International Journal of Health Sciences and Research
Vol.10; Issue: 5; May 2020
Website: www.ijhsr.org
Case Report *ISSN: 2249-9571*

Huge Ovarian Sertoli-Leydig Cell Tumor- A Rare Presentation Mimicking Advanced Ovarian Carcinoma: A Clinical Diagnostic Pitfall

Ikeanyi M Eugene[1], Udoye P Ezenwa[2], Jeremiah Israel[1]

[1]Department of Obstetrics and Gynecology, Niger Delta University Teaching Hospital, Okolobiri Bayelsa State Nigeria
[2]Department of Pathology, Niger Delta University Teaching Hospital, Okolobiri Bayelsa State Nigeria

Corresponding Author: Ikeanyi M Eugene

ABSTRACT

Sertoli-Leydig cell tumor is a very rare ovarian tumor constituting less than 0.5% of all primary ovarian tumors. It mostly occurs in second and third decades of life.
This is a case report of a rare presentation of a huge ovarian Sertoli-Leydig cell tumor presenting like an advanced ovarian cancer in a 62 year old seven years postmenopausal para eight woman. At surgery was a left well encapsulated multilobulated ovarian tumour measuring 28 x28x14cm, weighing 6.2kg and histologically containing clusters of Leydig cells and solid cords of Sertoli cells of intermediate differentiation**.** The patient presented with a year history of progressive abdominal swelling and irregular vaginal bleeding. She had total abdominal hysterectomy and bilateral salpingo-oophorectomy. About a year on follow-up and stable.

Keywords: Sertoli-Leydig cell, sex cord, stromal, ovarian, tumor, postmenopausal, neoplasm

INTRODUCTION

Ovarian Sertoli-Leydig cell tumor is one of the categories of sex cord-stromal tumors of ovary; defined by World Health Organization (WHO) as groups of tumors composed of granulosa cells, theca cells, Sertoli cells, Leydig cells and fibroblasts of stromal origin singly or in various combinations. [1,2] Sex cord-stromal tumors constitute about 8% of all ovarian neoplasms and are diverse in the clinical, histopathological, biologic behavior therefore pose diagnostic challenge**.** [3] Sertoli-Leydig cell tumor is very rare, constitutes <0.5% of primary ovarian tumors [4-5] characterized by excessive proliferation of sex cord (Sertoli cells) and stromal (Leydig cells), mostly unilateral and confines to the ovary. [6] It is reported mostly in second and third decades of life [6] but rarely in any age. It can contain heterologous elements and be functionally diverse. It contains testicular structures that secrete androgen with varying degrees of virilization based on the quantity of secreted androgen. [6] It can be benign or malignant based on the degree of its cellular differentiation. A case report of a rare presentation of Sertoli-Leydig ovarian tumor in a 62 year old postmenopausal woman with the clinical misdiagnosis as advanced invasive ovarian cancer is presented.

CASE REPORT

First seen on 25/2/19 was a 62 year old, para 8^{+5} alive 3, LCB 30 years, 7 years postmenopausal Ijaw woman, a petty trader who presented with one year history of progressive abdominal swelling and

irregular vaginal bleeding of about the same duration. She had leg swelling of about six months duration, associated weight loss, easy fatigability, easy satiety and loss of appetite. She attained menarche at 13, never used any form of contraception, was hypertensive, her mother died from complications of breast cancer. She received traditional medications prior to presentation. Repeated breast and abdominal ultrasonography (USS) tests prior to presentation showed breast tumor and features suggestive of ovarian and endometrial carcinoma.

On physical examination, she was emaciated elderly woman, pale, bilateral leg edema up to the knee

Abdomen was markedly enlarged about 36 weeks size irregular in shape, multi-nodular, non-tender, not attached, firm, demonstrable ascites. A Clinical diagnosis of advanced ovarian carcinoma was made.

