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Ovarian Fibro-thecoma a Diagnostic Trap – Case Report with Review of Literature

[1]Dr. Monika Kochhar, [2]Dr. Vivek Bharosay, [3]Dr. Hina Mittal
[1]Associate Professor, [2]Professor and Head, [3]Assistant Professor,
[1,2]Department of Pathology, [3]Department of Obstetrics and Gynecology,
Subharti Medical College, Jhajra, Dehradun- 248007

***Corresponding Author:**
Dr. Monika Kochhar
Associate Professor, Subharti Medical College, Jhajra, Dehradun- 248007

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Abstract

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Introduction

Ovarian thecoma-fibroma group (OTFG) are uncommon sex cord -stromal neoplasms. They are solid, benign tumours of the ovary, accounting for 1-4.7% of all ovarian neoplasms. (1-3) According to current revised World Health Organization (WHO) classification of sex cord-stromal tumors (2014), the previous thecoma-firoma group which had been classified under the granulosa-stromal cell tumor group division, were regrouped into pure stromal tumor category.(1) Tumours of the thecoma-fibroma group arising from the ovarian stroma and adopting female differentiation form a spectrum ranging from the typical fibroma at one end to the typical thecoma at the other. Occasionally there are overlapping histological and immunohistochemical features of both fibroma and thecoma, giving rise to term "fibrothecoma".(4) Affected women experience wide range of clinical features. The estrogenic effects are reflected by uterine bleeding , endometrial hyperplasia (15%) and even endometrial carcinoma (21%).(5) About 10-15% of ovarian fibrothecomas can be combined with ascites; however less than 1% can mimic malignancy when present in combination with both ascites and hydrothorax , known as Meigs syndrome.(6) We here by present a case which presented as diagnostic dilemma as neither the clinical presentation nor the preoperative investigations contributed to the diagnosis which was only established after histopathological examination .

Case Report

57-year female post-menopausal for 10 years, multi gravida (para 5 live 5) with all normal vaginal deliveries previously. She presented with pain abdomen off and on for 5 years and loss of appetite. Her medical history was not significant. There was no history of diabetes mellitus, hypertension, thyroid disorder or tuberculosis. On examination, per abdomen was soft and on per vaginal examination - small 4x4 cm mass felt in pouch of Douglas. PAP smear was negative for intraepithelial lesion and malignancy.

On Ultrasonic examination (Figure 1), uterus was post-menopausal status. Right ovary showed a well-defined cyst of size 4.1 X 3.6 cm with echogenic content within it. No internal vascularity /solid component was seen. Was reported as Right Ovarian Complex Cyst.

Contrast Enhanced CT pelvis (Figure 2) showed a thin walled unilocular, hypodense lesion (-20HU) measuring 5 X 4.3 X 4 cm in right adnexa not separately defined from the right ovary. No solid component / mural nodules/septations were seen within. No significant post contrast enhancement was seen. Findings were described as Endometrioma

Laboratory tests comprised routine hematological work up which was normal and other special tests

CA-125 7 U/ml, AFP 1.31 ng/ml and CEA 1.13 µg/L were with in normal ranges.

After informed consent patient was taken up for surgery. Total abdominal hysterectomy with bilateral salpingo-oophrectomy was done. Intra operative findings were, small post-menopausal uterus 4 x 4 cm with a solid mass arising from right ovary with intact capsule. Left tube and ovary were grossly normal. Intestinal surface, omentum, pouch of Douglas and other abdominal viscera were free of any adhesions and tumor deposits.

On gross examination (Figure 3) the uterus was already cut open with attached normal ovary and fallopian tube on the left side and right fallopian tube. The right ovarian mass was seen lying separately. Endocervical canal and endometrial cavity were patent. Endo-myometrial thickness varied from 1-1.2cm. The separately lying right ovarian mass measured 4.2 x 3.5 x 2 cm, was well encapsulated with no breach of the capsule. On cut section, it was solid homogenous with greyish-white to grayish - yellow areas. No necrotic or cystic areas were seen.

For microscopic examination the sections slides were stained with hematoxylin-eosin (HE) and were examined under light microscopes. The sections from right ovarian mass revealed a benign encapsulated tumor made of spindle cells arranged in groups and fascicles (storiform pattern) having bland ovoid centrally placed nuclei, with areas of round to polyhedral cells having round nuclei and moderate amount of pale vacuolated cytoplasm. Areas of hyaline plaques were noted. No capsular infiltration, pleomorphism or mitotic figures were seen.

