

BILATERAL FIBROTHECOMA OF OVARY IN A YOUNG FEMALE- A RARE ENTITY

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PRESENTATION OF CASE

16 years young unmarried girl presented with fever and abdominal pain of 5 days duration. Her menstrual history was normal with menarche at 13 yrs. General physical and systemic examination were normal.

Local examination of abdomen and pelvis revealed a hard mass of 20 x 10 cm in pelvis and lower abdomen.

- Lower component is causing mass effect on uterus, bladder.
- This mass is taking minimal flow on application of colour doppler
- Bilateral ovaries are not visualised.
- Mild ascites noted in pelvis and right, left iliac fossae.
- On screening chest, there is no pleural effusion noted.

CLINICAL DIAGNOSIS

Right Adnexal Mass-? Malignant Ovarian Tumour.

PATHOLOGICAL DISCUSSION

Blood Investigations-

| | |
|----------------------------------|------------------------|
| CEA (Carcinoembryonic Antigen) | 1.4 ng/ml {WNL} |
| CA 125 | 25.4 U/ml {WNL} |
| Sr. LDH | 165 U/L {WNL} |
| Beta-HCG | <0.5 micro IU/ml {WNL} |
| LFT | WNL |
| CBP (Complete Blood Picture) | WNL |
| CUE (Complete Urine Examination) | WNL |
| Sr. Electrolytes | WNL |
| Sr. Creatinine & Bl. Urea | WNL |

Abdomino-Pelvic Ultrasonography

A large mass lesion is noted in abdomen arising from with two components, occupying majority of abdominal space towards right side and also pelvis.

The upper component-is homogeneously hypoechoic in echotexture, and is noted from right side of pelvis extending into RIF, right lumbar, umbilical, left lumbar and LIF regions, it is measuring approximately 25 x 10 cm.

The lower component of this mass is heterogeneously hypoechoic with lobulated margins, with multiple dense hyperechoic calcifications in it.

- Upper component is causing mass effect on adjacent bowel loops.

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Image 1

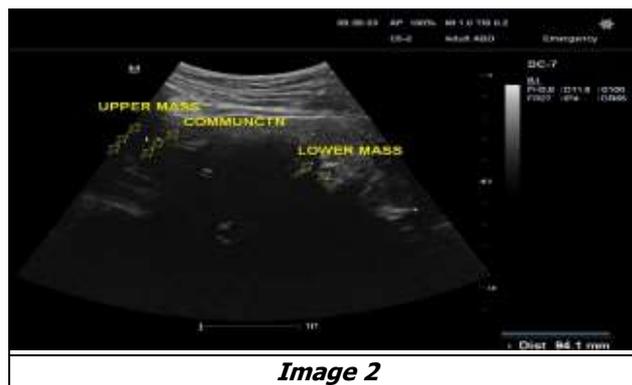


Image 2



Image 3



Image 4

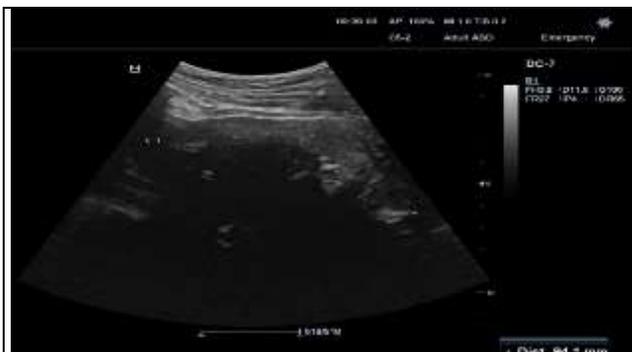


Image 5

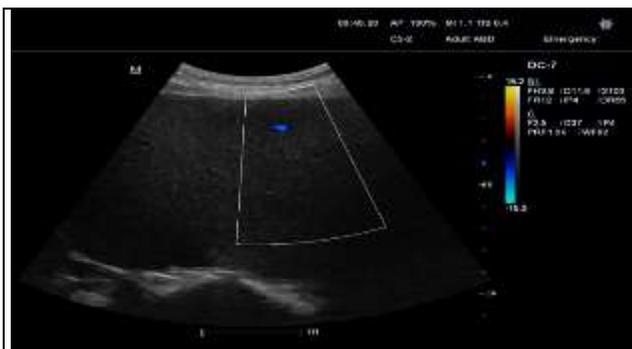


Image 6



Image 7

Impression- Large intraperitoneal hypovascular mass arising from pelvis with two components-

- Upper component showing homogeneously hypoechoic echotexture, occupying majority of lower abdominal space (rt>lt), causing mass effect on adjacent bowels.
- Lower component showing heterogeneously hypoechoic echotexture with multiple dense calcifications, occupying right side of pelvis, causing

mass effect on adjacent bladder and uterus- pushing uterus to left side.

