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Ovarian Fibrothecoma: Presented as a Large Pelvic Mass

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ABSTRACT

Fibrothecomas are benign sex cord-stromal ovarian tumors. We present here with a rare case of large fibrothecoma of ovary. A 75 year female presented with complaints of pain abdomen, distension and irregular per vaginal bleeding. On radio imaging reported as solid cystic ovarian mass suggestive of neoplasm? ovarian cystadenoma? ovarian fibrothecoma. Her serum CA 125 was normal. The exploratory laparotomy was done. The excision of left ovarian tumor with total hysterectomy with bilateral salpingo-oophorectomy with right oophorectomy was performed. On gross examination large left ovarian mass measuring 24 x 19 x 8 cm and weighing 950 gm was noted. The histopathological diagnosis was fibrothecoma with large simple cyst of left ovarian mass. We are presenting this case of aggressive behavior of fibrothecoma for it clinical, radiological and histopathological findings.

Keywords: Fibrothecoma, Ovarian tumor, Sex cord-stromal tumors.

INTRODUCTION

The sex cord-stromal tumors are a distinct group of ovarian tumors representing about 1%-4.7% of all ovarian neoplasms. [1,2,3] The fibroma / thecoma / fibrothecoma is a spectrum of benign sex cord-stromal tumors. The tumor is composed of fibroblastic stromal cells and/or luteinized theca-like cells. They are categorised as Ovarian Thecoma-Fibroma Groups (OTFG). [4] They are uncommon sex cord-stromal neoplasms and mostly are of solid tumors of the ovary. We are presenting this case of aggressive behavior of fibrothecoma for it clinical, radiological and histopathological findings.

CASE REPORT

A case of 75 year female presented with complaints of rapidly increasing pain abdomen, distension, irregular per vaginal bleeding of 4 month duration. There was no family history of malignancy. The systemic examination was normal. Her serum CA 125 was normal (31.2U/mL). An ultrasound showed left ovarian heterogeneous solid hypoechoic mass measuring 18.6 x 12 x 9.2 cm with large cyst attached to it 12.5 cm in diameter. On USG reported as large ovarian cyst suggestive of cystadenoma of left ovary. On MRI reported as solid cystic ovarian mass suggestive of neoplasm? ovarian cystadenoma? ovarian fibrothecoma. The imaging by computed tomography (CT) scan showed a well-defined heterogeneous lesion noted at left

ovary measuring about 19×11×10 cm. suggestive of ovarian neoplasm. The cytology of ascitic fluid was negative for malignancy.

The exploratory laparotomy proceeded to excision of left ovarian mass with total hysterectomy with bilateral salpingo-oophorectomy with right oophorectomy was performed. We received specimen of large left ovarian Mass (Figure:1) measuring 24 x 19 x 8 cm and weighing 950 gm. Cut section shows a large grey white to yellowish encapsulated solid mass measuring 14.5 x 11.2 x 8 cm (Figure:2) along with cyst measuring 12.5 cm in maximum diameter filled with clear fluid. The external surface of ovary was smooth. Uterus with cervix, bilateral fallopian tubes and right ovary were unremarkable. On cut section endometrial canal measured 4 cm in length with increased endometrial thickness at places. The myometrium was thickened. The right ovary measured 3.2 x 2.3 x 2 cm and cut section shows multiple cysts, largest cyst measuring 1 cm in maximum diameter.

On microscopic examination, multiple sections showed neoplastic cells composed of spindle stromal cells arranged in storiform, fascicular pattern and in sheaths. The cells were monomorphic with central nucleus. No mitotic activity could be identified. The theca cells were oval or rounded with moderate clear/vacuolated cytoplasm. The nuclei were round to oval and exhibit no atypia. The fibromatous component could be seen separating the sheets and nests of theca cells. (Figure:3,4) The histopathological diagnosis was fibrothecoma with large simple cyst of left ovarian mass. Other findings were chronic non specific cervicitis, nabothian cyst, squamous metaplasia of cervix. Also noted cystic endometrial hyperplasia with areas of Endometrial Intraepithelial Neoplasia (EIN) of endometrium. The focus of adenomyosis was noted. Right ovary showed simple cyst. The bilateral fallopian tubes were unremarkable. There was no evidence of tumor in the omentum.