Right ovarian mass showed features of Pure stromal benign tumor of the fibroma -thecoma spectrum (Figure 4), Fibro-thecoma Right ovary.

Discussion

OTFG neoplasms mostly occur in postmenopausal women, often with a good prognosis. In our case also the patient was of postmenopausal age group. Most patients visited the hospital because of pelvic masses found on routine physical examination, while 63.93% (39/61) of these patients showed no obvious clinical symptoms [7]. Symptoms such as abdominal pain and abdominal lump are common presenting symptoms in case of large tumors. Our case presented with abdominal pain with lump palpable in pouch of Douglas on per vaginal examination. Thecoma is a benign lesion and occasionally malignant. Presentation in the form of pelvic or abdominal pain was uncommon, but postmenopausal bleeding was relatively frequent.[8] Chen et al presented that fibroma and theco-fibroma which comprises 91.80% of cases showed no mensural bleeding.[9] In our case patient had no symptoms of post-menopausal bleeding. Zhang et al concluded that tumors of fibrothecoma group are unilateral occasionally bilateral with oval shape with well-defined border with no enhancement or slight enhancement; accompanied by small amount of ascites.[10] In our case the lesion was unilateral, oval with well-defined margins , showed no enhancement on CT evaluation and was without any ascites. Correlation analysis revealed that the diameter of tumors was statistically significantly correlated with CA125 level and the amount of ascites fluid ($p < 0.05$) [11]. Ascites formation may be due to transudation through the tumor surface which exceeds the peritoneum's resorptive capacity [12]. Irritation of the peritoneal surface by the tumor may explain the increased CA 125 levels [7]. In our case CA 125 was not elevated . as the size of the tumor was moderately enlarged (4.5 x 3.5 x 2 cm).

Pathologically thecomas , fibro-thecomas, and fibromas are stromal neoplasms and may be variants of single entity. These tumors are composed of admixtures of fibrous tissue and theca cells, and therefore have a tendency toward endocrine activity and may be associated with endometrial hyperplasia and endometrial polyps.[2] There were no such symptoms or signs pertaining to this in our patient.

Ultrasonographic (USG) imaging of the pelvis have been well documented , nowadays are the best diagnostic methods for organic pelvic masses as in the present case. USG in our case was suggestive of well defined Ovarian Complex Cyst with low level internal echoes. Fibrothecoma and fibroma show acoustic attenuation, in 96.30% (26/27) , as reported by Huie Chen et al. [9] The reason of the attenuation is most likely due to the low sound propagation of fibroblast tissue [13]. In general, the blood flow of OTFG tumor is not rich as also suggested by no enhancement or slight enhancement on contrast CT. [14]

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The characteristic CT manifestations of the group of ovarian thecoma-fibroma are that they are often unilateral solid mass with the shape of oval and well defined border; no enhancement or slight enhancement; accompanied by small amount of ascites. In a study by Bazot et al reported that 79% of ovarian fibrothecomas appeared as solid masses with delayed accumulation of contrast medium. On dynamic CT, they reported absence of arterial vessels and absence or slight early uptake of contrast enhancement which could be useful findings for preoperative diagnosis. When partly or mainly cystic (21% of cases), ovarian fibrothecomas could not be easily differentiated from other ovarian masses. We also reported similar findings. USG and CT findings with cystic and solid components make differential diagnosis from other ovarian masses difficult such as serous cystadenocarcinoma and other malignant lesions.[14]

MRI is an excellent modality for evaluation of ovarian fibromas and fibrothecomas. Troiano et al. reported the MRI findings of ovarian fibromas and fibrothecomas in 11 patients. All the lesions, regardless of size, had homogeneous low signal intensity on T1-weighted images, and all but one had predominantly low signal intensity on T2-weighted images.[15] Outwater et al., in their report of five ovarian fibromas, also showed that tumor components representing fibrous tissue showed low signal intensity on T2-weighted images.[16] Simiarly, Shinagare et al reported on T1-weighted images, the majority of fibromas and fibrothecomas were isointense to hypointense to both uterine myometrium and iliopsoas muscle. On T2-weighted images, the majority were isointense to hypointense compared with myometrium, and isointense to hyperintense compared with iliopsoas muscle. [17] MRI was not done in our case on account of financial reasons.

