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Case Report

Ovarian fibrothecoma with minor sex cord elements: a case report

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ABSTRACT

Ovarian fibroma is the most common sex cord stromal tumour of ovary accounting to 1-5% of all ovarian tumours. Minor sex cord elements in ovarian fibroma are a rare entity occupying less than 10% of tumour area. To the best of our knowledge only 20 cases has been reported till date. This case is presented because of its rarity. Authors reported a case of fibrothecoma with minor sex cord elements in a 70yr old postmenopausal women who presented with postmenopausal bleeding with abdominal mass.

Keywords: Fibroma, Minor sex cord elements, Postmenopausal women, Sex cord stromal tumours

INTRODUCTION

Sex cord stromal tumours are group of tumours representing 8% of all ovarian neoplasm and are composed of granulosa cells, sertoli and leydig cells, thecal cells and fibroblasts either singly or in various combinations.[1] Ovarian fibromas are the most common sex cord stromal tumours accounting to 1-5% of all ovarian neoplasms.[2,3] Young and scully in the year 1983 first described the presence of minor sex cord elements in ovarian fibroma.[4]

It is defined as the tumour with predominantly fibromatous or the comatous area containing scattered minor sex cord elements in less than 10% of tumour area on any slide and individual aggregate should not be more than 0.45mm. These cells form discrete tubules or small nests resembling granulosa cells, sertoli cells or indifferent cells of sex cord type either with or without steroid hormone production in spindled stroma.

CASE REPORT

A 70 yr old woman has come to our gynaecological department with history of postmenopausal bleeding with left adnexal mass. Her routine hematological investigation is normal with CA 125 levels of 39.43IU/ml. CT scan revealed a well defined heterogenous solid lesion measuring 6.5cm x 3.4cm in left adnexa.

She underwent total abdominal hysterectomy with bilateral salphingo opherectomy. Grossly uterus with cervix measured 10x8x6cm with bilateral tubes 4cm in length, left ovary 7x6x4cm with other ovary 2x2x1cm. Cut surface uterus revealed a polyp of size 1m arising from uterine fundus in endometrial cavity.

Left ovary was replaced by a tumour with solid, firm grey white mass. Microscopically the ovarian mass showed benign spindle shaped cells arranged in fascicles and in storiform pattern enclosing small aggregates of sex cord like elements occupying less than 10% of tumour area. These elements are arranged in tubular pattern. There are areas showing luetinization. Endometrium was in proliferative phase with polyp was a benign endometrial polyp. Solid firm homogenous mass with focal yellowish areas (Figure 1). Sheets of benign spindle cells arranged in fascicles and storiform pattern (4x H& E) (Figure 2). Small aggregates of sex cord like elements arranged in

tubular pattern embedded within the fibrothecomatous stroma (45x H&E) (Figure 3).

Figure 1: Gross appearance of ovarian mass.

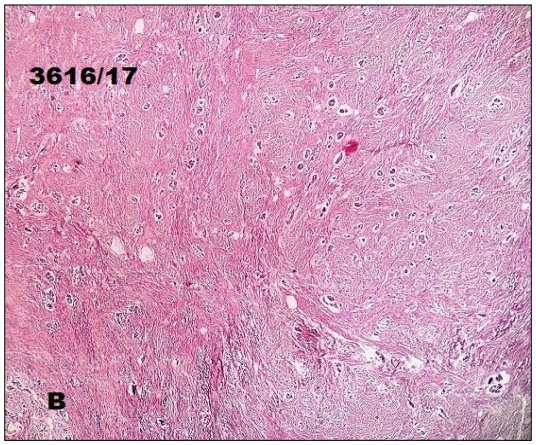

Figure 2: Microscopic appearance of ovarian mass.

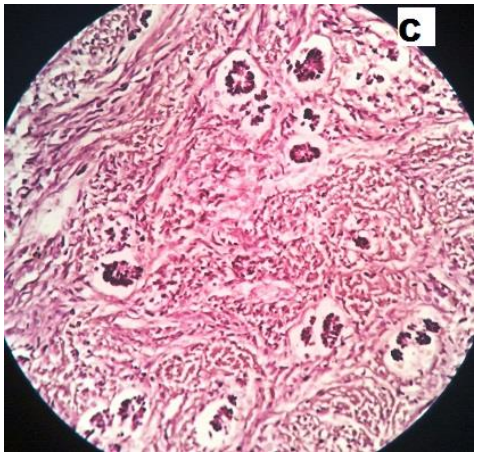

Figure 3: Sex cord like elements arranged in tubular pattern.

