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Primary Ovarian Fibrosarcoma – A Rare Case Report

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Abstract: *The ovaries, are the site of oogenesis, play important role in ova production, by the process of ovulation, in a cyclical manner. Ovaries are important site for many malignancies in female and most common tumor of ovaries are Surface epithelial tumors, comprising about 60% of all ovarian tumors. Primary Fibrosarcoma of ovaries is a very rare type of Sex cord–stromal tumors and it pose diagnostic difficulties because of similar features of other tumor. We report a case of fibrosarcoma of ovary in a 40 year old female who presented with complaints of lump abdomen, pain, and per vaginal bleeding. She underwent hysterectomy with bilateral salpingo-opherectomy. Histopathological examination of tumor confirmed the diagnosis of fibrosarcoma of ovary.*

Keywords – *cellular fibroma, fibrosarcoma, fibroma, ovary,*

I. Introduction

Sex cord or ovarian mesenchymal tumor constitute 5-12% of all ovarian tumor. Sex cord–stromal tumors of the fibroma–thecoma group account for 9% of ovarian tumors[1]. The most common malignant sex cord–stromal tumor is granulosa cell tumor, it accounts for only about 1% of ovarian tumors. Other sex cord–stromal tumors are rare[1]. Primary ovarian fibrosarcomas are very rare type of sex cord–stromal tumors. Fibrosarcomas were difficult to diagnose, other mesenchymal stromal tumor must be ruled out for exact diagnosis of fibrosarcomas. The ovarian fibromas, cellular fibromas and fibrosarcomas were included in the fibroma thecoma group of ovarian tumors. The ovarian fibroma is the most common sex cord–stromal tumor of fibroma thecoma group followed by cellular fibromas and fibrosarcoma.[2,3]

The clinical presentation is nonspecific. Fibromas occur in 20 to 80 years of age, with an average age of more than 50 years, cellular fibromas appear in somewhat younger age group, with an average age of 40 to 50 years, whereas fibrosarcomas occur in any age but most often in older women.[3] Large fibromas, cellular fibromas and fibrosarcomas causes palpable pelvic mass, abdominal pain, and ascites in 30% patients. Meigs syndrome is an unusual condition in which an ovarian fibroma is accompanied by ascites and hydrothorax.[4] Fibrosarcoma is a malignant mesenchymal tumor that has a poor prognosis and treatment consists of complete resection followed by chemotherapy.[2,5,6]

II. Case Report

A 40 year old Hindu married female visited gynecologic outpatient department of Pacific institute of medical sciences with the chief complaints of lump abdomen since 1 year and irregular, excessive vaginal bleeding since 1.5 years. Patient also complained of body ache, weakness, fatigue, abdominal pain, mild fever and off on and weight loss. On physical examination, a mass was palpated in the right pelvic area measuring 22 cm × 20 cm in size, firm, and immobile.

Per vaginal examination, revealed blood clots and a firm growth in the right side. Percussion of abdomen revealed no fluid in the peritoneal cavity. On auscultation, bowel sound were normal. A provisional diagnosis of ovarian tumor was made.

Pelvic ultrasonography revealed a large heterogeneous mass measuring 20 x 18 cm in the right ovary.

Pre-operative routine investigations were normal except microcytic hypochromic anaemia. The patient underwent total hysterectomy with bilateral salpingo-oophorectomy. Gross specimen was sent for histopathological examination to our department.

Grossly received specimen of uterus cervix with left sided adnexa, right tube and right ovarian mass. Uterus cervix measuring 8x5x5.4 cm on cutting endomyo thickness was 1cm and endometrial canal was patent. Left fallopian tube measuring 5cms. Lumen patent. Left ovary measures 3x2x1 cm. on cut surface corpus luteum seen. Right tube measuring 8cm.

Soft to firm right ovarian mass measuring 22 x 20 cms. External surface of mass was capsulated, bosselated and greyish white to greyish brown with yellowish areas. Cut surface was solid to cystic. Cystic areas filled with haemorrhagic and serous fluid at places.

Histopathologically sections from cervix showed squamous hyperplasia with marked non-specific chronic cervicitis, endometrium showed chronic endometritis, myometrium showed adenomyosis, and both tube showed no pathology. Sections from left ovary showed haemorrhagic corpus luteum and multiple follicular cysts.

Sections from right ovarian mass showed uniform fasciculated growth pattern consisting of fusiform or spindle-shaped cells with mild pleomorphism, with scanty cytoplasm and indistinct cell borders. Cells were separated by interwoven collagen fibres arranged in a parallel fashion. At places cells were arranged in curving or interlacing fascicles, forming a classic herringbone pattern. Tapered darkly stained nuclei with irregular chromatin and variably prominent nucleoli were seen. The average mitotic count was 6-7/10 hpf. No epithelial element was seen.

Hence, on the basis of clinical findings, gross and microscopic features a diagnosis of low grade fibrosarcoma of ovary was made.

Patient was referred to higher centre for immunophenotyping and further management.