Fibrothecoma can mimic like pedunculated intra-ligamentous leiomyomas and other solid ovarian masses such as Brenner tumors, granulosa cell tumors and dysgerminomas. Large tumors can undergo degenerative changes such as infraction, necrosis and cystic change, hence fibrothecoma can be mistaken as malignant ovarian tumor. [15,18,19]

For ovarian tumors, surgery is the treatment of choice The methods differ according to the size ,nature of the tumor, age of the patient, and clinical presentation. Bilateral Salpingo-oophorectomy can be considered for perimenopausal or post- menopausal women, and tumorectomy can be performed in young women. In addition, these procedures could be performed either by laparotomy or by laparoscopy. In the case presented Total abdominal hysterectomy with bilateral salpingo-oophrectomy was done as the patient was post-menopausal and there was no consensus on diagnosis only the histopathology revealed the diagnosis.[20]

Conclusion

Tumours of the thecoma-fibroma group arising from the ovarian stroma and adopting female differentiation form a spectrum ranging from the typical fibroma at one end to the typical thecoma at the other. They present as wide variety of clinical presentations. Preoperatively can mimic malignancy but these tumors carry good prognosis. Only after excision histopathology confirms the diagnosis.

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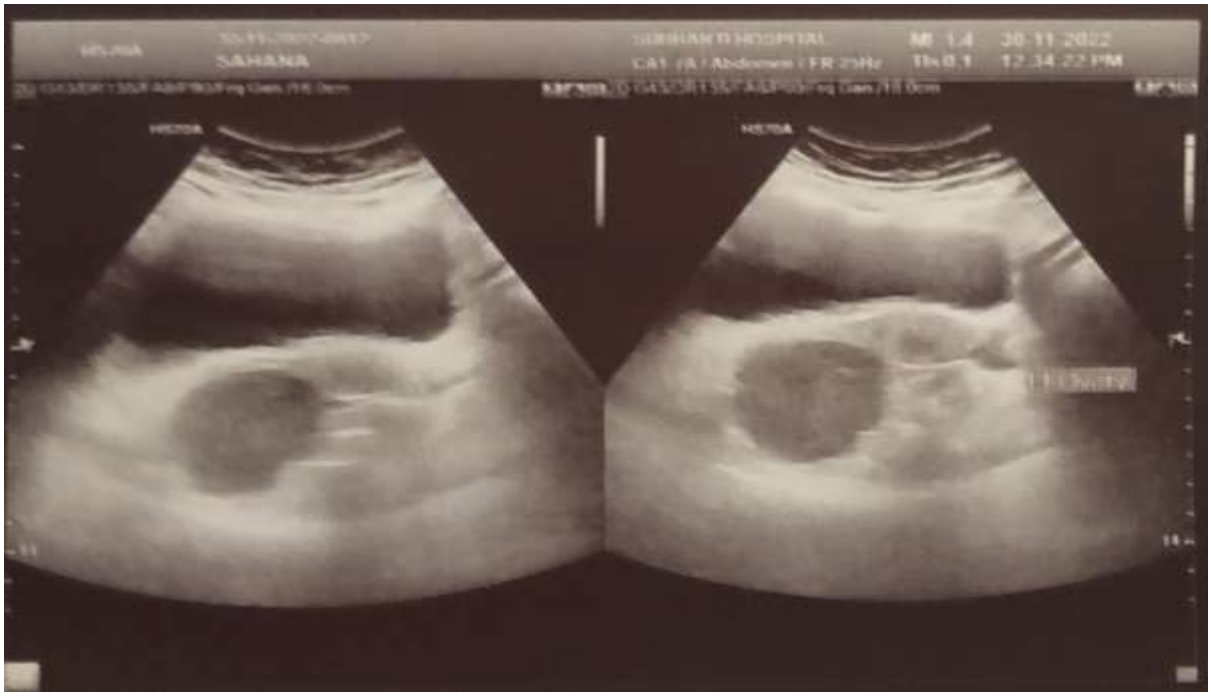

