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Dr. Prof. Nilesh Dalal
Obstetrics & Gynaecology, Department, MGM Medical College Indore, M.P, India

Dr. Anshita Goyal
Obstetrics & Gynaecology Department, MGM Medical College Indore, M.P, India

Dr. Somen Bhattacharjee
Obstetrics & Gynaecology. Department, MGM Medical College Indore, M.P, India

Correspondence
Dr. Prof. Nilesh Dalal
Obstetrics & Gynaecology, Department, MGM Medical College Indore, M.P, India

Virilizing Ovarian Fibrothecoma with Minor sex cord elements in a 17 year old girl: A rare case report

Dr. Prof. Nilesh Dalal, Dr. Anshita Goyal and Dr. Somen Bhattacharjee

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Abstract
Ovarian fibrothecomas is a rare, benign, sex cord cell neoplasm with a typically unilateral location in the ovary, and is characterized by mixed features of both fibroma and thecoma. Here we present an unusual clinical manifestation of virilizing ovarian fibrothecoma with abdominal distension in a young female.
Ovarian masses, although rare in children, must be included in the differential diagnosis of all girls who presented with abdominal pain with distension. Any solid ovarian tumour in adolescent age group should be considered malignant unless proved otherwise by histological examination.

Keywords: sex cord stromal tumor, laparoscopy, laparotomy, ovarian fibrothecoma, ultasonography

Introduction
Adolescence is a critical period and its assessment require a sensitive and age appropriate approach.
Sex cord stromal tumors represent approximately only 8% of all ovarian neoplasms, and are composed of granulosa, theca, Sertoli and leydig cells, together with fibroblasts, singly or in combination [1]. Fibromas and thecomas may show a significant morphologic overlap, which has led to the use of the term fibrothecoma.
This kind of tumor accounts for 3-4% of all ovarian neoplasms [2]. Another subgroup, which was first described by Young and Scully, comprises fibroma/fibrothecomatous with minor (<10%) sex cord elements; it is designated as stromal tumor with minor sex cord elements.1 On examination, it contains discrete tubules or small nests of cells resembling granulosa cells, Sertoli cells, or indifferent cells of sex cord type with or without steroid hormone-cell type in the spindled stroma [3]. This group is usually hormonally inactive, but functional differentiation in producing hormones by luteinized theca cells or leydig cells determines its course [1, 2, 4].
To the best of our knowledge, only 14 cases of ovarian stromal tumor with minor sex cord elements have been documented till date, with only 3 cases of fibrothecoma with minor sex cord elements [3, 5, 6]. Even though 11% of fibrothecoma can be androgenic, no case of virilizing fibrothecoma with minor sex cord elements has been found in the previous literature [4-10].

Case Summary
A 17-year-old, unmarried, female patient presented in gynae OPD of MGMMC MYH Indore with complaints of gradualy growing abdominal lump with dull aching pain in center since 1 year, secondary amenorrhoea since 6 months, and masculinizing features of enlarged clitoris, facial hair, deep voice since 3 month.
Menstrual history of the patient was normal with menarche at 15 years.
General physical and systemic examination were normal.
Local examination of abdomen and pelvis revealed a hard mass of 20x15 cm in midline, arising from pelvis. Ascites was absent.
Complete hemogram and routine blood biochemistry of the patient were within normal limits.
Hormonal Study-shows TSH-0.61 miu/l, FSH – 2.71miu/ml, LH- 4.48 miu/ml,
Testosterone-548.12ng/dl, DHEAS-164.4ug/dl, CA-125 was 11.8U/ml.
Abdomino-pelvic USG revealed a large hypoechoic lesion, arising from pelvis, reaching up to the umbilicus. The mass is well defined, solid, with cystic lesion of 6.5x5.5 cm within it and blood flow on color doppler was normal.
Magnetic resonance imaging revealed a large ill-defined right adnexal mass of size 24x20x10x cm with eccentric solid components. Uterus and left ovary were normal.

