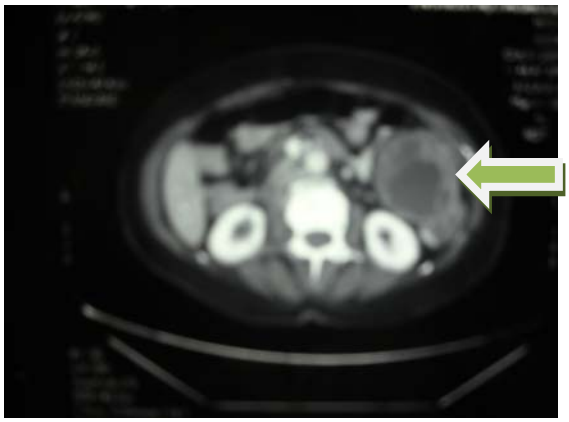

Fig. 1: Abdominal CT scan, shows the left upper quadrant abdominal cystic mass (pointed by arrow)

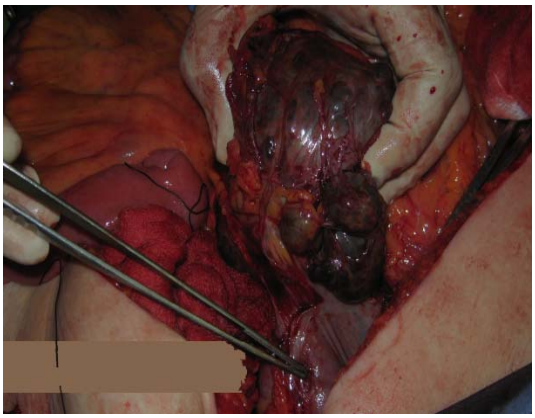

Fig. 2: An explarotory laparotomy showing the retroperitoneal mass attached to the splenic flexure

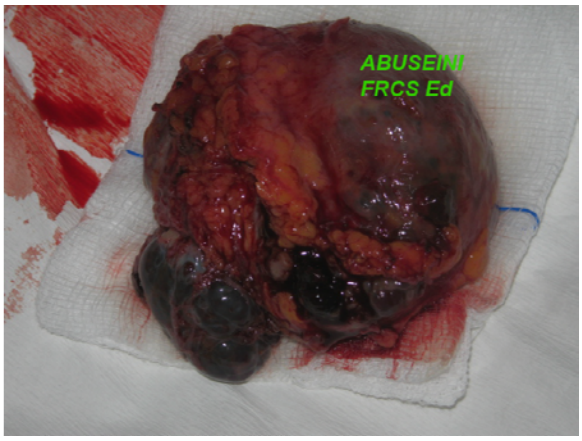

Fig. 3: The excised multiloculated cystic mass.

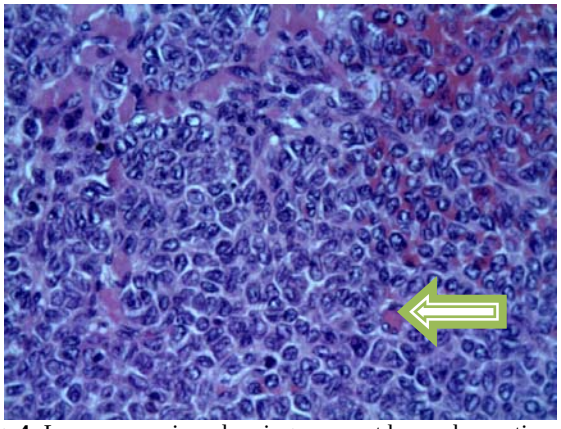

Fig. 4: Low power view showing compact hyperchromatic cells with scanty cytoplasm, high N/C ratio microfollicular pattern showing pale oval nuclei & nuclear grooves. Call-exner bodies (pointed by arrow). The mitotic index is low.

diagnosis of intra-abdominal hydatid cyst. The intraoperative findings revealed a large bulging cystic tumor arising from the retroperitoneal space posterolateral to the descending colon, inferior to the spleen and anterior to the left kidney and attached to the splenic flexure by a highly vascularized tissue band, giving the impression of a retroperitoneal sarcoma as shown in Fig. 2. Externally, it was grayish red and smooth with a row of adipose and fibrous tissue tags along one surface. The mass was dissected free and excised (Fig. 3). The patient had an uncomplicated postoperative recovery course and she was free of disease six months later.

Grossly, the mass was grayish brown in color, soft and cystic with hemorrhagic area measuring 8X9 cm. Cut section revealed solid homogenous grayish tumor with multiple small cystic areas and area of hemorrhage. Microscopic finding, showed small round to oval neoplastic cells with diffuse and trabecular patterns. The cells showed scanty cytoplasm and round to oval nuclei with nuclear grooves and Call-exner bodies (Fig. 4). With this typical histopathological feature, a diagnosis of extrovarian GCT was made. The tumor was positive for inhibin and negative for EMA, confirming the diagnosis extraovarian GCT.

Discussion

GCT is a rare tumor with only eleven cases reported in the English literature.[4] It can develop in the retroperitoneum,[3,5] broad ligament mesentry, omentum, liver or adrenals. Histogenic origin is thought to be from ectopic gonadal tissues from the meson ephros. [3] This case represents the first extrovarian GCT in Jordan. It was difficult to distinguish from several other histologically similar neoplasm, including undifferentiated carcinoma, small cell or neuroendocrine carcinoma, endometrial stromal sarcoma, thecoma, carcinoid, and malignant

melanoma.[6] There are varieties of techniques used to confirm the diagnosis of GCT and exclude alternatives, such as the use of reticulin stain and antibodies to inhibin, S-100, synaptophysin, chromogranin, pankeratin, estrogen receptor and progestron receptor.[7]

In the present case, the patient presented with left sided abdominal mass, associated with abdominal pain, the hormonal studies were not done, as the diagnosis of GCT was not suspected. The histopathology features of the tumor are typical of GCT, small pale, round oval nuclei with nuclear groove, and the tumor showed positivity for inhibin.

This case is reported for its rarity, and difficulty of diagnosis, and to describe its relevance to the histogenic origin. The diagnosis is made by characteristic histological features. Immunostains like inhibin help in definitive diagnosis.

Conclusion

GCT of the ovary is a rare tumor, and the extra-ovarian GCT is even more rare and can present as a retroperitoneal mass. Collaboration between the surgeon and the pathologist confirms its diagnosis.

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