DISCUSSION

Ovarian stromal tumours with minor sex cord elements are a rare neoplasm which was first described by Young and Scully in the year 1983. It is defined as tumour with predominantly fibromatous or the comatous area containing scattered minor sex cord elements in less than 10% of tumour area on any slide and individual aggregate should not be more than 0.45mm.[5] These cells form discrete tubules or small nests resembling granulosa cells, sertoli cells or indifferent cells of sex cord type with or without steroid hormone production in spindled stroma. They are polygonal cells with uniform nuclei and small amount of cytoplasm. The most common clinical symptoms are abdominal pain, bleeding per vaginum and adnexal mass. This case presented with history of postmenopausal bleeding. The average age groups affected are between 16-65yrs with median age of 59yrs.

Table 1: Comparison of ovarian fibroma with other tumours or tumour like conditions.

Features	Fibroma with MSCE	Fibromatosis	Brenner tumour	Adenofibroma	Metastatic carcinoid
Spindled cells in fascicles and whorls	+	+	–	–	Extensive stromal proliferation
Collagen	Variable	Abundant	Variable	Variable	-
Normal ovarian follicles	Replaced by fibrous sroma	Preserved	-	-	Replaced by tumour cells
Small nests of undifferentiated sex cord cell type cells	<10% of tumour area	–	–	–	Insular, trabecular, rarely solid pattern
Edema	-	+	-	-	-
Epithelial nests of transitional/mucinous cells	-	-	+	-	Uniformly small round cells with clumped chromatin
IHC	Positive for inhibin, calretinin, CD99, CD56	Negative for inhibin and calretinin	EMA CK+	-	Chromogranin Synaptophysin CD56

Table 2: Comparison of individual case reports reported in the literature with present study.

Author	Age in yrs	Clinical features	Endometrial changes	Stromal component	MSC component	IHC in MSC
Mathur et al[2]	52	Bleeding Per vaginum	Simple endometrial hyperplasia without atypia	Fibrothecomatous component	Poorly defined nests in solid, hollow tubules, cords	Inhibin Calretinin
Mandal et al[4]	45	Menorrhagia with incidental finding in normal sized ovary	Proliferative endometrium	Fibromatous component	Small aggregates of undifferentiated sex cord like cells with poorly defined tubular structures	Negative for SMA, Vimentin, EMA
Sujatha et al[6]	52	Abdominal mass and pain	-	Fibrothecomatous component	Granulosa cells in micro & macro nodular pattern	-
Kawatra et al[7]	65	Postmenopausal bleeding with abdominal pain	Simple endometrial hyperplasia with polyp	Fibromatous component	Uniform large cells with inconspicuous nucleoli in poorly defined nests or cords	Inhibin strongly positive
Sherwani et al[8]	52	Abdominal mass	Atrophic endometrium	Luetinized fibroma	Sertoli like cells arranged in tubular pattern	-
Lee et al[9]	69	Postmenopausal bleeding with abdominal mass	Endometroid adenocarcinoma	Fibrothecomatous component with focal fibrosarcomatous change	Small well demarcated closely packed aggregates of cells (Granulosa and steroid cells)	-
Kumar et al[10]	79	Postmenopausal bleeding with abdominal mass	Endometroid adenocarcinoma	Fibrothecomatous component	Granulosa cell aggregate	Inhibin calretinin Positive
Shilpa et al[11]	55	Postmenopausal bleeding with abdominal mass	Endometroid adenocarcinoma	Fibromatous component	Nests of cells with grooved nuclei and minimal cytoplasm	Inhibin strongly positive
Sood et al[12]	13	Masculinizing features with Abdominal mass	-	Fibrothecomatous component	Sertoliform cells of intermediate differentiation	Calretinin strongly positive
Sharma et al[13]	40	Masculinizing features with Abdominal mass	-	Fibrothecomatous component	Sex cord like cells	Inhibin and calretinin Positive
Yang et al[14]	58	Abdominal mass	-	Fibromatous component with mucinous cystadenoma	Uniform large cells in nesta and cords (Unclassified sex cord elements)	Inhibin
Asavari et al[15]	18	Abdominal mass and Pain	-	Fibrothecomatous component with serous cystadenoma	Few tubules lined by sertoli cells	Inhibin and calretinin Positive
Nalini[16]	36	Abdominal mass and Pain	Proliferative endometrium	Fibromatous component	Small tubular cells with round vesicular nuclei & inconspicuous nucleoli	-
Present study	70	Postmenopausal bleeding with abdominal mass	Proliferative endometrium with polyp	Fibrothecomatous component	Sex cord like cells in tubular pattern	-