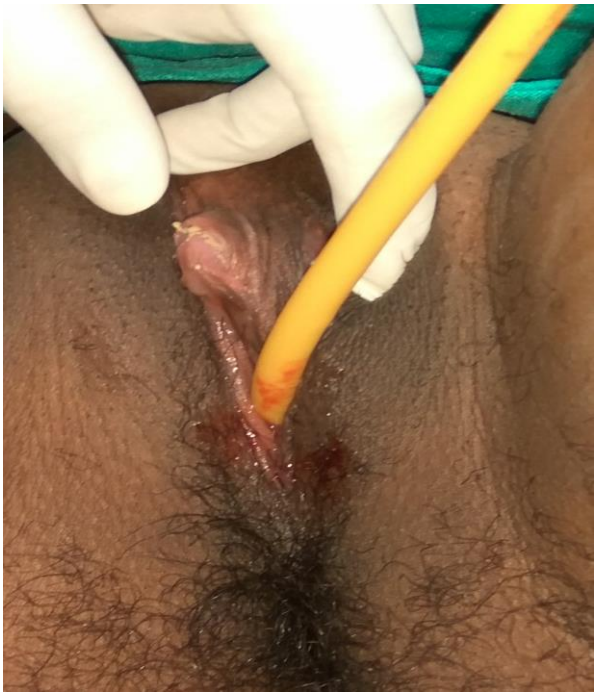

Fig 1: Clitoromegaly

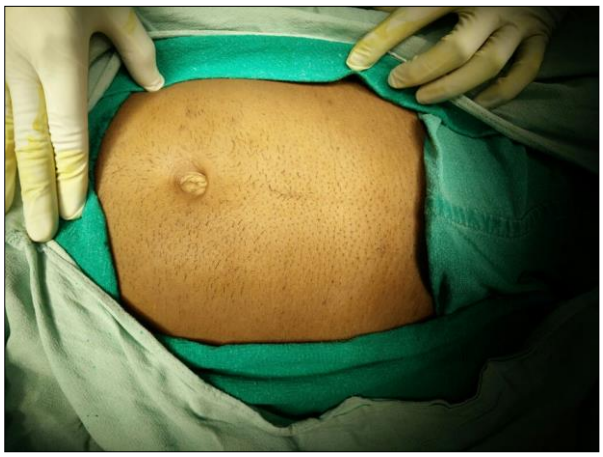

Fig 2: Male pattern abdominal hairs

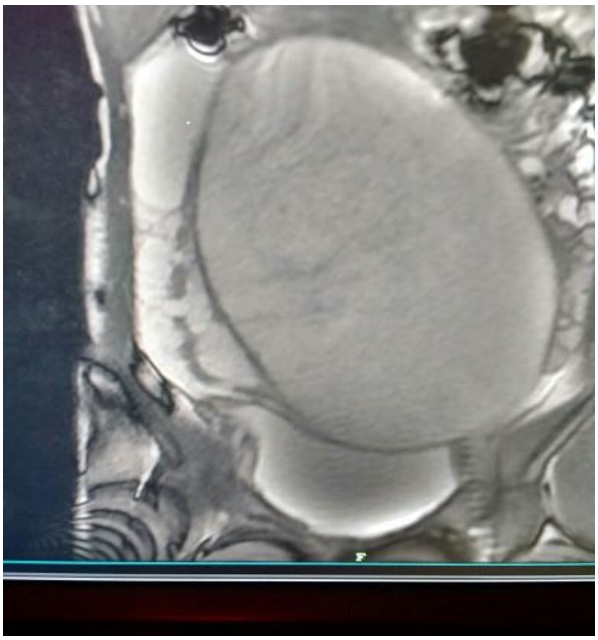

Fig 3: MRI image showing abdominal mass 24X20X10 cm

The patient underwent exploratory laparotomy which revealed left sided ovarian tumor with torsion 24x13 cm with 2 functional cyst of 7x6 cm. Left sided oophorectomy was done.

