Case Report

Ovarian fibroma with minor sex cord elements — an incidental finding

Rashmi M V[1], Dhanya A N[2], Ankita Das[2]

Ovarian fibroma with minor sex cord elements is rare tumor of benign nature with less than 20 cases reported till date. Our case was a 55yr old female, who presented with dysfunctional uterine bleeding and a solid ovarian mass was diagnosed incidentally on scan. The tumor was histopathologically termed as ovarian fibroma with minor sex cord elements. A knowledge of this entity and careful examination for the sex cord elements in ovarian fibromas and thecomas is needed so that these tumors are not missed or underdiagnosed. [*J Indian Med Assoc* 2019; **117:** 33 & 35]

***Key* words : Ovarian fibroma, minor sex cord elements.**

Ovarian fibroma with minor sex cord elements is a fibrothecomatous tumor containing <10% of sex cord elements. These tumors occur in women of any age and are usually hormonally inactive. They are solid tumors indistinguishable from fibromas and thecomas. Histopathologically, they show the features of fibroma or thecoma with sex cord elements appearing as fully differentiated granulosa cells to undifferentiated tubular structures similar to immature sertoli cells[1].

Case Report

A 55 year old female presented with menstrual irregularities. On ultrasound scan, a solid ovarian mass was reported. The patient underwent surgery and we received hysterectomy specimen with unilateral adnexa. Grossly, the uterus appeared normal. The ovarian mass measured 12 x 11 x 5 cms. External surface was nodular and cut section showed homogenous grey-white areas. Histopathology, showed thin spindle shaped cells arranged in fascicles. The cells were uniform with bland nuclei and scant cytoplasm. No mitotic figures were seen. Admist these fibroblasts were islands of sex cord stromal cells arranged in tubules and nests. They composed less than 10% of tumor (Fig 1). A diagnosis of ovarian fibroma with minor sex cord elements was made.

The incidence of ovarian fibroma is 4% of all ovarian tumors[2]. The occurrence of ovarian fibroma with minor sex cord elements is extremely rare. It is recognized as a distinct entity by WHO[1]. It is an uncommon benign tumor occurring commonly in middle aged females. It is usually hormonally inactive[2].

It was first described by Young & Scully in 1983[3] as a tumor predominantly composed of fibromatous or thecomatous area and containing minor sex cord elements in less than 10% of the tumor area[4].

This tumor occurs in a wide range of age group, from 16-65 years old females[3]. However, a literature search yielded the occurrence of this tumor in a girl as young as 13 years[5] and as old as 79 years[6].

The tumor is usually hormonally inactive but may a few cases of hormonally active tumors have been reported. Estrogen producing tumors may have endometrial hyperplasia or diffuse atypical hyperplasia or even at times adenocarcinoma of the endometrium[3,6,7]. It may be associated with virilisation in females. These patients present with deepening of voice, hirsutism, clitoromegaly or breast atrophy[6].

Most of these tumors present clinically with pain abdomen or bleeding per vagina or abdominal mass[4,7]. On ultrasound scanning, the size of the tumor varies from 1-10 cms. However, tumors as large as 20 cms have been reported. They are solid, firm, grey-white tumors and have a yellowish – white appearance whenever thecoma component is predominant[6,7].

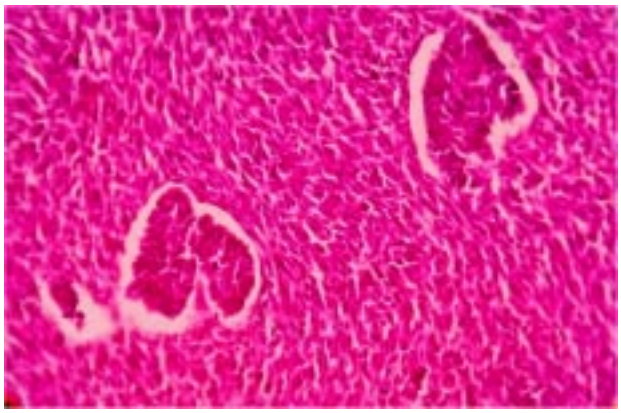

Fig 1 — Fascicles of spindle cells with islands of sex cord stromal cells arranged in the form of tubules and nests (H&E, 20X).

Microscopy shows a well – encapsulated mass with predominantly a fibromatous or a thecomatous tumor with minor component of sex cord elements occupying <10% or not more than 0.45mm of the tumor area[2,7]. The minor sex cord elements may be seen in nests or tubules and composed of cells resembling granulosa cells, sertoli cells, undifferentiated cells or steroid cells of sex cord type[3,5].