III. Discussion

Fibrosarcomas are rare fibroblastic tumour of the ovary that typically has 4 or more mitotic figures per 10 high power fields as well as significant nuclear atypia in most of the cases [7]. Fibrosarcoma has become, in large part, a diagnosis of exclusion.[8]

Fibrosarcomas are the most common ovarian sarcoma, occurring at any age but most often in older women7. In our case patient was of 40 year old female.

The tumor may occur in any soft tissue site but is most common in the deep soft tissues of the lower extremities, particularly the thigh and knee, followed by the upper extremities and trunk. There are numerous reports of fibrosarcoma in the head and neck, including the nasal cavity, paranasal sinuses, and nasopharynx.[8] Rare examples of this tumor have been reported in virtually every anatomic site including the breast, ovary, thyroid, heart, liver, and central nervous system.[7,8]

Ovarian fibrosarcomas are rarely associated with Maffucci syndrome and the nevoid basal cell carcinoma syndrome.[8] Chrisatman JE et al reports a case of fibrosarcomas of ovary associated with Maffucci's syndrome in a young female[9] Kraemer BB et al report a case of fibrosarcoma of ovary in an 8 year old female child associated with nevoid basal cell carcinoma syndrome[10] but in our case it is not associated with any of this syndrome.

Most patients present with a solitary palpable mass ranging from 3 to 8 cm in greatest dimension. It is slowly growing and usually painless.[10]

Biplab k Biswas et al[11] reported a case of giant primary ovarian fibrosarcomas measuring 25x17x12 cms in a 23 year old female in our case tumor size was 20x20cm in 40 year old female.

Prat and scully[2] establishes diagnostic criteria for the diagnosis of ovarian fibrosrcoma by mitotic count as the most important feature. The mitotic figure count up to 3/10 hpf was considered as benign lesion (cellular fibromatous lesions) whereas mitotic count more than 3/ 10hpf was considered as malignant lesion with significant survival difference in their study of 17 cases. In our case mitotic figure were 6-7/10 hpf. The cytologic atypia was less reliable prognostic indicator in fibrosrcomas2.

Poor prognostic factors of fibrosarcoma includes high grade, high cellularity with minimal collagen, mitotic rates >20/10 hpf, necrosis, and little collagen.

Fibrosarcoma is exceptionally rare in the ovary. Many ovarian tumors that have been reported as fibrosarcomas in the past would now be considered mitotically active cellular fibromas. Fibrosarcomas are typically large tumors that often have spread beyond the ovary at diagnosis. The differential diagnosis includes leiomyosarcoma, a gastrointestinal stromal sarcoma, and various types of primary or metastatic soft tissue sarcomas.

Immunophenotyping of Fibrosarcomas are positive for vimentin and very focally for smooth muscle actin, representing myofibroblastic differentiation. Some cases arising in dermatofibrosarcoma or solitary fibrous tumour are CD34 positive.

IV. Conclusion

Primary ovarian fibrosarcomas is a rare neoplasm of ovary but it must be considered as differential diagnosis of unilateral solid ovarian mass in all age group. Preoperative diagnosis is helpful in most cases for proper intraoperative management and postoperative management.

V. Figures and Tables

Figure With Legend

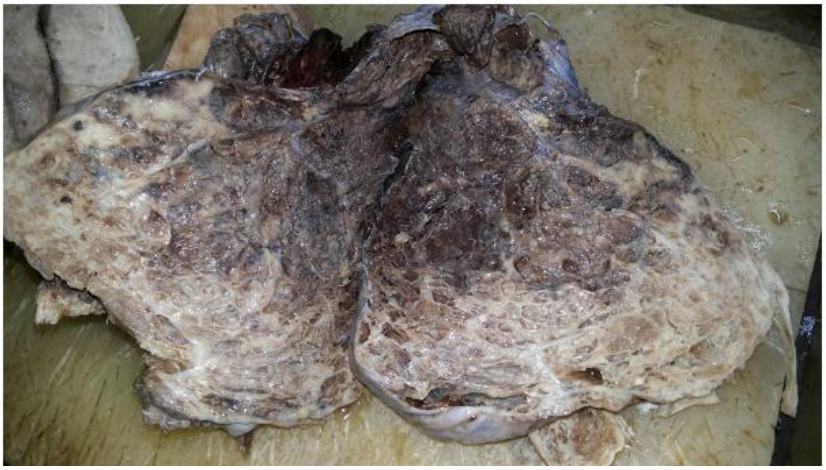

Figure-1 Cut surface of tumor mass showing cystic and solid areas with haemorrhage.

