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A Rare Histopathological Variant of Sertoli-Leydig Cell Tumor with Asymptomatic Bicornuate Uterus

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ABSTRACT

Sertoli-Leydig Cell tumor; a variant of Sex-cord stromal tumor, accounts less than 0.5% of neoplasm of ovary. They are mostly prevalent in second & third decades of life. Literature review documents that histopathologically Sertoli – Leydig cell tumors may be well differentiated, intermediately differentiated, poorly differentiated and some may have heterologous elements. Among the histopathological variants, the intermediate variant has the lowest incidence. In this instance, we report a Sertoli-Leydig tumor with intermediate differentiation. A bicornuate uterus was noticed in this woman as an incidental finding. The incidence of Mullerian duct defects is less than 3%. These Mullerian anomalies are often complicated by repeated pregnancy losses, preterm labor, labor dystocia, dysmenorrhoea. However, this patient had uneventful obstetric history thus clearly indicating that asymptomatic Mullerian anomalies do not require surgical intervention. Only one case of Simultaneous occurrence of ovarian neoplasm with bicornuate uterus as an incidental finding has been reported. We report this case for the rarity of histopathological variant of ovarian neoplasm & associated uterine malformation.

Key words: Sertoli-Leydig cell tumor, Mullerian ducts, Dysmenorrhoea

INTRODUCTION

Sertoli-Leydig cell tumor (SLCT); classified under the category of sex-cord stromal tumors, constitute less than 0.5% of neoplasms of ovary. [1] They are seen commonly during the second and third decades of life and are unilateral. Sertoli-Leydig cell tumor (SLCT) may be associated with DICER1 mutation [2] and have autosomal dominant inheritance pattern. Sertoli-Leydig cell tumor (SLCT); are detected by the presence of testicular structures that produce androgens causing virilisation. About 29% of SLCTs exhibit heterologous elements like glands, cysts etc. [3] In this instance, we present a clinical report of of rare histopathological variant of SLCT in a young women along with endocrine abnormalities along with associated uterine malformation.

CASE REPORT

A 31 year old married women coming from a sub-urban back ground attended the Outpatient department of Gynaecology department of Medical College & Hospital with complaints of facial hair growth with itching for 3 months (Figure 1). She had complaints of amenorrhoea for past 2 years. In addition, she complained of abdominal discomfort and pain. She was married for 10 years with two living issues and last child birth was 7 years back. Her general examination was satisfactory except for the presence of breast atrophy. She had documented a weight loss of 10 kilograms in preceding 3 months. There was recession of temporal hairline. The clitoral index was 140 sq.mm. The sexual maturity rating (SMR) was B5 (atrophy), P5. There was no change in voice or increase in libido. However, the patient

had muscular phenotype (+) with increase in muscle power. The serum testosterone was 14.88 nmol/L (0.38-1.97nmol/L). The DHEA was 254.5 µg/dl (45-270 µg/dl). However, normal FSH & CA-125 excluded ovarian failure & epithelial ovarian malignancy. However, Computed tomography of abdomen revealed right adnexal space occupying lesion (55X34 mm) along with a bicornuate uterus. Based on the above, clinico-radiological findings a strong suspicion of androgen producing neoplasm was there. Exploratory laparotomy was done and right sided salpingo-oophorectomy was done. Histopathological examination revealed a Sertoli-Leydig cell tumor grade II (sertoli cells in lobular aggregates) with intermediate differentiation. No Capsular breech was seen (Figure 2).No heterologous elements were noticed. However, a bicornuate uterus (Left cornu= 89.4x33x56.4 mm) was noticed but the lady had no history of obstetric complications thus clearly indicating that asymptomatic Mullerian anomalies do not require surgical intervention. The virilisation symptoms disappeared after the surgical intervention (Figure 3). No postoperative adjuvant radiotherapy or chemotherapeutic drugs were prescribed. The patient was advised follow-up of 12 months.

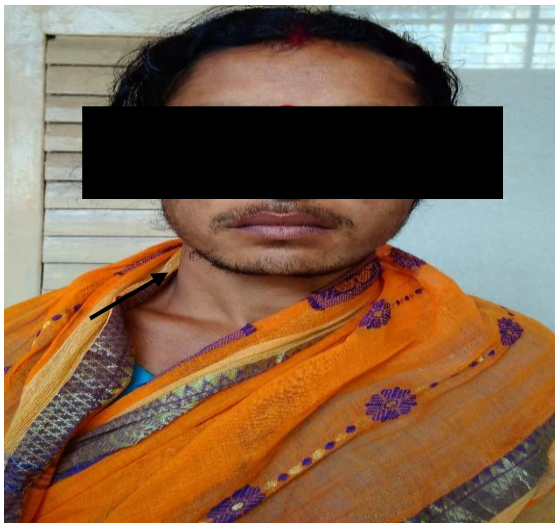

Figure 1. Showing course facial hairs.