Postoperative course was uneventful. The patient is well and asymptomatic after surgery and advised follow up.

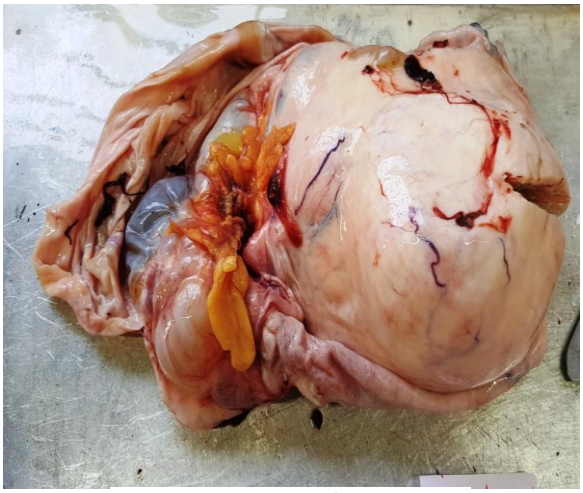

Figure:1- an excised left ovarian mass

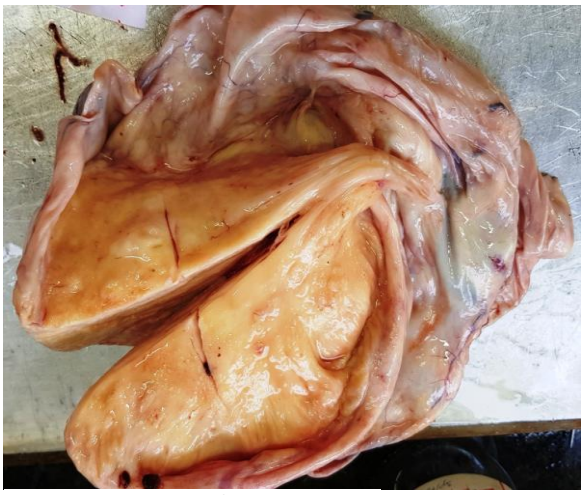

Figure:2- cut open left ovarian mass showing a large grey white to yellowish encapsulated solid , firm mass.

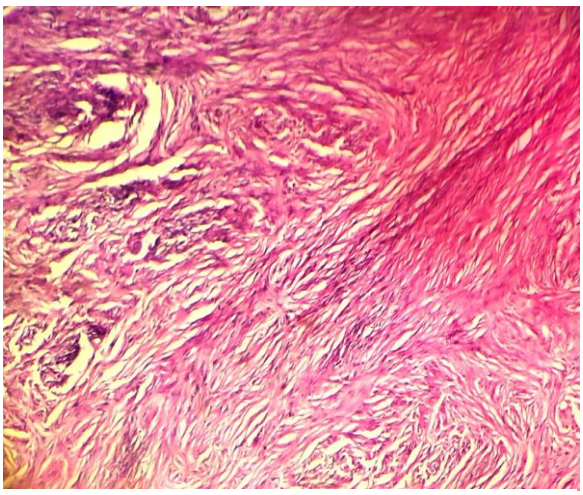

Figure:3- photomicrograph showing neoplastic cells composed of spindle stromal cells arranged in storiform ,fascicular pattern and in sheaths.(H & E Stain,40x)

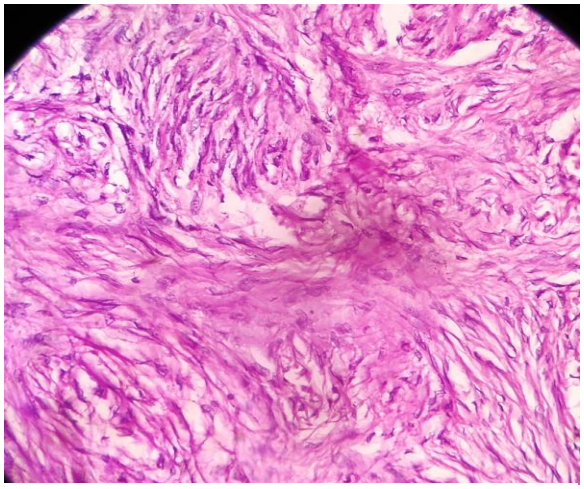

Figure:4- photomicrograph showing neoplastic cells composed of spindle stromal cells with theca cells.(H & E Stain,100x)

DISCUSSION

The ovarian fibrothecomas are benign sex cord-stromal tumors of the ovary, accounting for 1%–4.7% of all ovarian neoplasms.

