

## Sex cord stromal tumours of the ovary, experience at Shifa International Hospital Islamabad

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### Abstract

This descriptive study was carried out at Pathology Department, Shifa International Hospital from 2007 to 2016; all sex cord stromal tumours diagnosed during this time period were included. Epithelial, germ cell and metastatic tumours were excluded from the study. A total of 1254 Ovarian tumours were brought to Shifa of which 47 (4%) were labeled as sex cord stromal tumours. Of these 36 (76 %) were granulosa cell tumour (adult 33, juvenile 3), 7 were labeled as sertoli leydig cell tumours (15%), 3 as thecoma/ fibroma group (7%) and only one case was labeled as microcystic stromal tumour of the ovary (2%). Overall age range for sex cord stromal tumours was 42 (12-71). Immunohistochemistry was done in 41 out of 47 cases.

Sex cord stromal tumours of the ovary are rare tumours comprising 4% of the total. Adult Granulosa cell tumour is the commonest tumour seen in our study.

**Keywords:** Sex cord, Tumours, Ovary.

### Case Series

This study was planned at a tertiary care hospital over a time period of nine years, from 2007 to 2016, all the cases of ovarian tumours diagnosed in the histopathology department were retrieved. Reports of 1254 ovarian tumours diagnosed, were reviewed by two pathologists. Primary or metastatic carcinoma, germ cell tumours and all mesenchymal tumours of the ovary were excluded only sex cord stromal tumours were included in the study.

There was no bar on age limit all age groups were included. Slides of sex cord stromal tumours were reviewed and that case where Immunohistochemistry was done was also viewed. Permission from institutional ethics committee was taken.

A total of 1254 Ovarian tumours were diagnosed out of these 47 (4%) were labeled as sex cord stromal tumours. Of these 36 (76 %) were granulosa cell tumours (adult 33, juvenile 3), 7 were labeled as sertoli leydig cell tumours

(15%), 3 as thecoma/ fibroma group (7%) and only one case was labeled as microcystic stromal tumour of the ovary (2%). Overall age range for sex cord stromal tumours was 42 (12-71). Age range for granulosa cell tumour was 15- 71 mean age 48 years, age range for sertoli leydig cell tumours 12- 67 mean age 39 years. The age range for thecoma fibroma group was 33-41 mean age  $37 \pm 1.5$  years. The solitary patient of microcystic stromal tumour was 53 year old.

Immunohistochemistry was done in 44 out of 47 cases in total. Thirty four out of 36 Granulosa cell tumours (32 out of 33 adult and 2 out of 3 juvenile granulosa cell tumours), all cases of sertoli leydig cell tumours, three of fibroma/ thecoma cases and one case of microcystic stromal tumour.

Immunohistochemistry was done on 44 out of 47 cases. Thirty four cases of Granulosa cell tumour were stained for Inhibin, Calretinin and Cytokeratin. All tumours were positive for Calretinin and Inhibin whereas cytokeratin was focally positive in 12 out of 34 cases. Sertoli leydig cell tumours showed diffuse positivity of CK in all seven cases with co-expression of Inhibin. Three cases of thecoma fibroma group was weakly positive for cytokeratin and Inhibin. The microcystic stromal tumour was positive for CK, CD10, WT1 and beta catenin and negative for Inhibin.

### Discussion

Sex cord stromal tumours of the ovary are rare tumours, accounting for 5-7% of the total ovarian tumours. These include granulosa cell tumours, fibromas, thecomas, and sertoli and leydig cell tumours. These tumours can occur singly or in combinations.<sup>1</sup>

Molecular pathogenesis of these groups of tumours is unknown; some have suggested mutation in FOXL 2 gene a transcription factor. However this hypothesis has not been proved.<sup>2</sup> These tumours are largely known to be positive for Inhibin, Calretinin and EMA. A helpful feature in the diagnosis of sex cord stromal tumours is lack of squamous differentiation.<sup>3</sup>

Sex cord-stromal tumours are rare tumours accounting for less than 7% of all ovarian tumours. These usually present in the younger age group.<sup>4</sup> Younger women, 20-30 years

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of age are usually involved. Sertoli-Leydig cell tumours are more morphologically diverse than pure Sertoli cell tumours. The classification proposed by Meyer into well, intermediate, and poor differentiation, remains important prognostically. More recently, heterologous and retiform differentiation has been described. Granulosa cell tumours are most frequently seen. From practical viewpoint, the most helpful immuno-histochemical findings are the negative staining of sex cord tumours for epithelial membrane antigen, and positive staining for Inhibin and Calretinin.<sup>5</sup> Behaviour is usually benign.

We have reported 47 cases of sex cord tumours of the ovary in nine years. Of these, majority were granulosa cell tumours; our data is comparable to what has already been reported in Pakistan by Saroona et al.<sup>1</sup> They have reported 480 tumours over a 20 year period of which majority were adult granulosa cell tumour (43%), which is comparable to our study.<sup>6</sup>

Sertoli leydig cell tumours were the next common group in our series. Sertoli cell tumours account for 4% as reported in international literature.<sup>7</sup>

Thecoma fibroma group is our third most common group; these are approximately one third as common as granulosa cell tumours.<sup>8,9</sup>

We have not reported any case of sclerosing stromal tumour of the ovary however these are reported in the literature.<sup>10</sup>

We have also encountered a rare case of microcystic stromal tumour of the ovary. This tumour was associated with FAP (familial adenomatous polyposis).<sup>11</sup>

## Conclusion

Sex cord stromal tumours of the ovary are rare tumours, granulosa cell tumour being the commonest in our series which is comparable to international and regional data.

Our data shows these have typical morphological and immuno-histochemical features.

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**Conflict of Interest:** None.

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