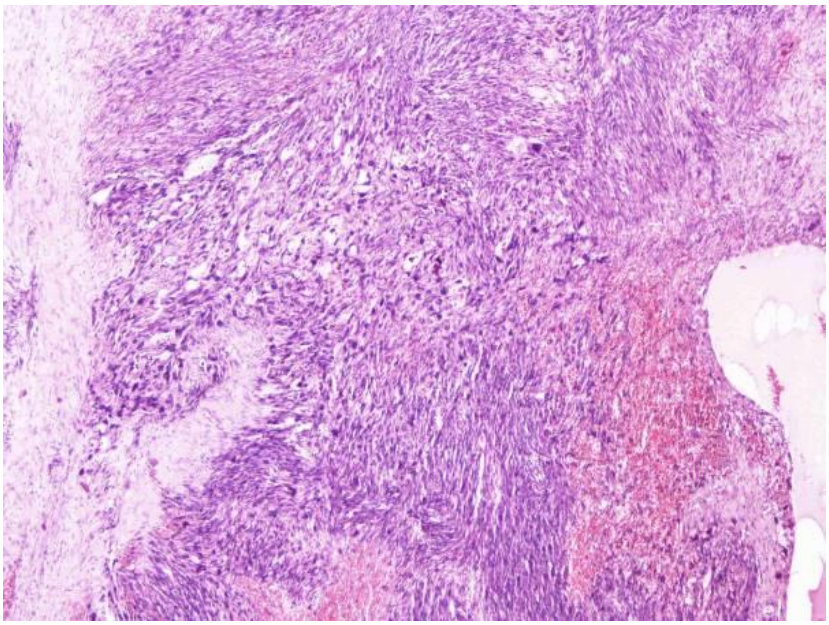

Figure 2- section show spindle cells arranged in herringbone pattern (Low power 40x).

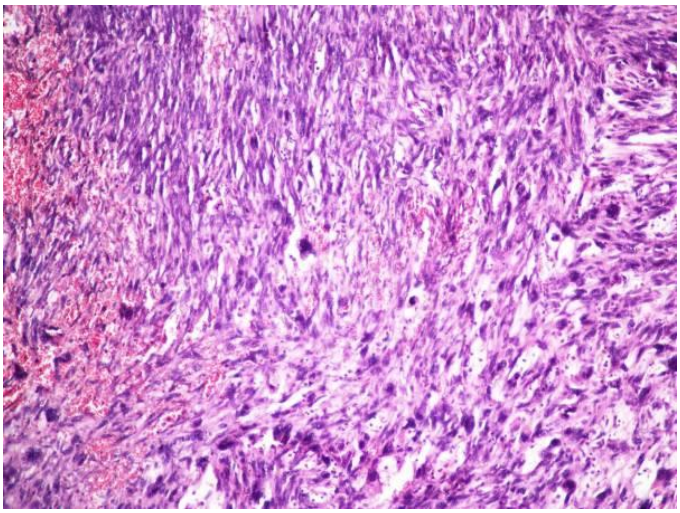

Figure 3- section show spindle cells arranged in fascicular pattern and mitotic figure at places (Low Paper, 100x)

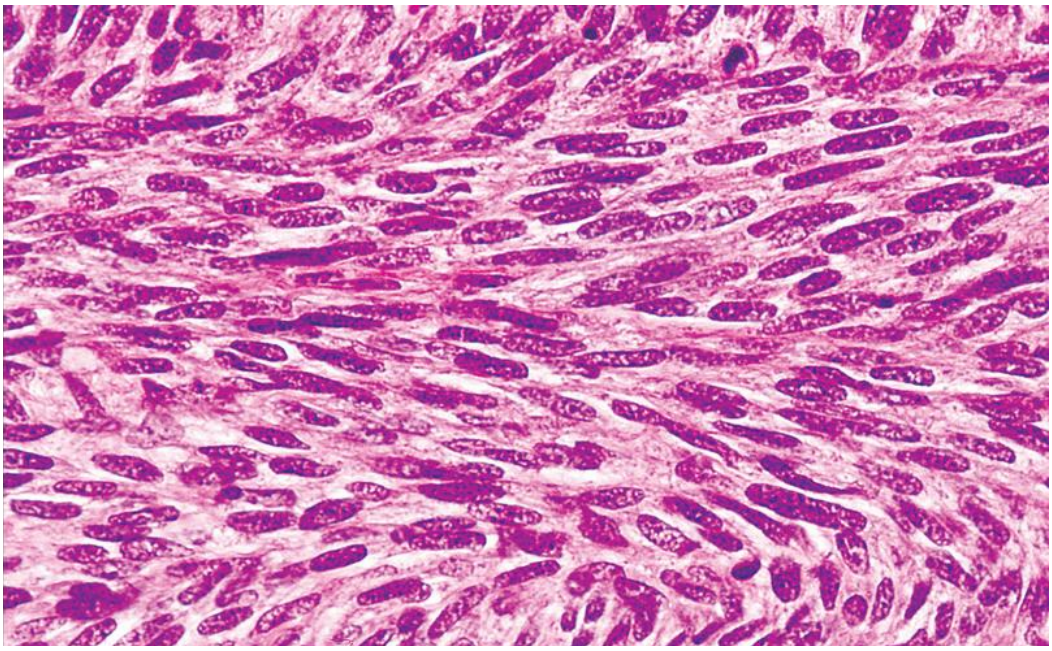

Figure 4 section shows spindle cells with mild pleomorphism and mitotic figure (High Power, 400x).

VI. Conclusion

Primary ovarian fibrosarcomas is a rare neoplasm of ovary but it must be considered as differential diagnosis of unilateral solid ovarian mass in all age group. Preoperative diagnosis is helpful in most cases for proper intraoperative management and postoperative management.

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