Investigations were packed cell volume (PCV) 17%, Carcinoembryonic antigen (CEA) 3.7 ng/ml (smoker up to 8.5ng/ml, non-smoker up to 5.0 ng/ml), Cancer antigen 125 (CA 125) was 83.4 (ref 0-35 u/ml) Liver function and renal function tests were normal. Ultrasound scan (USS) imaging investigation showed bulky poorly visualized uterus, abdominal mass suspicious of right ovarian malignancy, right cystic breast mass.

For the preoperative management she was transfused a total of seven units of blood (preoperative PCV -37%) and two units postoperative. She had bowel preparation.

The results and clinical diagnosis were explained to her and she was counseled and consented to the line of management; cytoreductive surgery and chemotherapy.

At Surgery, a general surgeon was in attendance and the intraoperative findings included clean pelvis, straw colored ascitic fluid about 100mls, a huge left well encapsulated multicystic multilobulated ovarian tumor on a thick stalk, smooth surface, no excrescence measuring about 31 x 29 x18 cm and weighed 7.3kg, an enlarged uterus 11x9x6.5cm, grossly healthy looking right ovary, fallopian tubes and other intra-abdominal organs; no peritoneal or intra-abdominal organ deposits or lymph node enlargement. Left salpingoophorectomy was done first (to ensure intact removal of the huge tumor) followed by total abdominal hysterectomy with the removal of right ovary and the tube.

Figure 1a: Left Oophorectomy in Progress

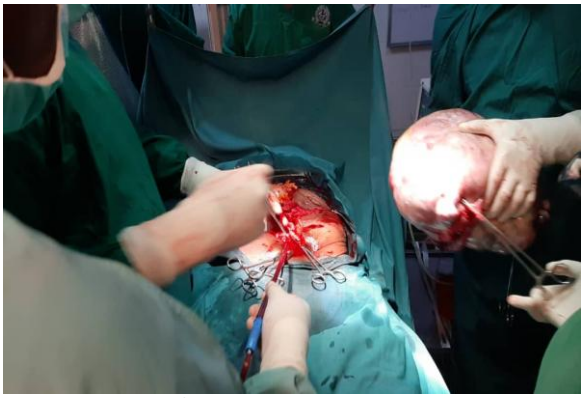

Figure 1b: Left Oophorectomy, Total Abdominal Hysterectomy with removal of the tubes and the Right Ovary in progress

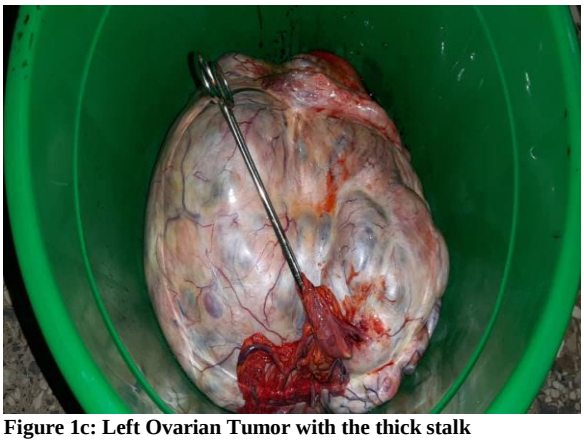

Figure 1c: Left Ovarian Tumor with the thick stalk

Figure 2: A huge left ovarian mass weighing 6.2kg and measuring 28cm x 28cm x14cm

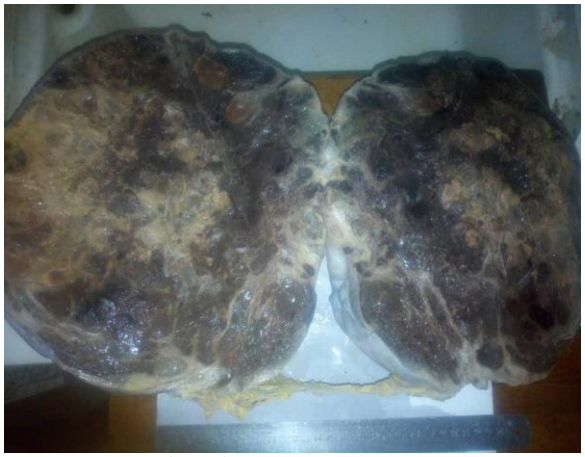

Figure 3: Cut surface of the ovarian mass showing solid cystic and partly haemorrhagic areas

