

# Poorly differentiated Sertoli-Leydig cell tumor of ovary: A diagnostic challenge

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## ABSTRACT

Sertoli-Leydig cell tumors (SLCTs) account for less than 0.5% of all ovarian tumors. The patients can present with androgenic as well as estrogenic manifestations. Poorly differentiated tumors are very difficult to diagnose under microscope. Here, we report a rare case of poorly differentiated SLCT (Meyer's type III) presenting with androgenic manifestations. Histopathologically, we got a variety of features from which the final diagnosis was quite impossible. Preoperative clinical features, hormonal level, and immunohistochemistry played a great role in finalizing the diagnosis.

**Key words:** Ovary, Sertoli-Leydig cell tumor, virilization

## INTRODUCTION

Sex cord-stromal tumors account for approximately 8% of all ovarian tumors, with fibromas accounting for approximately half the cases.<sup>[1]</sup> Sertoli-Leydig cell tumors (SLCTs) account for less than 0.5% of all ovarian tumors. These are mostly (97%) unilateral.<sup>[2]</sup> SCLTs occur in all age groups but are encountered most often in young women. The most striking mode of presentation of SLCTs is virilization. But the patients can present with estrogenic manifestations.<sup>[1]</sup> Poorly differentiated tumors may be hard to distinguish from pure sarcomas and the diagnosis of poorly differentiated SLCT should always be strongly considered when the diagnosis of a pure sarcoma is being entertained. Most otherwise, featureless poorly differentiated SLCTs have small diagnostic areas composed of clusters of dark blue Sertoli cells and some cells that are consistent with Leydig cells or their precursors that will confirm the diagnosis.<sup>[3]</sup> The prognosis of SLCTs is correlated most meaningfully with the degree of differentiation and

stage of the tumor.<sup>[4]</sup> Here, we report a rare case of poorly differentiated SLCT (Meyer's type III) presenting with androgenic manifestations in a young woman without having any definitive diagnostic area histopathologically.

## CASE REPORT

A 29-year-old, married, nulliparous female presented with secondary amenorrhea, vague abdominal pain, and lower abdominal fullness for last 2.5 years. On clinical examination, hirsutism (on face, lip, breast, abdomen), deepening of the voice, laryngeal protuberance, and clitoromegaly were noted. She was not sexually active. Abdominal examination revealed a swelling in the pelvic region. Pelvic ultrasound revealed 14 cm × 8 cm cystic space occupying lesion (SOL) with multiple septae in the right adnexal region. A computed tomography (CT) scan of the abdomen and pelvis demonstrated a complex pelvic mass measuring 16 cm × 8.5 cm that appeared to arise from the right adnexa. The adrenal glands were normal both by ultrasound and CT. Routine blood counts and liver function test were also normal. Serum total testosterone and free testosterone level were elevated and were 23 nmol/l (normal: 0.52-2.4 nmol/l) and 120 pmol/l (normal: 3.5-29.5 pmol/l), respectively. Serum thyroid stimulating hormone (TSH), estrogen, and progesterone level of the patient were within normal range. The urinary 17-ketosteroid values were also normal. On the third postadmission, day, right salpingo-oophorectomy was done. A 17 cm × 9 cm solid cystic mass [Figure 1a] with

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intact but congested surface was received in the pathology department. On cut-section, the mass showed partly cystic and partly solid areas along with areas of hemorrhage and necrosis. Blood-mixed mucoid material came out on cutting the cystic spaces. The specimens were routinely processed and stained with hematoxylin and eosin (H and E). We got a variety of histopathological features from various parts of the tumor mass [Figures 1b-d and 2]. The sections mostly demonstrated a tumor mass formed by short spindle and oval to round cells forming fascicles and storiform pattern. Areas mimicking sheets of lipoblasts, round cell tumor, sarcomatoid carcinoma, sarcoma, seminoma, and hemangiopericytomatous patterns were also noted. Cytoplasm of the cells was clear as well as eosinophilic. Mitotic figures were very conspicuous in most of the areas (>20/10 high power field (HPF)). No specific cellular differentiation was identified. Immunohistochemistry gave diffused cytoplasmic positivity with inhibin, suggesting its sex-cord stromal origin. Finally; considering the patient's age, clinical features, hormonal parameters, gross, histopathological features, and immunohistochemical staining pattern of the tumor mass; the diagnosis of poorly differentiated SLCT (Meyer's type III) was made. The patient received four cycles of adjuvant chemotherapy consisting of cisplatin, etoposide, and bleomycin (PEB). Most of the virilizing manifestations regressed within 6 months. The patient resumed normal menstruation after 4 months. Serum testosterone level also came down to normal range.

