Fibrothecoma of the Ovary- A Rare Case Presentation
A Dhull, V Kaushal, S Mathur, R Agarwal

Citation

Abstract
Ovarian fibrothecomas represent an ovarian stromal neoplasm developing in a wide spectrum of clinical settings. Fibrothecomas have been described as rare ovarian neoplasms. Here we present an unusual clinical manifestation of ovarian fibrothecoma with abdominal distension in a young female, which must be differentiated from massive edema of the ovary and sclerosing stromal tumor of the ovary.

INTRODUCTION
Both thecoma and fibroma of the ovary are included in stromal tumors of the ovary. Thecomas are histologically composed of lipid-containing cells that resemble theca interna cells. Fibromas are composed entirely or almost entirely of spindle, oval, or round cells forming variable amounts of collagen. The differentiation between thecomas and fibromas is occasionally imprecise because of the histological and immunohistochemical overlap between them. Therefore, the term “fibrothecoma” has been frequently used1. Ovarian tumors of the thecoma-fibroma group have been reported to show myxoid change or degeneration2,3. Recently, we encountered a case of fibrothecoma with abdominal distension, which was difficult to distinguish from massive edema of the ovary, ovarian myxoma, and sclerosing stromal tumor. We herein report this rare case of fibrothecoma in a young female.

CASE SUMMARY
An eighteen years young female presented with abdominal distension and mild constipation of one month duration. She had lost 10% of body weight in one month. Menstrual history of the patient was normal with menarche at 13 years. General physical and systemic examination were normal. Local examination of abdomen and pelvis revealed a hard mass of 20x18 cm in midline, arising from pelvis. Ascites was present.

Complete hemogram and routine blood biochemistry of the patient were within normal limits. CA-125 was 181U/ml. Abdomino-pelvic ultrasonography revealed a large hypoechoic lesion, arising from pelvis, reaching up to the umbilicus. The mass had few cystic areas within it and blood flow on color doppler was normal. However bilateral ovaries and uterus could not be visualized due to mass lesion which was suspected to be ovarian mass. Free fluid was seen in Morrison’s pouch and pre-hepatic region. The cytology of the ascitic fluid was negative for malignancy.

The patient underwent exploratory laparotomy which revealed bilateral ovarian tumor, 16x16 cm on left side and 4x2 cm on right side. Left sided salpingoovariotomy and right sided complete removal of tumor mass was done. Histopathology of bilateral ovarian tumor revealed fibrothecoma. There were no malignant changes in the tumor. Omental and peritoneal biopsy revealed areas of congestion only. Postoperative ultrasonography of abdomen and pelvis was normal. Postoperatively patient is disease free and has been advised close follow-up.

DISCUSSION
The name fibrothecoma because the theca cells of normal ovary have more of the characteristic of connective tissue elements. Stromal tumors of the ovary include thecoma and fibroma, yet as differentiation between these two types may be difficult the term fibrothecoma has emerged in recognition of the similar immunohistochemical features present in both. The exact incidence of fibrothecoma is unknown, although they have been described as rare ovarian neoplasms1. Here we present an unusual clinical manifestation of ovarian fibrothecoma with abdominal distension in a young female. Grossly, the resected left ovarian mass measured 16x16x10 cm and right ovarian mass measured 3.5x2x0.4 cm. Cut surface of both was firm, grayish white fascicular pattern with few yellow areas (Fig.1). No normal ovarian tissue was identified.
Fibrothecomas are round, oval or lobulated solid tumors that cast stripy shadows and are associated with fluid in the pouch of Douglas, and most manifest minimal to moderate vascularization. A fibrothecoma with atypical ultrasound appearance may be mistaken for a malignancy, in particular if associated with fluid in the pouch of Douglas or ascites, high color content and raised CA 125 levels, as in the present case report.

**Figure 1**
Figure 1: Cut surface of the resected ovarian mass showing firm, grayish white fascicular pattern with few yellow areas.