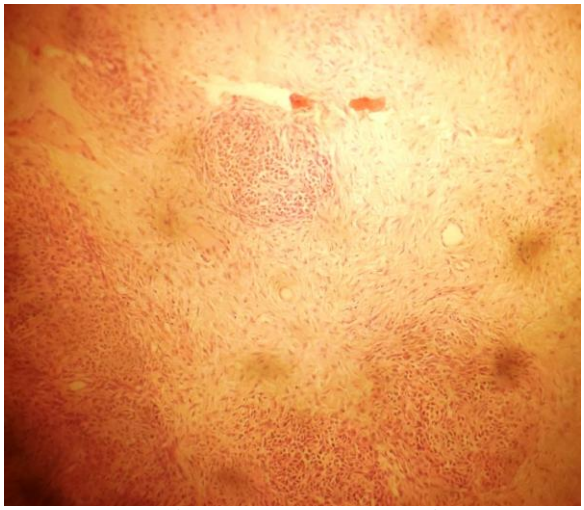

Figure 4a: Nests of Leydig cells at the edges of the aggregates of Sertoli cells adjacent to an edematous area. (H&E x 100).

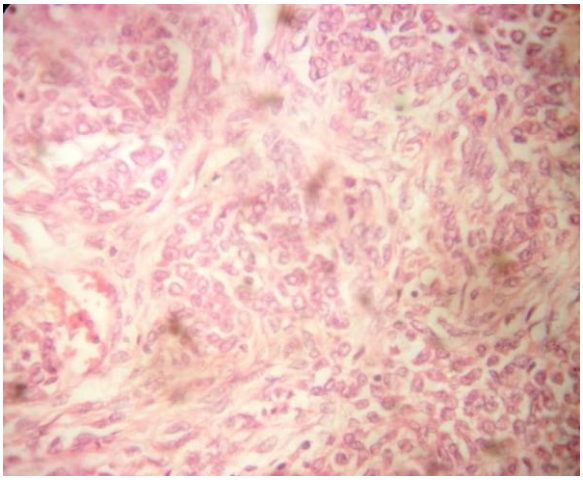

Figure 4b: Solid cords of Sertoli cells surrounding clusters of Leydig cells in their centers (H&E X 400)

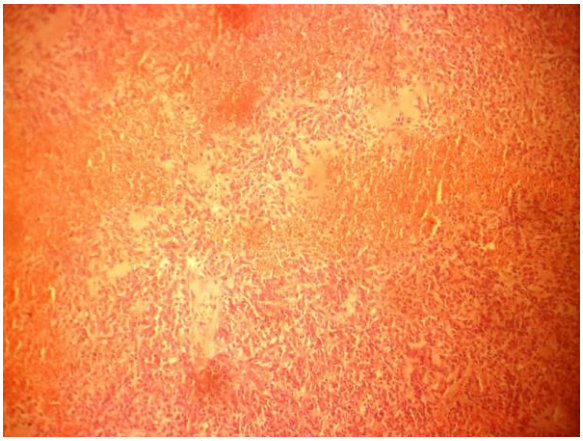

Figure 4c: Areas of hemorrhage and necrosis within the tumor. (H&E x 100)

The histopathology Gross: a huge well circumscribed left ovarian tumor weighing 6.2kg, measured 28 x28x14cm on a thick pedicle (Figures 1, 2) and the cut surface showing solid cystic and partly haemorrhagic areas (Figure 3). Microscopy (Figures 4a-c) show cellular lobules of hyperchromatic spindle –shaped gonadal stromal cells with poorly defined cytoplasm merging with cords and poorly developed tubules at the periphery of cellular lobules. There were very scanty mitotic figures. Some aggregates of Sertoli cells were adjacent edematous areas. Nests of Leydig cells at the edges of the aggregates of Sertoli cells adjacent to an edematous area (Figure 4a) Solid cords of Sertoli cells surrounding clusters of Leydig cells in their centers (Figure 4b)There were wide areas of hemorrhage and necrosis. Areas of hemorrhage and necrosis within the tumor