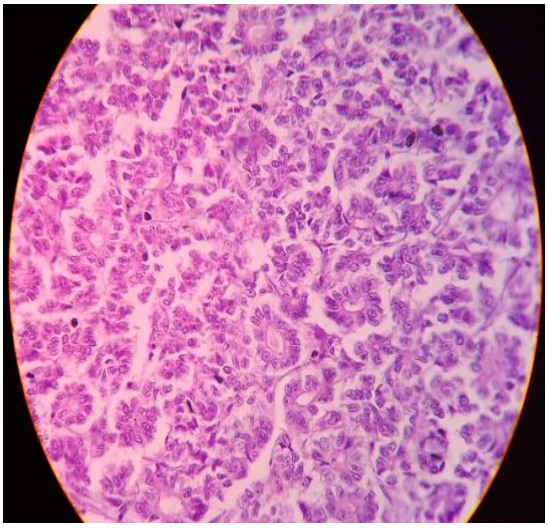

Figure 2. Showing Histopathological variant of SLCT

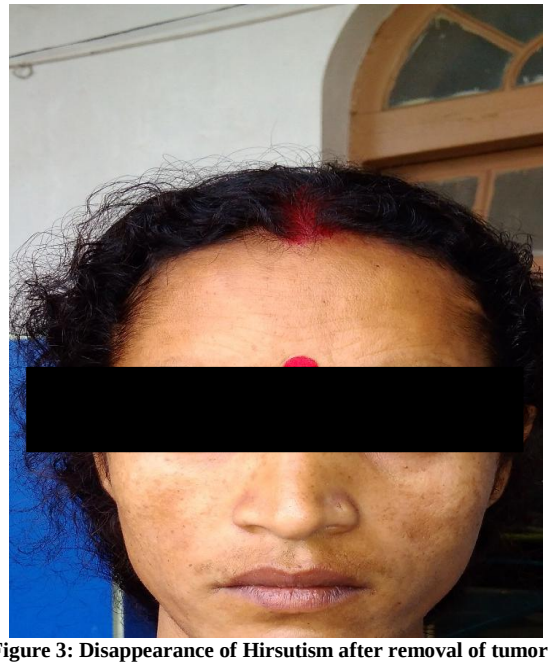

Figure 3: Disappearance of Hirsutism after removal of tumor

DISCUSSION

Literature review suggest that there is limited information about the clinical behaviour and outcome of SLCT due to very low incidence of the tumour (<1%). The tumour is mostly unilateral and one third of cases may suffer from symptoms pertaining to excess of androgen. A case-series study Gui et al suggested that the virilisation is conversely related to the degree of differentiation where as in our case report it was a reverse finding. [4] It was a well-differentiated tumour along with virilisation symptoms corroborative with the findings of Roth et al. [5] There are various school of thoughts about the treatment protocol of the treatment. The important prognostic factors for the disease recurrence

are grade and stage of this rare neoplasm. As this disease is mostly noticed in reproductive age group female fertility sparing surgery is the main treatment protocol. The rate of malignant transformation is almost 11% in poorly differentiated tumours and very high in tumours with heterologous elements. [6] There are however chances of malignant transformation to exist in intermediate variety. [7] However, in this case the patient was kept on follow up and all the virilisation symptoms disappeared. Moreover, the post-operative levels of serum testosterone also reduced to baseline levels. Moreover, the actual prevalence of Mullerian duct anomalies may be difficult to assess due to asymptomatic nature. [8] They are noticed in investigating infertility. Routine pelvic USG or CT-scan has lower detection rate in comparison to pelvic MRI. [9] Rarely, the SLCT may secrete estrogen and may be well associated with endometrial carcinoma, till date 2 cases have been reported. [10] To the best of our knowledge, this is the first case in the English literature of with a Sertoli-Leydig cell tumor of intermediate differentiation co-existing with Bicornuate uterus.

CONCLUSION

SLCT should be excluded in young females with virilisation symptoms. Management protocol depends on the histopathological variant of the tumor. Tumors with intermediate differentiation need an individualized treatment protocol. The actual prevalence of asymptomatic Mullerian anomalies requires proper investigation. Moreover, proper follow up is required in such patients as they have impending risk to develop endometrial malignancy.

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Conflict of Interest: The authors have no conflict of interest

REFERENCES

1. Young RH, Scully RE. Ovarian Sertoli-Leydig cell tumours. A clinicopathological analysis of 207 cases. Am J SurgPathol. 1985;9:543–569.
2. Kock L, Terzic T, McCluggage WG, et al. DICER1 mutations are consistently present in moderately and poorly differentiated Sertoli–Leydig cell tumors. Am J SurgPathol. 2017; 41:1178–87.
3. Lantzsch T, Stoerer S, Lawrenz K, Buchmann J, Strauss H-G, Koelbl H. Sertoli-Leydig cell tumour. Arch Gynecol Obstet. 2001;264:206–208.
4. Gui T, Cao D, Shen K, Yang J, Zhang Y, Yu Q, *et al.* A clinicopathological analysis of 40 cases of ovarian Sertoli-Leydig cell tumors. GynecolOncol 2012;127:384-9.
5. Roth LM, Anderson MC, Govan AD, Langley FA, Gowing NF, Woodcock AS. Sertoli-Leydig cell tumors: A clinicopathologic study of 34 cases. Cancer 1981;48:187-97.
6. Zhang M, Cheung MK, Shin JY, Kapp DS, Husain A, Teng NN, *et al*. Prognostic factors responsible for survival in sex cord stromal tumors of the ovary–an analysis of 376 women. GynecolOncol 2007;
7. Pellerito JS, McCarthy SM, Doyle MB, Glickman MG, DeCherney AH. Diagnosis of uterine anomalies: Relative accuracy of MR imaging, endovaginal sonography, and hysterosalpingography. Radiology 1992; 183:795-800.
8. O'Hern TM, Neubecker RD. Arrhenoblastoma. Obstet Gynecol 1962; 19: 758-70.
9. Elford KJ, Spence JE. The forgotten female: Pediatric and adolescent gynecological concerns and their reproductive consequences. J PediatrAdolescGynecol 2002;15:65-77.
10. Akman L, Ertas IE, Gokcu M, Terek MC, Sanci M, Sanli UA, Zekioglu O, Ozsaran AA. Ovarian sertoli–leydig cell tumors: A multicenter long-term clinicopathological analysis of 27 patients. J Can Res Ther 2016; 12:290-4

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