Figure 1: Ultrasound images of the Tumor

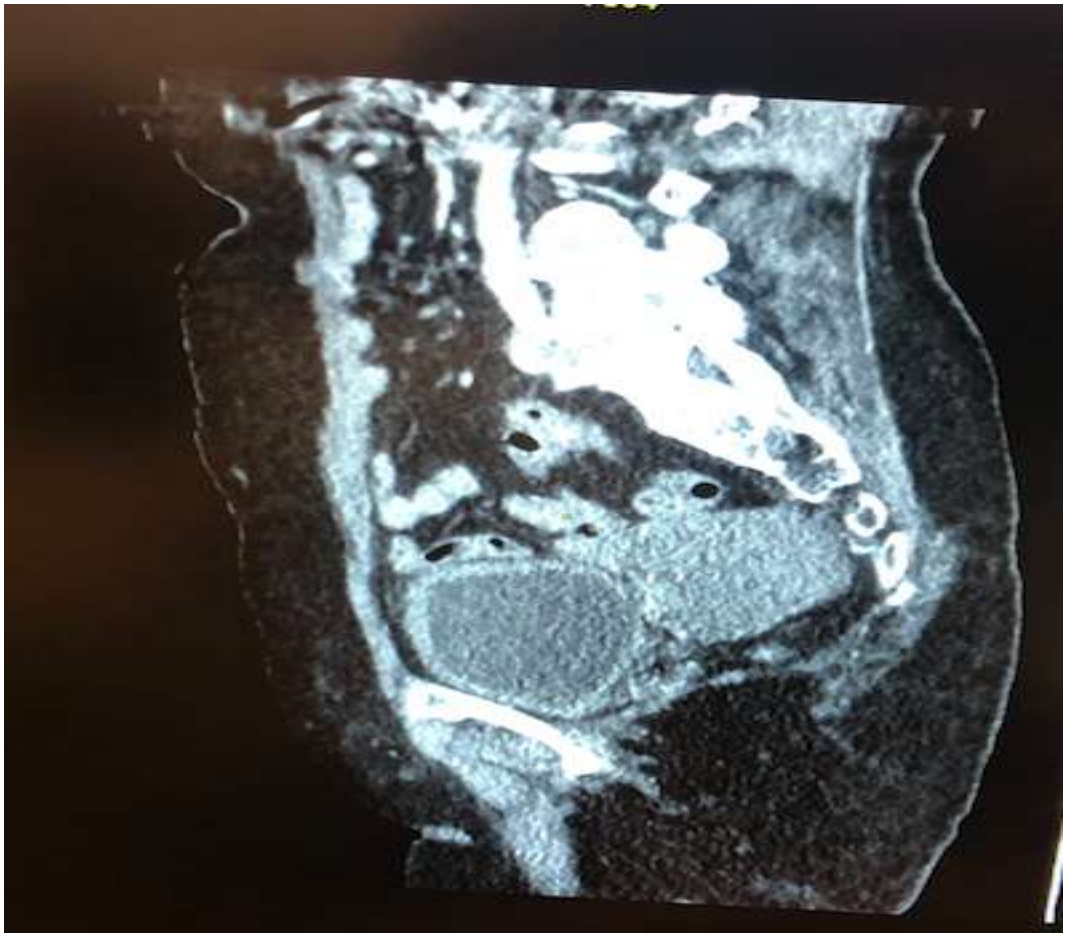

Figure 2: Contrast CT images of the tumor a) Sagittal image

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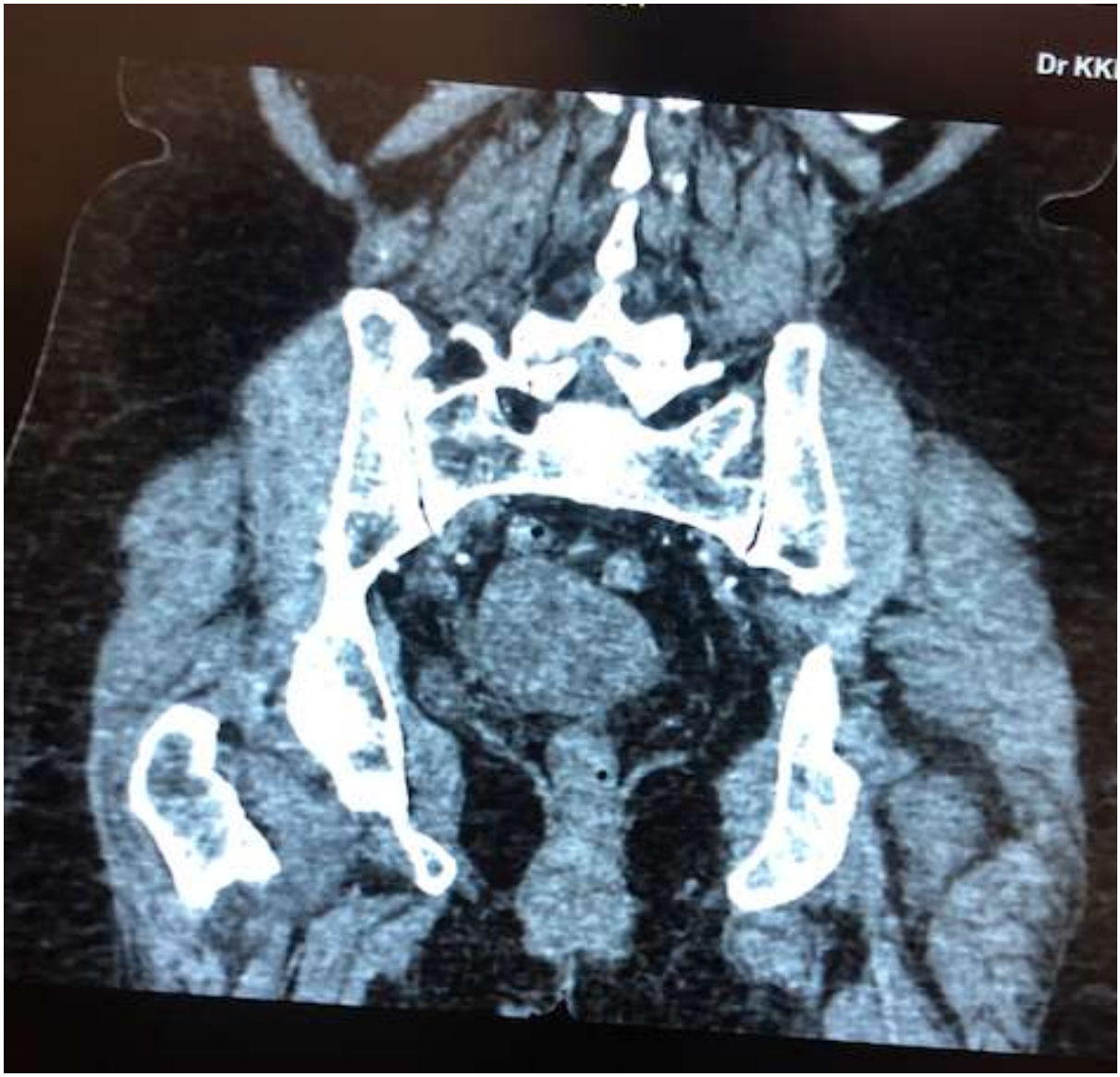