On staining for reticulin stains, a pericellular pattern in the spindle cell area with grouping around the sertoliform cells can be appreciated[5].

The sex cord elements are immunophenotypically positive for inhibin, calretinin, CD99, CD56, antikeratin antibody KL1 & MIC. They are negative for SMA, CK, EMA & vimentin[4,5,7].

The spindle cells are positive for vimentin, smooth muscle actin, and weakly for inhibin. They are negative for calretinin, CD34 & CK[4,5].

The nearest differential diagnoses are ovarian fibromatosis, Brenner tumor and adenofibroma[7].

There is proliferation of spindle cells with abundant collagen formation and focal edematous areas with preservation of normal follicular structures of the ovary in ovarian fibromatosis whereas they are replaced by fibrous tissue in ovarian fibromas[4,6].

The epithelial rests in Brenner tumor have cystic lumina with

(Continued on page 35)

Department of Pathology, Sri Siddhartha Medical College, Tumkur, Karnataka 572104
[1]MBBS, MD, Professor and Corresponding author
[2]MBBS, Postgraduate

(Continued from page 33)

eosinophilic secretion. Also the presence of nuclear grooves is an indication of transitional epithelial derivation. They can be further distinguished immunohistochemically. Brenner tumors are positive for CK & EMA, whereas they are negative in OST with MCE[3,9].

In adenofibromas, abundant, large tubular glands of variable sizes are seen in contrast to uniform tubules seen in OF with MSC elements. Immunohistochemically, both inhibin and calretinin are negative in adenofibromas[3].

Many studies have reported these tumors to be of benign nature and without any prognostic significance[2-4]. However, a co-existent endometrial carcinoma may be seen as reported by a few authors[6,7].

Conclusion

OF with MSC element is a distinct clinicopathologic entity. A knowledge of this entity and a thought to look for these minor sex cord elements in all ovarian fibromas or thecomas is needed so that these tumors are not under reported. They need to be differentiated from the nearest differentials and immunohistochemical staining may be essential to differentiate them. An association with other endometrial pathologies needs to be ruled out.

References

1 Tavassoli FA, Mooney E, Gersell DJ — Stromal sex cord tumours. In: Tavassoli FA, Devilee P, eds. WHO pathology and genetics of tumours of the breast and female genital. 1st ed. Lyon: IARC Press, 2003: 152-3.

2 Arasi E — Ovarian fibroma with minor sex cord elements – A rare case report. *IJRHS* 2016, **4:** 24-6.

3 Giriyan SS, Honakeri KR, Gupta D — Ovarian stromal tumor with minor sex cord elements – A case report and review of literature. *IOSR-JDMS (Internet)* 2016; **15:** 79-83. Available from: e – ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 15, Issue 6 Ver. IV (June.2016), PP 79-83.

4 Mandal S, Mahajan D, Roy S, Singh M, Khurana N — Fibroma with minor sex cord elements – an incidental finding in a normal sized ovary A case report with literature review. *Diagnostic Pathology* 2007; **2:** 46.

5 Sood N, Nigam JS, Goyal G, Ranjan R — Virilizing ovarian fibrothecoma with minor sex cord elements in a 13 year old girl: a rare case. *Pediatric Reports* 2014; **6:** 5447: 37-9.

6 Kumar S, Mathur S, Subbaiah M, Singh L — Ovarian stromal tumor with minor sex cord elements with coexistent endometrial carcinoma. *Ind J Med Paediatr Oncol* 2013; **34:** 44-6.

7 Shilpa G, Gomathy E, Harendra Kumar ML, Anil Kumar Sakalecha M — A rare benign tumor and an associated malignancy: Case report of ovarian fibroma with minor sex cord elements and uterine endometrioid adenocarcinoma. *J Obstret Gynecol Ind* 2016; **66:** S632-S635.

8 Sharma R, Vij A, Mahajan N, Chaudhary U, Sharma A, Kumar A — Ovarian stromal tumor with minor sex-cord element with virilizing menifestations: a rare case report. *Int J Repord Contracept Obstet Gynaecol* 2015; **4:** 2113-6.

9 Ross JA, Salgam O — Stromal overgrowth in a Brenner tumor or ovarian fibroma with minor sex cord elements? *Cancer control* 2015; **22:** 366-8.