(Figure 4c). Histopathological diagnosis was Sertoli-Ledig cell tumor (SLCT) of intermediate differentiation. Apart from adenomyoma from the uterine sections, the cervix, endometrium, right ovary and fallopian tubes showed normal histology. Her follow-up is ongoing.

DISCUSSION

Ovarian Sertoli-Leydig cell neoplasm formerly arrhenoblastoma and androblastoma [4,7] is very rare accounting for <0.5% of all primary ovarian tumors. [4-6,8-9] It is characterized by excessive proliferation of Sertoli and Leydig cells in varying proportions and differentiations with the Sertoli cells and not the Leydig cells as the actual neoplastic component. [4-5,7] Though it can occur in any age group, three in four majorities of the cases were reported in second and third decades of life while fewer than one in ten cases in premenarchal and postmenopausal. [4,8,10] The index case was 62 years old and seven years postmenopausal. This was left unilateral tumor confined to the ovary similar to another report [8] and predominantly unilateral [4, 7-8] with only about 2% reported bilateral. [4-5,8] There was mild ascites in the index patient in keeping with the rarity (4%) of ascites in the disorder. [11] The index tumor was grossly well circumscribed and encapsulated, multicystic and multilobulated with intervening fibrous bands consistent with another report [8] on a thick vascularized stalk. Histologically this was intermediately differentiated tumor though it can be well or poorly differentiated [5, 7] retiform or show heterologous elements. [8, 12] There was no ectodermal or mesodermal germ cell heterologous components; rarely seen in about 20% of the cases. [11] The presence of heterologous elements depends on the degree of tubular differentiation of the Sertoli cell component and the amount of primitive gonadal stromal part. [4-5, 7, 11] Alpha fetoprotein (AFP) produced by the Sertoli cells, Leydig cell and heterologous hepatocytic cells is a marker of Sertoli-Leydig tumor mostly detected in about 75% of the under 30 year age group. [4] This however was not tested for in the index case. The presence of germ cell heterologous elements introduces challenges to the diagnosis which is best resolved by immunohistochemistry staining positive to inhibin and calretinin and negative to epithelial membrane antigen (EMA) for SLCT [4,8] but the index case was not subjected to immunohistochemistry test as there was no histopathological diagnosis dilemma. Heterologous differentiation may include mucinous elements as in this case similar to another report, [8] endodermal like cysts and glands, mesodermal like bone, cartilage or skeletal muscle. [9] The sex cord stromal tumors (SCSTs) with heterologous elements tend to be cystic and larger in size relative to the pure SLCT. [4] This was explained in the index case which was multicystic, contained mucin, weighed 6.2kg and measured 28 x28 x14 cm.

The index case presented with abdominal swelling, postmenopausal vaginal bleeding and nonspecific symptoms. The typical clinical presentation is largely related to either abdominal mass, hormonal production, heterologous differentiation or any of the combinations. About 30-50% of patients present with androgenic symptoms of virilization. [4, 9] The index patient did not manifest with any obvious feature of hyperandrogenism; hirsutism, hoarse voice, breast atrophy. However, this patient presented with features suggestive of estrogenic hormone; enlarged uterus, postmenopausal vaginal bleeding. Rarely the case presents with features of excessive estrogen secretion. [5] Ultrasonography was able to diagnose ovarian tumor but not sensitive enough to characteristically specify the diagnosis of Sertoli-Ledig tumor. Other imaging techniques like magnetic resonance imaging may improve the characterization and diagnosis. Histopathological testing is key in the final cellular diagnosis of Sertoli-Leydig cell tumor as was in this case. This was achieved by the identification of clusters of Leydig

cells in interstitial stromal and coexisting proliferating tubules with lining Sertoli cells with varying degrees of mitotic figures. Because there is associated risk of malignancy of 11% and 59% in moderate and poorly differentiated cases respectively, [4,8,10] and up to 80% 5-year survival in Sertoli-Leydig cell neoplasm, the patients will benefit from long-term follow up. [4-5] The follow up for the past ten months with repeat abdominopelvic USS and CA125 has been uneventful.