Figure 2 b) Coronal image

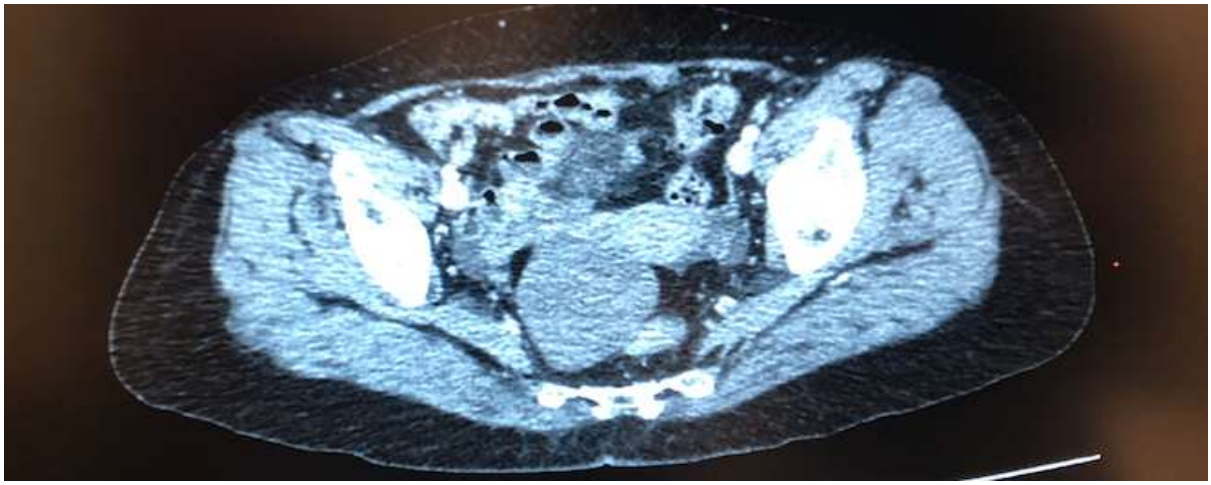

Figure 2 c) Axial Image

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Figure 3: Cut Surface of the lesion with Hysterectomy specimen

