

# Sertoli-Leydig cell tumour of ovary with heterologous elements—A case report

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## Citation

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## Abstract

Sertoli-Leydig tumors are rare tumors of the ovary, occurring in young women in the majority of cases. They constitute less than 0.5 % of all ovarian neoplasms and 1% of sex-cord tumors. Here, we report a case of a 26 year old multiparous woman presenting with abdominal lump, pain, secondary amenorrhea and virilizing signs and symptoms. A diagnosis of Sertoli-Leydig cell tumors of the ovary of intermediate differentiation with heterologous elements was given. Unilateral salpingo-oophorectomy followed by Bleomycin, Etoposide and Cisplatin combined chemotherapy was given. The patient was healthy and symptom-free at the 6 month follow-up.

## INTRODUCTION

Sertoli-Leydig cell tumors of the ovary have been defined by World Health Organization (WHO) as tumors composed of variable proportions of Sertoli cells, Leydig cells and in the case of intermediate and poorly differentiated neoplasms, primitive gonadal stroma and sometimes heterologous elements<sup>1</sup>. These are rare tumors accounting only for about 1% of sexcord-stromal tumors<sup>2</sup> and constitute < 0.5% of all the ovarian neoplasms; intermediate and poorly differentiated forms being the most common. SLCTs have been reported in females from 2-75 years of age with a mean age of 23-25 years in different studies<sup>1</sup>. 75% of the cases present within 30 years. The majority of these tumors are benign, and most are localized unilaterally. Bilaterality is reported in only 2% cases<sup>3</sup>.

It has been reported that about one-third of the patients present with virilizing symptoms and signs like amenorrhoea, hirsutism, breast atrophy, clitoral hypertrophy and hoarseness. Some patients may have estrogenic manifestations like isosexual pseudo-precocity and menometrorrhagia. It has been seen that the androgenic manifestations are more common in the poorly differentiated tumors. 20% cases have been reported to have slightly raised AFP. However, 50% of these patients may have no endocrine symptomatology and instead have abdominal pain or swelling. Heterologous elements are seen in 20% of the SLCTs<sup>4</sup>.

## CASE REPORT

A 26 year old married woman with 4 children, presented with secondary amenorrhea, pain abdomen and lower abdominal fullness for the last 2 years. On examination, hirsutism, hoarseness, breast atrophy and clitoromegaly were noted. Abdominal examination revealed a hard mass of about 20 weeks gestation size in the pelvic region. Ascites was absent. USG revealed a 12 × 8 cm cystic SOL with multiple septae in the left ovarian region. Chest X-ray and ECG were within normal limits. US guided FNAspiration yielded blood mixed mucoid material and microscopically seen to have poor cytological yield with only thick mucinous material. Estimation of androstenedione, by Radio-ImmunoAssay (RIA), showed a high value of 5.38 ng/ml (the normal value for adult female being 0.47-2.68 ng/ml.) The free testosterone level was 5.2 pg/ml. The normal level of free testosterone in cases of adult females usually ranges from 0.7 to 3.6 pg/ml. The estrogen and progesterone levels were within normal limits. A left sided salpingo-oophorectomy was done. Following the histopathology report, combination chemotherapy, usually with Bleomycin, Etoposide and Cisplatin (BEP) was started.

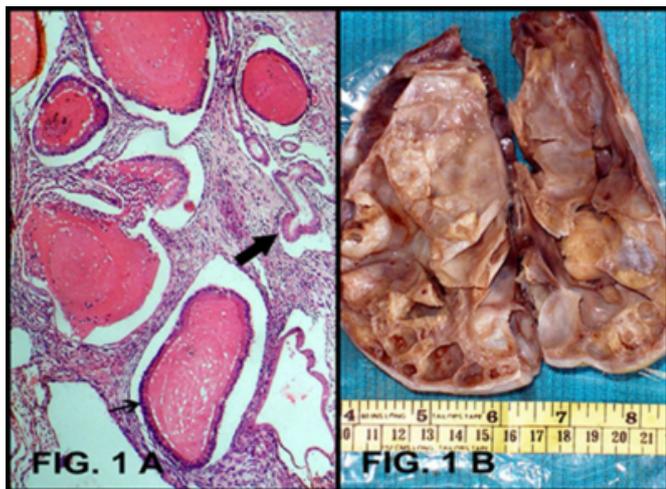
Pathologic findings: Gross examination showed a bilateral ovarian tumor of size 13 × 10 cm. The outer surface was congested. The cut section showed mainly cystic areas with one solid area. The cysts contained mucinous material. Multiple sections were given from representative areas.

