IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 15, Issue 6 Ver. IV (June. 2016), PP 79-83
www.iosrjournals.org

Ovarian Stromal Tumour with Minor Sex Cord Elements- A Case Report and Review of Literature.

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Abstract:
Objective: *Ovarian stromal tumour with minor sex cord elements is a rare tumour. It is defined as a predominantly fibromatous or thecomatous tumour containing minor sex cord elements in less than 10% of the tumour area. It can pose a diagnostic dilemma to pathologists. These tumours may be hormonally active and predispose to carcinoma endometrium. We report the 16th case of ovarian stromal tumour with minor sex cord elements.*
Case report: *We report the case of a 52 year old lady who presented with pain abdomen and mass per abdomen. Histopathologically, the ovarian tumour was a stromal tumour with minor sex cord elements.*
Conclusion: *Ovarian stromal tumours with minor sex cord elements are a distinct clinoco-pathologic entity. The knowledge of this entity is crucial in order to avoid misdiagnosis. Such patients require long term follow up as hyperoestrogenemia may predispose to endometrial cancer.*
Keywords: *Ovarian fibromata, thecoma, sex cord-stromal tumor, inhibin, granulosa cells*

I. Introduction

Sex cord –stromal tumours are ovarian tumours composed of granulosa cells, theca cells, Sertoli cells, Leydig cells and fibroblasts of stromal origin singly or in various combinations. Overall they account for 8% of ovarian neoplasms.[1] Fibromas and thecomas may show a significant morphological overlap, which has led to the use of term fibrothecoma. They account for 3-4% of all ovarian neoplasms.[2,3]

Ovarian stromal tumour with minor sex cord elements was first described by Young and Scully in 1983.[3,4,5,6] It is defined as a predominantly fibromatous or thecomatous tumour containing minor sex cord elements in less than 10% of the tumour area.[1,3,5] To the best of our knowledge, only 15 cases of ovarian stromal tumour with minor sex cord elements have been reported till date.[2,3,4]

II. Case Report

A 52 year old lady came with the complaints of severe abdominal pain and mass per abdomen since one month. Bilateral salpingo-oophorectomy with dissection of bilateral pelvic nodes and omentum was done and the specimen was received for histopathological examination.

2.1 Gross

The left ovarian mass consisted of a single irregular mass measured 25 ×20× 15 cms. External surface (Figure 1) was nodular, cystic at places. Congested blood vessels were seen. On sectioning it drained hemorrhagic serous fluid. Cut surface (Figure 2) showed multiple cystic and solid fleshy yellow areas. Largest cyst measured 15 cms in diameter. The attached, stretched left fallopian tube measured 9cms in length, the external and cut surface of which was unremarkable.

Separately sent right ovary measured 2.5×2.2 ×1.5 cms. External surface was cystic and cut surface showed a single cyst measured 1.7 cms in diameter. The right fallopian tube measured 3.5 cms in length, the external surface and cut surface of which was unremarkable.

Separately sent right pelvic lymphnodes consisted of 2 pieces of fibrofatty tissue, from which 2 lymphnodes were dissected, the larger measured 2×1×0.7 cms. Cut surface of the lymphnodes showed tan pink areas.Separately sent left pelvic lymphnodes consisted of 2 pieces of fibrofatty tissue, from which 2 lymphnodes were dissected, the larger measuring 1.5 cms in diameter. Cut surface of the lymphnodes showed tan pink areas.Separately sent omentum consisted of single piece of fibrofatty tissue measured 42×20×4cms, from which one lymphnode was dissected, measured 0.4 cms in diameter.

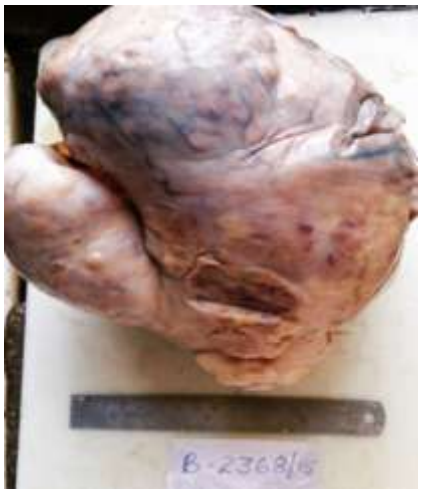

Figure 1- External surface of left ovarian tumour.