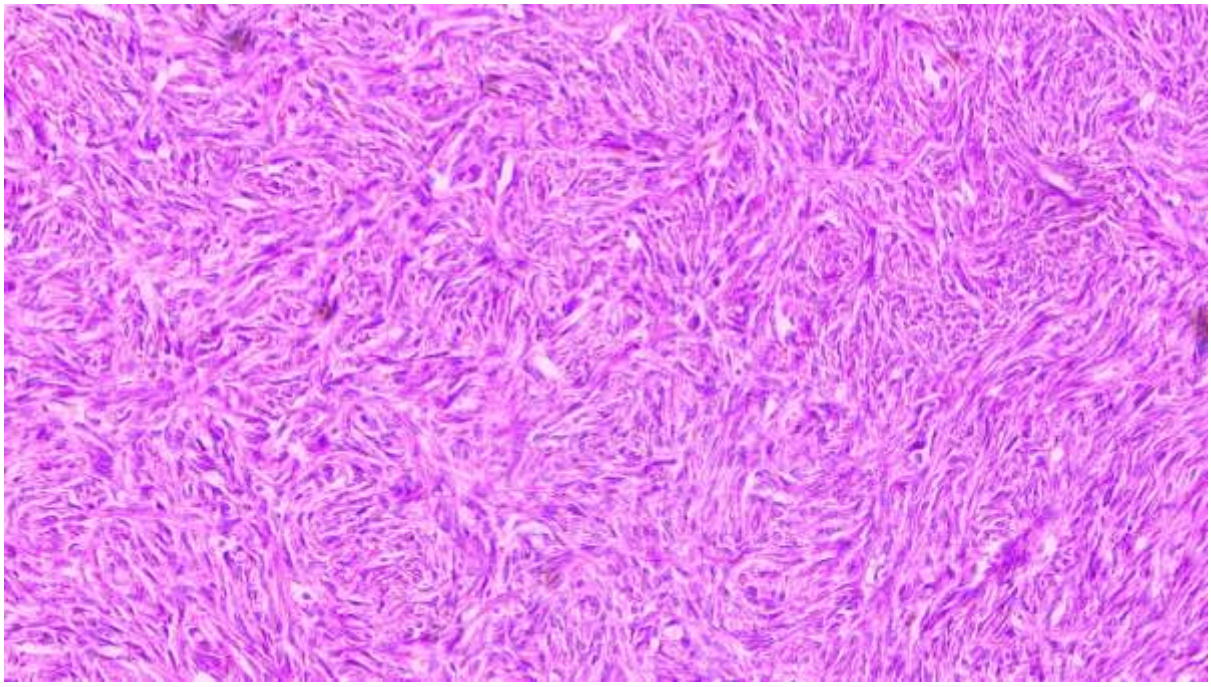

Figure 4: a) The Fibrous component comprising bland spindle cells arranged in groups and fascicles (Storiform pattern) HE 20X