- Mild Ascites

On CT-

- This mass appears heterogeneous in density with few areas of hypodensity and is seen to cause mass effect on adjacent bowels and vessels.
- Another heterogeneously hypodense mass is noted occupying mid part pelvis with multiple irregular scattered dense sheets of calcifications noted through out the pelvic mass.

Contrast studies couldnot be performed on this patient because of allergic reaction to the test dose and known previous history of drug allergies.

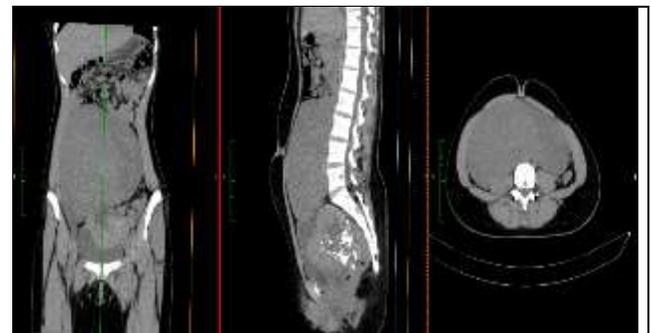


Image 8



Image 9

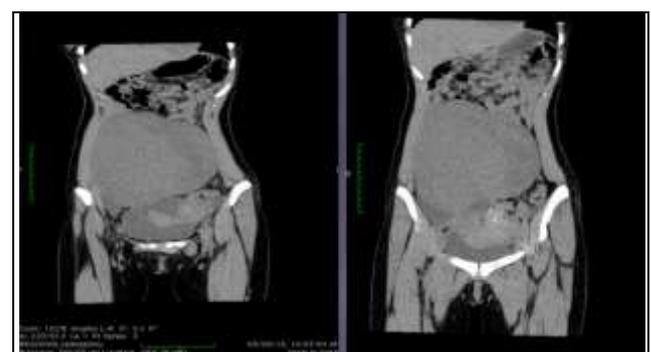
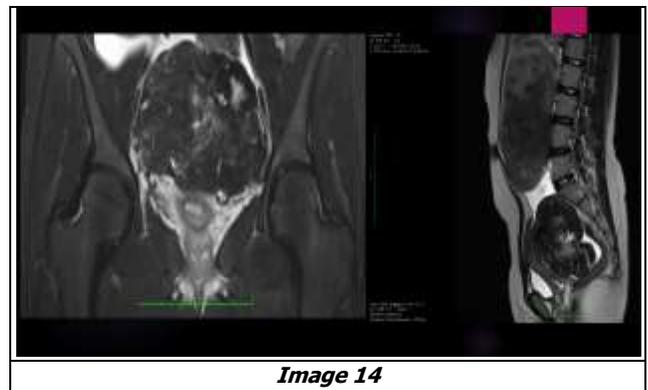
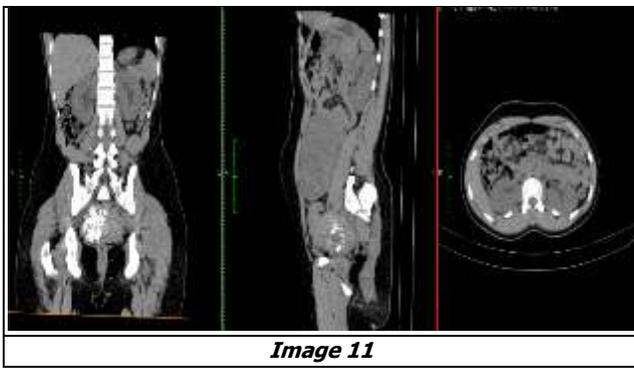