**Figure 2**
Figure 2: Standard H&E stain - Low power view of fibrothecoma (10X)

On microscopy, the ovarian mass was composed of spindle stromal cells which were randomly distributed or arranged in a fascicular fashion. An appreciable amount of the stromal neoplasm was characterized by thecal cells which were oval or rounded to polyhedral with moderate to abundant pale or vacuolated cytoplasm (Fig. 2 & 3). The nuclei were round to oval and pale and exhibit little or no atypia. The fibromatous component could be seen separating the sheets and nests of theca cells. The histology using special stains with Sudan III, showed the thecal cells with abundant intracytoplasmic neutral fat and Reticulin stain demonstrated reticulin fibres surrounding individual cells arranged in a box-in-appearance. Since the differentiation between thecomas and fibromas is occasionally imprecise due to the histological overlap between them, some have used the designation “fibrothecoma” for tumors in the intermediate zone between thecoma and fibroma. The ovarian mass in the present case demonstrated both spindle cells associated with collagen bundles and lipid containing cells with a box-in-appearance which indicate fibroma and thecoma, respectively. Therefore, the histologic findings in the present case were consistent with fibrothecoma. Moreover immunohistochemical study with Vimentin was positive for more than half of the tumor cells in the cellular area.

Edema may be observed in fibrothecoma, and it can be massive edema if fibrothecoma may be induced by a stasis of lymphatic drainage, such as massive edema of the ovary. A partial intermittent torsion has been thought as a cause. Samanth et al. reported that ovarian tumors larger
than 10 cm in diameter tended to be associated with myxoid change, and insisted that a discrepancy between arterial supply and venous and lymphatic drainage could lead to stromal edema. Fibrothecoma with abdominal distension and raised CA125 must be differentiated from the edema of the ovary, ovarian myxoma and sclerosing stromal tumor. Edema of the ovary is characterized by a proliferation of ovarian stromal cells with marked intercellular edema preserving the overall structure of both the ovarian cortex and medulla, probably secondary to intermittent torsion. Edema affects young women with an average age of 22 years, whereas 84% of the patients with thecoma are postmenopausal. Fibromas occur at all ages, but are most frequent during middle age. Ovarian myxoma has recently been reported as a new distinct pathological entity that shows a myxoid, moderate cellular proliferation of spindle and stellate cells interspersed with areas of fibrosis, hemorrhage and delicate vascular spaces. Myxoid change has also been reported in fibrothecoma in up to 40% of cases. Costa et al. insisted that ovarian myxoma may be at one end of the spectrum of differentiation in the thecoma-fibroma group of tumors, because the myxoid change in fibrothecoma was indistinguishable histologically and immunohistochemically from ovarian myxoma.

Sclerosing stromal tumors are rare ovarian neoplasms occurring predominantly in young women, and their hypocellular and edematous area may be confused with edematous area of fibrothecoma. The histological features of sclerosing stromal tumor are a pseudolobular pattern of cellular areas and hypocellular areas, prominent vasculature with a hemangiopericytomatosus pattern and cellular heterogeneity of vacuolated luteinized theca-like cells and spindle-shaped fibroblast-like cells in the cellular area. In the present case these findings were not observed. We reported a rare case of ovarian fibrothecoma, which must be differentiated from massive edema of the ovary and sclerosing stromal tumor of the ovary. The age of the patient was also important. Patient has been advised for close follow up with ultrasonography and routine investigations on every follow up.

**CONCLUSION**

Ovarian fibrothecomas represent an ovarian stromal neoplasm developing in a wide spectrum of clinical settings. Particularly if oophorectomy is stated to have been performed remote from the time of index presentation, the status of the ovaries must be considered whenever pelvic pathology is encountered.

**References**

Author Information

Anil Kumar Dhull, MD
Senior Resident, Department of Radiation Oncology, Pt. BD Sharma Post Graduate Institute of Medical Sciences

Vivek Kaushal, MD, DNB
Senior Professor & Head, Department of Radiation Oncology, Pt. BD Sharma Post Graduate Institute of Medical Sciences

S.K. Mathur, MD
Senior Professor, Department of Pathology, Pt. BD Sharma Post Graduate Institute of Medical Sciences

Ruchika Agarwal, MD
Demonstrator, Department of Pathology, Pt. BD Sharma Post Graduate Institute of Medical Sciences