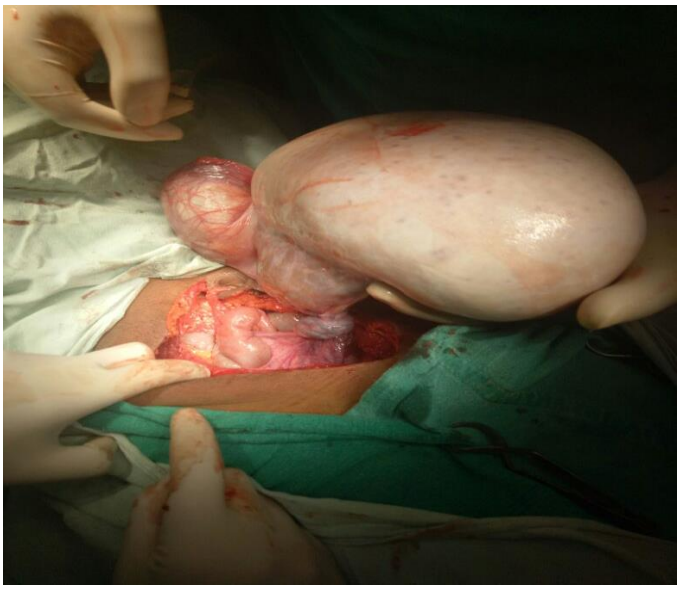

Fig 4: Intraoperative Findings

Histopathology of ovarian tumor revealed fibrothecoma. There were no malignant changes in the tumor. Omental and peritoneal biopsy revealed areas of congestion only.

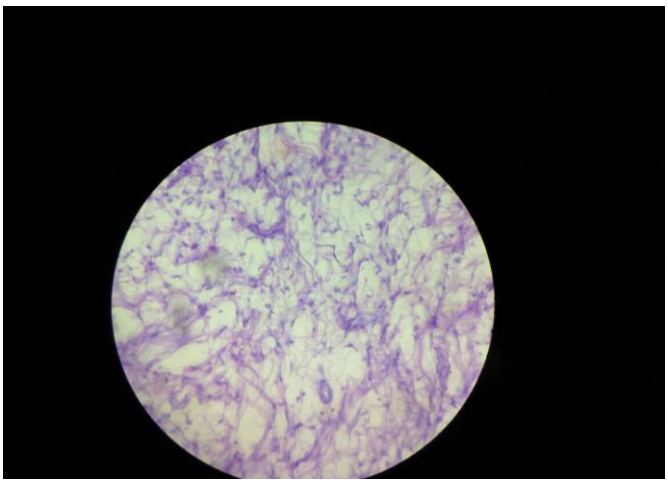

Fig 5: Histopatholgy Reporting

Postoperatively patient is disease free and has been advised close follow-up.

Discussion

Young and Scully were the first to describe ovarian stromal neoplasms, predominantly fibromatus or thecomatous tumors, containing scattered minor sex cord elements (as above) in less than 10% of the tumor area, with the age of presentation ranging from 16 to 65 years, with a mean age of 59 years [3]. These patients generally present with abdominal pain, bleeding per vaginum and adnexal masses, rarely with androgenic features [3, 8], However, in the present case, the patient was only 17 years old, much younger than usual, and she presented with secondary amenorrhoea, masculinizing features and abdominal lump. These tumors are usually hormonally inactive, but have been occasionally associated with estrogenic features, e.g. endometrial hyperplasia or diffuse complex atypical hyperplasia

or even adenocarcinoma [1, 5, 9]. In the present case, tumor was hormonally active, but with masculinizing features and with postoperative correction.

Out of seven reported cases of fibromatous tumors of the ovary by Young *et al*. [3] five cases were ovarian fibroma with minor sex cord elements. Two of them were luteinized thecomas with steroid-hormone cell types without reinke's crystalloids, as in this case, and stromal- leydig cell tumor with steroid-hormonecell type with reinke's crystalloids, but sertoliform areas were not described in these two cases unlike this case.3 Zhang *et al*. [8] reported fifty cases of ovarian stromal tumors and found steroid hormone secreting cells in a background of predominant of pattern of fibroma/thecoma. He also reported the cases of steroid cells without reinke's crystalloids, as found in the present case [8]. However, combination of sertoliform cells and steroid cells/theca cells, as in our case, have not been described by them, but only recently by Sherwani *et al*. (without virilization) [6].

Conclusion

In conclusion; this case highlights the challenges in our day to day scenario. But a proper approach, sound knowledge, diagnostic modalities (USG, MRI, Hormonal studies) have opened many ways to simplify the diagnostic evaluation of difficult cases.

Ovarian masses, although rare in children, must be included in the differential diagnosis of all girls who presented with abdominal pain with distension.

Any solid ovarian tumour in adolescent age group should be considered malignant unless proved otherwise by histological examination.

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