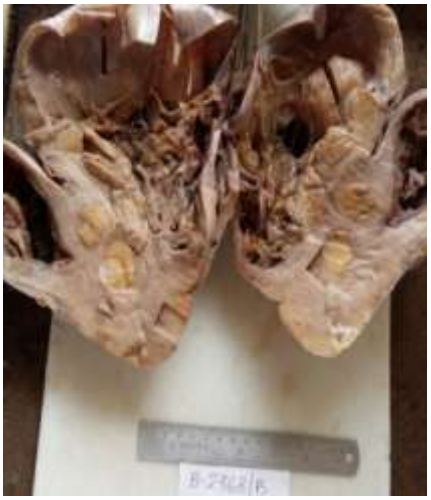

Figure 2- Cut section of left ovary showing solid and cystic areas.

2.2 Microscopy

Sections studied from the left ovary showed well encapsulated mass with majority of tumour cells arranged in intersecting fascicles composed of oval to spindle shaped cells with scant cytoplasm and bland nuclei (Figure 3). Also seen were cells with abundant pale vacuolated cytoplasm and uniform bland oval to spindle shaped nuclei (Figure 4). These cells constituted fibroma- thecoma component of tumour. Less than 5% of the tumour cells showed granulosa cells with scant eosinophilic cytoplasm and vesicular nucleus with nuclear grooves, arranged in diffuse sheats along with micro and macrotubules (Figure 5 and 6). Features were suggestive of stromal tumour with minor sex cord elements. The omentum and lymphnodes were free of tumour.

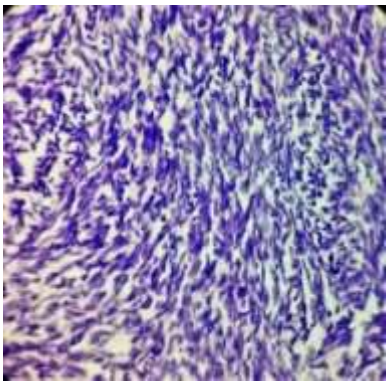

Figure 3- Section showing tumour cells arranged in intersecting fascicles composed of spindle shaped cells with scant cytoplasm and bland nuclei. H&E 10x.

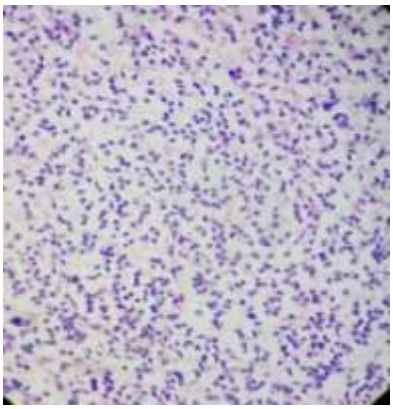

Figure 4- Section showing cells with abundant pale vacuolated cytoplasm and uniform bland oval to spindle shaped nuclei. H&E 10x.

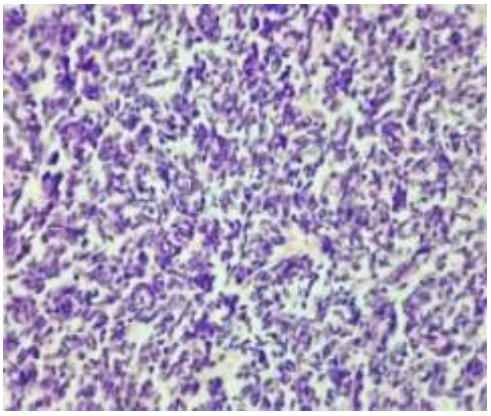

Figure 5- Section showing granulosa cells with scant eosinophilic cytoplasm and vesicular nucleus with nuclear grooves. H&E 10x.