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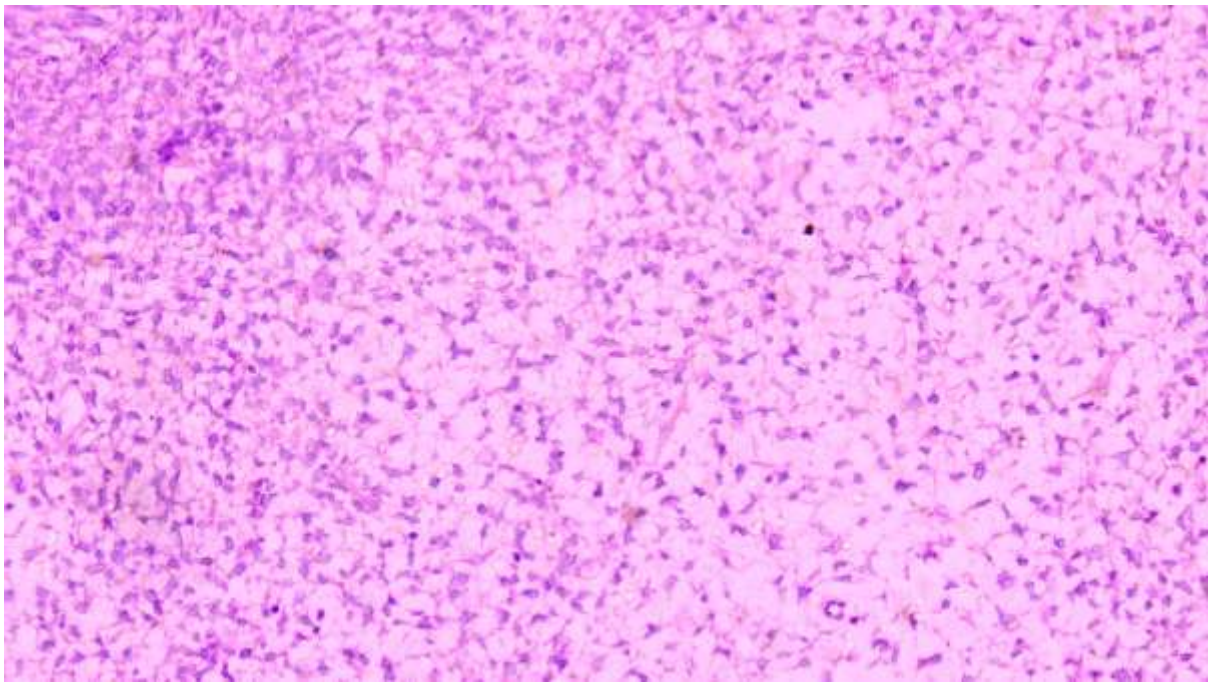

Figure 4:b) The Thecomatous component HE 20X

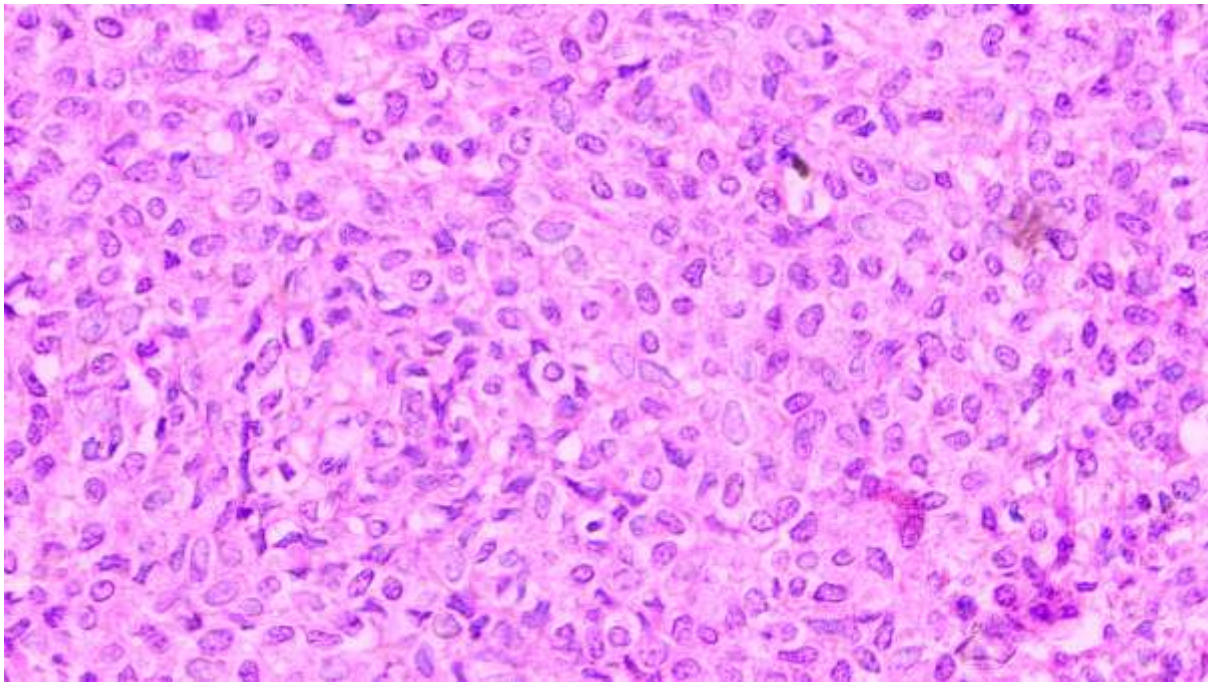

Figure 4 c) The Thecomatous component comprising polyhedral cells HE 40X

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