## DISCUSSION

Based on a comprehensive review of 207 cases, Young and Scully<sup>[4]</sup> opined that the majority of individuals (97.5%) of ovarian SLCTs presented with stage I disease. Furthermore, on histological analysis, 11% were well differentiated,

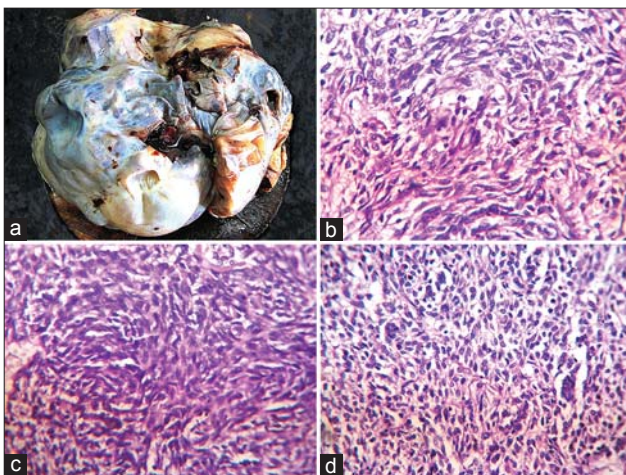
54% intermediate, 13% poorly differentiated, and 22% heterologous. They also observed that 100% of individuals with the poorly differentiated type recurred with a survival rate of 30% only.

In another study, McGuire *et al.*,<sup>[5]</sup> found that 11% of SLCTs with intermediate differentiation and 59% of those with poor differentiation were malignant, with 10-year survival rates of 87 and 41%, respectively. Gui and colleagues<sup>[6]</sup> concluded that patients with high risk factors (intermediate or poor differentiation, beyond stage I, retiform pattern, or with heterologous elements) should receive adjuvant chemotherapy and long-term follow-up.

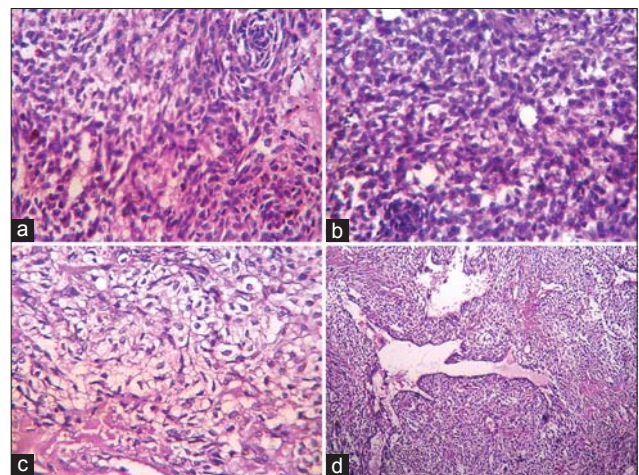
SLCTs are rare hormone producing tumors that must be considered when a young woman presents with virilizing manifestations and a pelvic mass. If the tumor is poorly differentiated, chemotherapy is required in addition to surgical management.<sup>[7]</sup>

Misdiagnosis most often occurs in patients with poorly differentiated tumors with or without heterologous elements. Histologic appearance may be indistinguishable from that of a pure sarcoma, carcinosarcoma, granulosa cell tumors, primary or metastatic endometrioid stromal sarcomas, alveolar area mimicking dysgerminoma, carcinoid tumors, and many more. However, the application of immunohistochemistry can improve the diagnostic accuracy to a greater extent.<sup>[1,8]</sup>

A long list of differential diagnosis<sup>[7,9]</sup> consisting of poorly differentiated Sertoli-Leydig cell tumor (SLCT), sarcomatoid variant of adult granulosa cell tumor, luteinized thecoma, mitotically active fibroma, fibrosarcoma, and endometrial stromal sarcoma (ESS) of ovary was considered initially



**Figure 1:** (a) Ovarian Sertoli-Leydig cell tumors with solid and cystic components, (b) Tumor mass showing spindle cells with high mitotic counts (H and E,  $\times 400$ ), (c) Short spindle cells arranged in storiform pattern (H and E,  $\times 400$ ), (d) Oval to round cells with clear cytoplasm (H and E,  $\times 400$ )



**Figure 2:** (a) Oval to round cells with eosinophilic cytoplasm (H and E,  $\times 400$ ), (b) Small round cell tumor like areas in the tumor mass (H and E,  $\times 400$ ), (c) Sheets of lipoblasts like areas (H and E,  $\times 400$ ), (d) Hemangiopericytomatous pattern (H and E,  $\times 100$ )

in our case. Granulosa cell tumors mostly present with estrogenic features not with androgenic features. Fibroma, fibrosarcoma and ESS are commoner in higher age group. The tumor specimen was not yellow and solid like luteinized thecoma or white and solid like fibroma. Characteristic thick-walled vessels of ESS resembling spiral arteries with surrounding neoplastic cells mimicking endometrial stromal cells were typically absent in this tumor. Conspicuous mitotic figures as in the present case are not a feature of luteinized thecoma. As there was no definitive area favoring the diagnosis of SLCT, we had to face a diagnostic challenge to arrive at the final diagnosis. But the clinical features, hormonal study and finally immunohistochemistry helped a lot to solve the diagnostic problem.

## CONCLUSION

Poorly differentiated SLCT can be confused with many other tumors of the ovary. The diagnosis is especially important from therapeutic point of view.

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