CONCLUSION

The rarity of Sertoli-Leydig cell tumor, the diverse histo-morphologic and clinical pattern, and heterologous elements make accurate diagnosis challenging with implications for consequent quality therapy and prognostication. The malignant potential necessitates long term follow up of cases.

REFERENCES

1. Tavassoli FA, Mooney E, Gersell DJ, e tal. Sex cord-stromal tumors. Tavassoli FA, Devilee P, eds. *Pathology and Genetics of Tumors of the Breast and Female Genital Organs*. Lyon, France: IARC Press; 2003. 146-62.
2. Mircea R, Anton E, Anton C, et al. Specific features of the stromal ovarian tumors. *Rev Med Chir Soc Med Nat Iasi*. 2012; 116(4):1123-30..
3. Lim D, Oliva E. Ovarian sex cord-stromal tumours: an update in recent molecular advances. *Pathology*. 2018 Feb. 50 (2):178-89.
4. Swati Sharma,Manna Valiathan,Sandeep Kumar,et al. Ovarian Sertoli-Leydig Cell Tumour with Heterologus Elements Masquerading as Mucinous Tumour on Frozen Section: A Case Report. J Clin Diagn Res. 2015; 9(7): ED01–ED03
5. Abu-Zaid A, Azzam A, Alghuneim LA, et al. Poorly differentiated ovarian sertoli-leydig cell tumour in a 16-year-old single woman: a case report and literature review. Case Rep Obstet Gynecol. 2013;2013: 858501.
6. Rimpy Tandon, Poonam Goel, RPS Punia.A Rare Ovarian Tumor-Sertoli-Ledig Cell Tumor with Heterologous Element MedGenMed 2007; 9(4):44
7. Rohini Dhanya CS, Padhi S, Varghese RG. Sertoli- leydig cell tumour of ovary- a diagnostic dilemma. J Clin Diagn Res. 2014; 8(3):127–29.
8. Jonathan E. Zuckerman, Neda A.Moatamed. Sertoli-Leydig cell tumor of the ovary masquerading as a mucinous adenocarcinoma: a frozen section pitfall.Rare Tumors 2017; 3:9(3):6861 doi: 10.40881/rt.2017.6861
9. Young RH. Sex cord-stromal, steroid cell, and other ovarian tumors with endocrine, paraendocrine, and paraneoplastic manifestations. Kurman RJ, Hedrick Ellenson L, Ronnett BM, eds. Blaustein's pathology of the female genital tract. 6th ed. New York: Springer; 2011. p. 785-846.
10. Young RH, Scully RE. Ovarian Sertoli-Leydig cell tumors. A clinicopathological analysis of 207 cases. Am J Surg Pathol 1985; 9:543-69
11. Chen L, Tunnell CD, De Petris G. Sertoli-Leydig cell tumour with heterologous element: a case report and a review of the literature. Int J Clin Exp Pathol. 2014; 7(3):1176–81.
12. Deavers MT, Oliova E, Nucci MR. sex cord-stromal tumors of the ovary. Nucci MR, Oliva E, eds. Gynecologic Pathology. 1st ed: Churchill Livingstone; 2009. p. 445-500.

How to cite this article: Ikeanyi EM, Udoye EP, Jeremiah I. Huge ovarian Sertoli-Leydig cell tumor- a rare presentation mimicking advanced ovarian carcinoma: a clinical diagnostic pitfall. Int J Health Sci Res. 2020; 10(5):135-139.

******