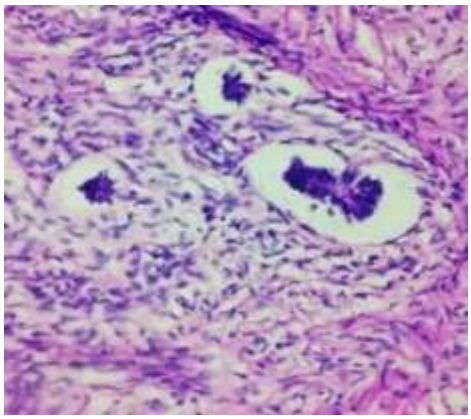

Figure 6- Section showing granulosa cells arranged in tubular structures. H&E 40x.

III. Discussion

Ovarian sex -cord stromal tumors arise from ovarian stroma, sex-cord derivatives, or both.[7] They comprise only 8% of all ovarian neoplasms.[1,3,6] Ovarian stromal tumour with minor sex cord elements was first described by Young and Scully in 1983.[3,4,5,6] It is a rare fibrothecomatous tumour containing scattered sex cord elements. By definition the sex cord element must account for less than 10% of the composition of the tumour.The minor sex cord elements are seen as small nests or tubules of cells resembling granulosa cells, sertoli cells, or indifferent cells of sex cord type. It is recognised as a distinct entity by the WHO. [1]Literature search revealed 15 cases of ovarian stromal tumour with minor sex cord elements. We report the 16th case of ovarian stromal tumour with minor sex cord elements with review of literature.

The average age of presentation ranged from 16 to 65 years, with a mean age of 59 years.[1,2] Our patient was 52 years old. Tumour is usually hormonally inactive, but has been associated with oestrogenic features, e.g. endometrial hyperplasia or diffuse complex atypical hyperplasia or even adenocarcinoma. [1,2,4,5] These patients generally present with abdominal pain, bleeding per vaginum and adnexal mass.[4] Our patient presented with abdominal pain and adnexal mass.

Macroscopically, usually the tumour is solid, not indistinguishable from fibroma or thecoma, and ranges from 1-10 cm in diameter.[1,4] They are firm, solid, yellow white to yellow grey in colour.[5] In our patient the size of the tumour was 25 ×20× 15 cms, and solid and cystic areas were present. Microscopically they are composed of spindled cells, arranged in intersecting fascicles with variable amount of collagen deposition and intermingled sex cord elements. The nucleus of stromal cells is elongated and cigar shaped without prominent nucleoli. The cells of minor sex cord elements resemble granulosa, sertoli, undifferentiated sex cord, or steroid cells. The term minor component of sex cord elements is defined as sex cord elements occupying no more than 10% of the area of the tumour on any slide. The individual aggregate of these minor sex cord elements should not be greater than 0.45mm. [1,4,5,6]

Immunohstochemically, the minor sex cord elements are positive for inhibin, calretinin, CD99, CD56, antikeratin antibody KL1 and MIC.[2,5,6]Differential diagnoses include ovarian fibromatosis, Brenner tumour, and adenofibroma. In ovarian fibromatosis, there is a proliferation of spindle- shaped cells with abundant collagen formation and focal areas of oedema. The normal follicular structures of the ovary are preserved in ovarian fibromatosis.[1,2] In adenofibroma, the glands are abundant, larger, and tubular and more variable in size

when compared to uniform tubules of minor sex cord elements. Both these entities are inhibin and calretinin negative. The epithelial aggregates of Brenner tumour are composed of transitional cells, mucinous cells or both and sometimes have a central lumen containing eosinophilic secretions and cells are EMA and CK positive.[4,5,6] **Table I** shows the comparision of the histological features of above mentioned tumours.

Table I: Comparison of key histologic features to the mentioned differential diagnosis.[6]

Histologic features	**Fibroma with minor sex cord elements**	**Ovarian fibromatosis**	**Brenner tumor**	**Adenofibroma**
Spindled cells in fascicles and whorls	Present	Present	Present	Present
Collagen	Variable	Abundant	Variable	Variable
Normal ovarian follicles	Replaced by fibrous stroma	Preserved	Absent	Absent
Small nests of undifferentiated sex cord type cells	Present (<10% of tumour area)	Absent	Absent	Absent
Edema	Absent	Present	Absent	Absent
Epithelial nests of transitional/ mucinous cells	Absent	Absent	Present	Absent

In 1983, **Young and Scully[8]** reported 7 cases of stromal tumours of the ovary with a minor component of sex cord elements. 5 cases were ovarian fibroma with minor sex cord elements and the other 2 were classified as luteinized thecoma and stromal-Leydig cell tumour with minor sex cord elements. Two out of seven cases had well-differentiated adenocarcinoma in the endometrium.