Usually these tumours are hormonally inactive but presence of lutenized thecal cells or sertoli like cells leads to hormone production, 50% of tumours have estrogenic effect leading to endometrial hyperplasia, diffuse complex atypical hyperplasia or even adenocarcinoma and 11% of cases have androgenic effect.[6,7] Most of the tumours are unilateral with predominantly solid in appearance and attain maximum size of 1-10cm in

diameter. If the tumour is associated with the comatous component the tumour appear solid whitish to yellow in colour.

Microscopically the tumour cells are spindle shaped arranged in interlacing bundles and fascicles with variable amount of collagen and intermingled sex cord elements. These sex cord elements appear either fully differentiated granulosa cells or indifferent tubular structures resembling sertoli cells. The minor sex cord elements shows variable staining positivity for Inhibin but show specific positive staining for Calretinin. Other immunohistochemical markers that are useful in differentiating these tumours from other tumours are positivity for CD56, CD99, Antikeratin antibody k1, MIC and negative staining for Vimentin, SMA and EMA.[5,8]

The common differential diagnosis includes ovarian fibromatosis, Brenner tumour, adenofibroma and metastatic carcinoid.[3,8,9] The common differentiating features are depicted in Table 1. Young and Scully first reported 7 cases of fibromatous tumors of ovary in the year 1983. Out of 7 cases, 5 cases had ovarian fibroma with minor sex cord elements. Out of these 5cases two cases associated with coexisting adenocarcinoma. 2 cases are lutenized thecoma and stromal leydig cell tumours with MSCE.

Out of 50 cases of luteinized thecomas and stromal leydig cell tumours reported by Zhang et al only 2 cases had sexcord elements with granulosa cell morphology.[6]

The following table 2 demonstrates similar individual case reports reported in the literature.

Endometrial changes related to these tumours are attributable to the hormone production either by thecal cells in stroma or by minor sex cord elements. Five cases reported in literature had endometrial adenocarcinoma secondary to hormone production by these cells.[3,9-11] In our case also we had proliferative endometrium with a polyp in a 70 yrs women.

In general, Fibromas are hormonally inactive. But the minor sex cord elements are hormone producing based on the presence of either granulosa cell or sertoli cells. Most of the tumours reported in literature showed estrogenic features but only 2 cases presented with signs of virilization (Sood et al, and Sharma et al,).[12,13] This case also showed estrogenic features with postmenopausal bleeding.

Though there are pure fibromas with MSCE, some case reports of fibroma associated with epithelial ovarian tumours have also been reported. Yang et al, in the year 2001, reported a case of mucinous cystadenoma with stromal tumour with minor sex cord elements in 58yr old women.[14] In 2014, Aswari et al, reported reported a case of serous cystadenoma with coexisting stromal tumour with sex cord elements in an 18yr old unmarried girl.[15]

Two cases of diffuse stromal proliferation with minor sex cord elements in epithelial ovarian tumour have also been reported in literature. Dillon et al, reported in serous cyst adenofibroma with stromal sex cord elements whereas Ueda et al, reported in mucinous epithelial ovarian tumour with stromal sex cord elements.[16,17]

Most of the tumours reported in literature are unilateral tumours except in one literature bilateral ovarian fibroma with minor sex cord elements are reported (Gupta et al).[18] Few cases in extraovarian site also reported as fibroma with minor sex cord elements in broad ligament (Omori et al).[19]

CONCLUSION

This case is being presented for its rarity. Minor sex cord elements in ovarian stromal tumours are prognostically insignificant as these tumours are benign in nature. But extensive histopathological examination is required in these tumours as the potential source for estrogen production can occur from these minor sex cord elements which leads to endometrial changes so that patient may be regularly followed up by the clinician.

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