Microscopic findings: The sections from the tumor showed

presence of immature Sertoli cells having small, round, oval, or angular nuclei arranged in ill-defined masses, creating a lobulated appearance. The Sertoli cells arranged in solid and hollow tubules, nests and cords reminiscent of the sex cords of the developing testis were seen along with clumps of Leydig cells. Heterologous elements in the form of surface epithelial serous and mucinous differentiation and structures resembling acini were seen. Apart from these hypercellular areas, the stroma was hypocellular, fibrous and loosely edematous at places. Some cyst like spaces with eosinophilic secretions resembling thyroid acini were seen. The epithelium and the secretions were Periodic acid Schiff (PAS) positive. A diagnosis of Sertoli-Leydig cell tumors of the ovary of intermediate differentiation with heterologous elements was given.

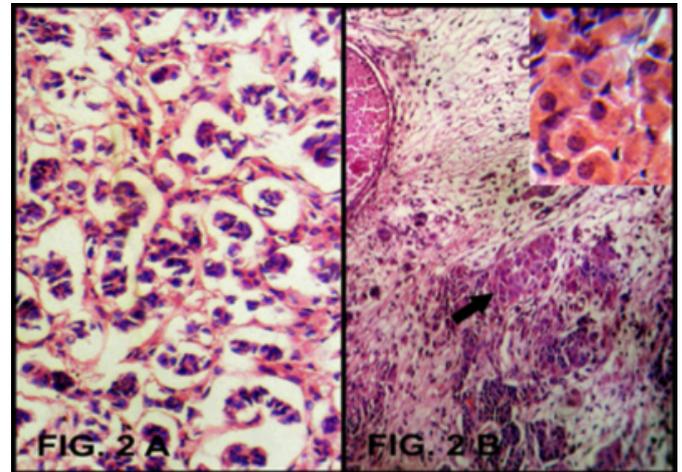
**Figure 1**

Figure 1A : Glandular structures lined by tall columnar epithelial cells without secretions (bold arrow) and with secretions (thinner arrow). Structures resembling thyroid acini seen on upper right (H & E stain, 100x magnification). Figure 1B : Gross appearance of the tumour with multiple cystic areas and few solid areas.



**Figure 2**

Figure 2A: Sertoli cells arranged in tubules and cords (H & E stain, 400x magnification). Figure 2B : Nests of Leydig cells present in an edematous background (H & E stain, 100x magnification).



Inset shows Leydig cells at a higher magnification (H & E stain, 400x magnification)

Following the histopathology report, combination chemotherapy with Bleomycin, Etoposide and Cisplatin (BEP) was started. At 6 months follow-up, the virilizing symptoms and signs were found to have receded to a large extent. There was neither clinical nor sonographical evidence of recurrence and the sex hormone levels had returned to within normal range.

**DISCUSSION**

Sertoli-Leydig cell tumors are rare tumors of the ovary occurring more commonly in young women. The term Sertoli-Leydig cell tumors have replaced the previous terminologies like arrhenoblastomas and androblastomas. Pure Sertoli cell tumors are also included in this group but pure Leydig cell tumors are included under the heading of lipid cell tumors<sup>6</sup>. It is also interesting to note that the Sertoli and not the Leydig cells form the neoplastic component of these tumors<sup>7</sup>. Heterologous elements are seen in 20% of the SLCTs predominantly belonging to the moderately and poorly differentiated categories. The heterologous components may be in the form of mucinous glands, cartilage, bone, skeletal muscle, smooth muscle and fat<sup>4</sup>. However, smooth muscle element is described as rare occurrence<sup>5</sup>. Heterologous mesenchymal elements are usually found in the tumors that have sarcomatoid background. Several theories have been proposed to explain the histogenesis of heterologous elements in Sertoli-Leydig cell tumor. According to Young RH, Kleimen GM and

Scully RE, the histogenesis involves neometaplasia, in which the cells undergoing neoplasia change into other types of cells that are not normally present in the tissue of origin of the tumor<sup>8</sup>. According to Hartz<sup>9</sup>, these tumors are of teratogenic origin. Heterologous mesenchymal elements (skeletal muscle or cartilage) or neuroblastoma imply a poor prognosis while gastrointestinal epithelium or carcinoid as the heterologous element does not have prognostic significance. Some authors opine that although overall SertoliLeydig cell tumors have a relatively good prognosis, intermediate tumors with retiform pattern and those containing heterologous elements have poorer prognosis. Tumor staging is extremely important from the prognostic point of view and guides the treatment too. For young women with stage I tumors, unilateral salpingo-oophorectomy is usually done and if poorly differentiated elements or heterologous elements are present, adjuvant therapy may be indicated with radiation or combination chemotherapy, usually with Bleomycin, Etoposide and Cisplatin (BEP). For tumors of Stage II or higher, a total abdominal hysterectomy with bilateral salpingo-oophorectomy is done and adjuvant therapy is often considered. Our case was treated with left

sided salpingo-oophorectomy and BEP chemotherapy. At 6-month follow up, it was seen that the virilizing symptoms and signs had receded, sex hormone levels normalized and there was no complaint.

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