The fibrothecomas occurs in a wide range, with an average patient age around 50 year. These tumors can occur at any age, but most commonly arise in middle-aged to postmenopausal women. [5] They are not hormonally active in most cases. Most of them are unilateral; however, bilateral cases may occur, especially in patients with Gorlin syndrome. They most commonly present as unilateral pelvic mass. The clinical presentation may be nonspecific such as pelvic and abdominal pain or distension. In our case presented with complaints pain abdomen, distension, irregular per vaginal bleeding

Ovarian fibromas can be part of Meigs' syndrome, a triad of ovarian fibroma, ascites, and pleural effusion, which disappears spontaneously after the tumor is removed. Large tumors were often associated with torsion, hemorrhage, calcification, or complicated with other cystic lesions. In our case tumor was large however there was no any secondary complication noted. Torsion occurs in about 8% of the patients. The fibrothecoma are rarely malignant. [7] Patients may also present with endocrine manifestations such as estrogenic effects with abnormal vaginal bleeding, endometrial hyperplasia or endometrial carcinoma. [8] In our case endometrium showed hyperplasia with areas of Endometrial Intraepithelial Neoplasia. They less commonly shows androgenic effect related to hormonally active thecoma elements. Luteinized the comas may be functional which secrete androgen and causes hirsutism or virilization.

Ovarian fibroma is often difficult to diagnose preoperatively on clinical and radio imaging. [9] Important radiologically because appears as a solid mass, mimicking malignancy. They are usually misdiagnosed as uterine liomyoma/sarcoma. On ultrasound tumor is seen as a homogeneous hypoechoic mass. About 80% of ovarian fibrothecomas appear as solid masses with a delayed accumulation of contrast medium. While on MRI these lesions shows predominantly homogeneous low signal intensity.

Fibrothecomas are usually small in size, it ranges from 1-10cm in diameter. They are bilateral in 5 to 10 % of cases. Grossly, fibrothecomas are usually round, oval or lobulated encapsulated hard gray white masses covered by intact ovarian serosa. Edema and cystic degeneration are relatively common. The fibrothecoma of more than 10cm tend to be associated with myxoid change or degeneration. In our case tumor was large (14.5 cm) but it does not showed any secondary changes.

On histopatholgy these tumors are characterized by the presence of spindle, oval, or round cells having nuclei which are bland, wavy and fusiform. Also contain theca cells composed of tumor cells with abundant pale or vacuolated cytoplasm. The intersecting bundles of spindle cells, collagen, and hyalinized tissue are seen. Tumor cells having nuclei which are bland, wavy and fusiform with whorled appearance resembling uterine leiomyoma. It is also important to differentiate them from broad ligament leiomyoma. [11] Other differential diagnosis includes fibrosarcoma or recurrent STUMP.

There was no specific tumor biomarker can be identified in the majority of ovarian fibrothecomas. Fibrous tumors of ovary are immunoreactive to vimentin and calretinin and typically stain positive for CD10 and inhibin. Newer markers such as FOXL2, steroidogenic factor 1 and CD56 are often positive in fibromas which differentiate from leiomyoma. [11]

Early diagnosis and surgical resection is the treatment of choice for ovarian fibrothecomas. The modality of treatment could be tumor excision alone, uni- or bilateral salpingo-oophorectomy with or without hysterectomy depending on the patient status and the aggressiveness of the tumor. [1] Most cases are benign and surgical resection is curative, effective and having good prognosis.

CONCLUSION

The ovarian fibrothecomas are uncommon benign solid tumor of the ovary. We are presenting this case of aggressive behavior of fibrothecoma for it clinical, radiological and histopathological findings. .

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