Image 10



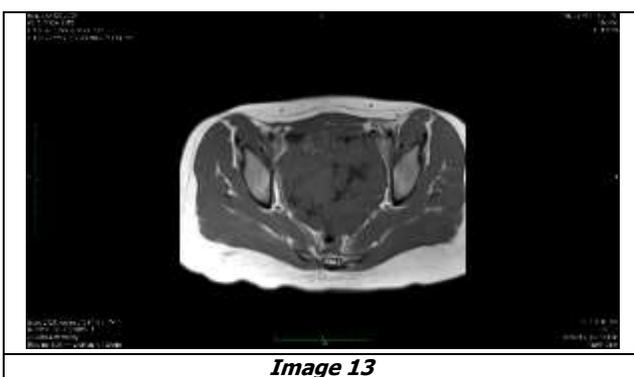
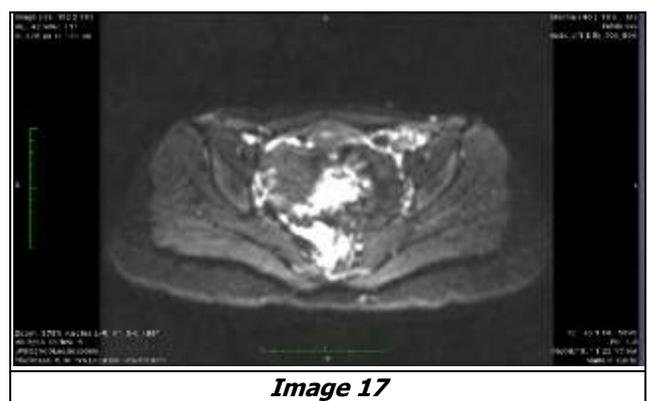
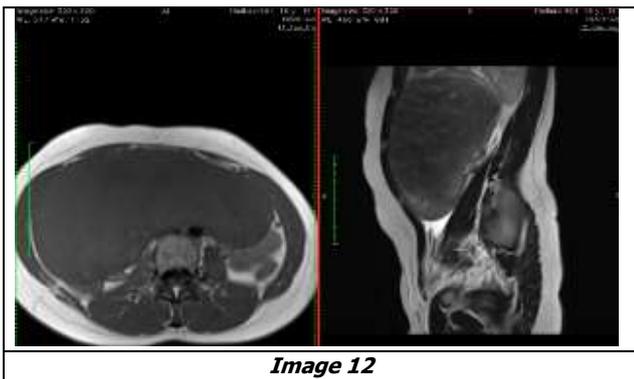
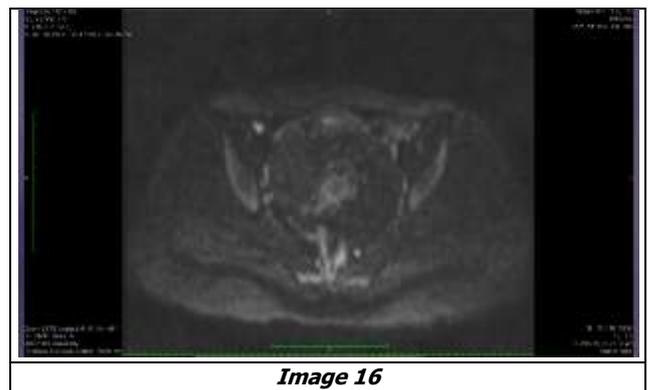
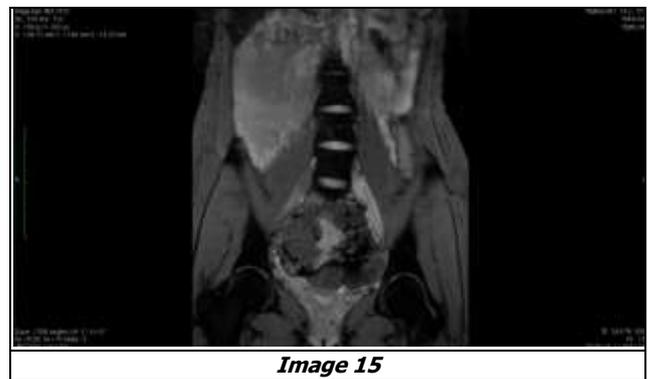
On MRI

- On T1- it appears heterogeneously hypointense with few hyperintense areas.
- On T2 - Heterogeneously hypointense with hyperintense components on T2W & STIR showing restriction on DWI reversal on ADC.

S/o. Large intraperitoneal mass lesion with solid & necrotic component causing mass effect on adjacent bowels & vessels.

Another mass appears.

- T1- Heterogeneously hypointense with irregular scattered hypointensities.
- T2 & STIR- Heterogeneously hypointense with central irregular hyperintensities.
- DWI- Central hyperintensities on T2 show restriction with reversal on ADC.
- This mass is seen to cause mass effect on adjacent organs- uterus, bladder, rectum.
- T2 GRE- Blooming is noted in lower aspect of this mass.



Differential Diagnosis

- 1) Dysgerminoma
- 2) Immature Teratoma
- 3) Brenner Tumour

DISCUSSION OF MANAGEMENT

As occurred in our case, ovarian fibroma is often difficult to diagnose, and the tumour is not often diagnosed accurately until the time of surgery.

Thus, after the surgery, histological confirmation of the prior preoperative diagnosis becomes mandatory for accurate diagnosis, particularly in premenopausal patients.

The treatment plan is surgical resection of the tumour and is associated with very low recurrence rates.

Now-a-days laparoscopic surgery can be an effective and safe alternative approach.

In this case, patient underwent exploratory laparotomy + bilateral salpingo-oophorectomy.

Bilateral salpingo-oophorectomy done & sent for histopathology examination.

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

Intraoperative Image

Gross Findings



Image 18

-Right ovarian solid pedunculated mass of size 20 x 10 cms. with smooth anterior surface but posterior surface showing papillary spiculation.