In a separate study of 50 luteinized thecomas and stromal Leydig cell tumours in 1982, **Zhang et al[9]** reported 2 of the 50 tumours to contain sex cord elements consistent with granulosa cells. 23 of the fibrothecomatous tumours showed the coexistence of cystic hyperplasia, atypical hyperplasia, or adenocarcinoma of the endometrium. These findings were attributed to the production of oestrogenic hormone by the tumours.

Most of the cases of ovarian stromal tumour with minor sex cord element have behaved in a benign fashion similar to that of fibroma.[6] However, a single case report of fibrothecoma with minor sex cord elements showing focal fibrosarcomatous change has been described by **Lee et al[10].** The criteria used for the diagnosis of fibrosarcoma were increased mitotic counts in association with hypercellularity and mild to moderate nuclear atypia. The uterus showed multifocal endometrial adenocarcinoma of endometroid type.

Sood et al[2] reported a case of virilising ovarian fibrothecoma with minor sex cord elements in a 13 year old girl, which showed negativity for inhibin and positivity for calretinin.[3] They concluded that the incidental presence of sex cord elements in ovarian stromal tumour needs to be confirmed by both inhibin and calretinin to categorise this distinct entity.**Mandal et al[6]** reported the first case of fibroma with minor sex cord elements, in a normal sized ovary, as an incidental finding. **Table II** details about comparison between the present study and other studies.

Table II: Comparison of various case reports of ovarian stromal tumour

	Present case	**Kumar et al**	**Kawatra et al**	**Mandal et al**	**Sood et al**	**Lee et al**	**Gupta**
Age	52 years	79 years	65 years	45 years	13 years	69 years	36 years
Symptoms	Pain in abdomen and mass per abdomen	Post menopausal bleeding, pain in lower abdomen.	Bleeding and pain abdomen since 6 months	Menorrhagia and prolapse since 6 months	Oligomenorrhoea and masculinizing features since 1 month	Postmenopausal bleeding	Abdominal pain and mass since 1 month
Unilateral/ bilateral	Unilateral	Unilateral	Unilateral	Unilateral	Unilateral	Unilateral	Bilateral
Size of ovary	25 ×20× 15 cm	21x14x12 cm	5x4x2 cm	2.5x2.5x1.5 cm	10 cm diameter	13x8.5x6 cm	14x12x8 cm and 9x8x8 cm.
Cut surface ovary	Multiple cystic and solid fleshy yellow areas.	Homogenously fleshy with areas showing yellowish discoloration.	Firm grey white solid mass with focal yellowish firm areas.	Well circumscribed yellowish nodular firm area.	Solid with focal cystic and hemorrhagic areas	solid, firm, yellow-grey mass containing an area of cystic degeneration	firm and grey white
Endometrium	---	Well differentiated	Simple glandular hyperplasia	Proliferative	--	complex atypical hyperplasia	Proliferative phase

		endometroid adenocarcinoma	without atypia			and focal adenocarcinoma.	
Stromal component	Fibroma	Fibroma-thecoma	Fibroma	Fibroma	Fibrothecoma	Fibrothecomawith focal fibrosarcomatous change	Fibroma
Minor sex cord component	Granulosa cells	Aggregates of granulosa cells	Uniformly large cells with inconspicuous nucleoli in clusters and cords.	Aggregates of undifferentiated sex cord type cells with poorly defined tubular structure.	Sertoliform cells of intermediate differentiation	Granulosa and steroid islands	Tubular structures filled with eosinophilic material

IV. Conclusion

Ovarian stromal tumours with minor sex cord element are rare tumours and a distinct clinoco-pathologic entity. They can pose a diagnostic dilemma to pathologists. The knowledge of this entity is crucial in order to avoid misdiagnosis. A close follow up of the patient should be done, since the tumour has been associated with oestrogenic features, and may predispose to endometrial cancer. Also the clinical behaviour and risk of recurrence in these patients needs further evaluation.

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