-Left ovarian hard calcified mass of size 15 x 10 cms. seen.

Histopathology

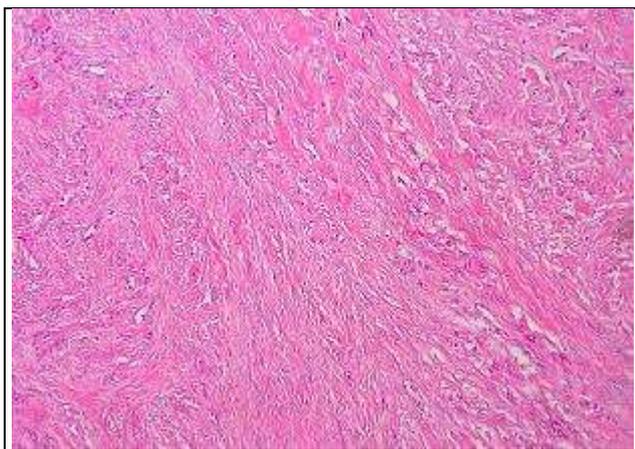


Image 19

Sections studied from both the lesions show fibrothecomatous nature. Spindle cells are arranged in haphazard storiform pattern interspersed with vacuolated cells.

FINAL DIAGNOSIS

Bilateral Ovarian Fibrothecoma.

DISCUSSION

Fibrothecoma-

- The fibrothecomas categorised as sex cord stromal mesenchymal tumours. Their origin is from ovarian stroma, consisting of theca like elements and fibrous tissue.
- Ovarian sex cord tumours are defined as tumours that arise from granulosa cells, theca cells, Sertoli cells, Leydig cells and fibroblasts of stromal origin.¹
- The term 'fibrothecoma' of the ovary is a new term for a tumour of gonadal stromal cell origin and is very rare as it only accounts for 1.2% of all ovarian cancers.²
- Fibrothecoma term has been traditionally used for neoplasms which are intermediate between the theca cell tumours and fibromas.
- The majority of the fibrothecoma usually behave in a benign fashion and their malignant variants are exceedingly very rare.
 - Constitutes approximately 3-4 % of all ovarian neoplasms.
 - Usually seen in perimenopausal age group.
 - Its occurrence before age of 20 yrs. is extremely rare.³
 - Unilateral. Only about 1% are bilateral.
 - Asymptomatic, usually incidental finding. Associated with ascites +/- pleural effusion. (classic Meigs syndrome- only 1% of cases)
 - Can be part of Gorlin Goltz syndrome in which 15-25% of females with this syndrome show fibromas.⁴
 - On USG: lobulated hypoechoic, solid, homogenous internal echogenicity.
 - Minimal to moderate vascularity.
 - Dramatic sound attenuation causing posterior acoustic shadowing.
 - With or without calcifications.
 - On MR: hypo to isointense to pelvic muscles on T1W and low signal intensity on T2W1.
 - On T1W1 c+ -mild enhancement is seen.
 - On CT: NECT: - nonspecific adnexal mass isodense to uterus +/- calcifications.

CECT-

- i) Early: hypovascular with negligible enhancement.
- ii) Delayed: progressive enhancement.

In case of young patients, laparoscopic tumorectomy is recommended over laparotomy. Whereas in postmenopausal women, radical surgery in terms of bilateral salpingo-oophorectomy is indicated.⁵

Histologically, these tumours are characterized by presence of spindle, oval or round cells forming various amount of collagen and a smaller population of theca cells.⁶

DIFFERENTIAL DIAGNOSIS

- 1) **Pedunculated Uterine Fibroids:** Uterine in origin with bridging vessel sign positive.
OVARY is seen separately.
c/f- fibrothecomas enhance significantly less than uterine fibroids.
- 2) **Brenner Tumour:** Usually incidentally detected while operating on other ovary small in size.
Associated with raise in CA125 levels.
It is a hypovascular tumour.
It appears hypoechoic/soft tissue density generally associated with calcifications.
- 3) **Dysgerminoma**
 - Adolescence/ early adulthood
 - Multi-loculated with vascularised enhancing septa
 - These septae cannot be appreciated on T1W1 and they are hypo or isointense on T2W1
- 4) **Immature Teratoma**
 - Predominantly solid heterogenous mass with scattered foci of fat and calcifications.
 - Calcifications are coarse irregular shaped, scattered throughout the tumour.
 - Seen in young females of 15-19 yrs.
 - Usually unilateral.
- 5) **Malignant Serous Epithelial Tumours**
 - It is associated with lymphadenopathy.
 - Spread by peritoneal carcinomatosis.
 - Associated with tumour invasion to pelvic organs.
 - Raised ca-125 levels noted in these tumours.
 - Mean age of presentation is 62 yrs.

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