International Journal of Biomedical And Advance Research

ISSN: 2229-3809 (Online)
Journal DOI:10.7439/ijbar
CODEN:IJBABN

Case Report

Pregnancy with recurrent Sertoli- Leydig cell tumour

Bangal V B.[*1], Gupta Kanika[2], Singh Pushpanjali[2], Gavhane S P[3] and Akuskar Roshni[3]

[1]*Professor and head, Dept. of Obstetrics and Gynaecology, Rural Medical College of Pravara Institute of Medical Sciences, (Deemed University) Loni, Maharashtra, India*
[2]*Postgraduate Student (MS), Dept. of Obstetrics and Gynecology, Rural Medical College of Pravara Institute of Medical Sciences, (Deemed University) Loni, Maharashtra, India*
[3]*Assistant Professor Dept. of Obstetrics and Gynaecology, Rural Medical College of Pravara Institute of Medical Sciences, (Deemed University) Loni, Maharashtra, India*

***Correspondence Info:**
Dr. Vidyadhar B. Bangal
Professor and Head,
Dept. of Obstetrics and Gynecology,
Rural Medical College of Pravara Institute of Medical
Sciences (Deemed University), Loni, Maharashtra, India
E-mail- vbb217@rediffmail.com

Abstract

Sertoli -Leydig cell tumor (SLCT) is a rare variety ovarian tumor that belongs to the group of sex-cord stromal tumors. These constitute less than 0.5% of malignant ovarian tumors. A recurrence of Sertoli-Leydig cell tumor of right ovary in a 21-year-old pregnant woman with twenty weeks pregnancy is reported. Oligo-menorrhoea, obesity, mild hirsutism and other features of virilism were associated with the condition. Normal level of testosterone and alpha-fetoprotein (AFP) were found preoperatively. Exploratory laparotomy and right sided salpingo- ovariotomy was performed in fifth month of pregnancy, under general anaesthesia. There were no pelvic or peritoneal metastatic deposits. Histopathology and immuno-histo chemistry confirmed the diagnosis of poorly differentiated Sertoli- Leydig cell tumour. Onco-surgeon and physician were consulted for tumor surveillance and further management during pregnancy. It was decided to start cancer chemotherapy after the delivery of baby. After the laparotomy, patient had uneventful antenatal period. She was delivered by an elective caesarean section at the completion of 38 weeks. A healthy male baby with 3 kg birth weight was born with good Apgar score and without any congenital malformation. A single peritoneal metastatic deposit of 3 to 4 cms in size was found in the pelvic region during caesarean section. This tumour deposits was removed during caesarean section and sent for histopathology. Histopathology report was suggestive of poorly differentiated Sertoli- Leydig cell tumour. Patient was treated with paclitaxel and carboplatin regimen in the postpartum period. This is one of the rare reports on a Sertoli- Leydig cell tumor which showed recurrence and rapid growth during pregnancy.
Keywords: Sertoli-Leydig cell tumor

1. Introduction

Sertoli-Leydig cell tumour is a member of the sex cord-stromal tumour group of ovarian cancers.[1] It arises from male directed cell rests in the hilum of the ovary, from Granulosa cells or from teratomas. Sertoli or Leydig cell tumour can occur at any age, but it occurs most often in young adults. It accounts for less than 0.5 percent of all ovarian tumours.[2] The tumours are either small or medium in size and usually unilateral. Due to excess testosterone secreted by the tumour, one-third of female patients present with a recent history of progressive masculinization. Masculinization is preceded by anovulation, oligomenorrhea, amenorrhea and defeminization. Additional signs include acne and hirsutism, voice deepening, clitoro-megaly, temporal hair recession, and an increase in musculature.[1]

2. Case Report

Twenty years old, second gravida presented with history of 5 months of amenorrhoea and an ultrasound report suggestive of right sided ovarian tumour. Patient had attained menarche at the age of 17 years. Her menstrual cycles were irregular, occurring after every 3 months with menstrual flow lasting for 8 days. She was married for last three years and was treated for ovarian tumour one year back.

One year before the development of recurrence, she had undergone diagnostic laparoscopy and subsequent exploratory laparotomy for pain in abdomen and 6 weeks of amenorrhoea with the provisional diagnosis of ruptured hemorrhagic cyst or ruptured ectopic pregnancy. There was evidence of hemoperitonem due to rupture of right sided ovarian tumour. Considering her age and parity at the time of laparotomy, she had undergone right sided ovarian cystectomy /excision of right sided ovarian mass (13x12 cm solid and cystic tumour) with ovarian reconstruction. Histopathology report of ovarian tumour was suggestive of Sertoli-Leydig cell tumour of intermediate grade (Meyers type II). Her beta HCG (1.20miu/ml and Serum Testosterone values (32.29ng/dl) were within normal limits. Her CA 125 values were 120.2 U/ml. Post operative CT Abdomen did not find any abnormality. Following the diagnosis of Sertoli- Leydig cell tumour, patient received 4 cycles of chemotherapy consisting of Inj. Carboplatin and Paclitaxel. Patient was asymptomatic after the chemotherapy. She conceived after chemotherapy and was diagnosed pregnant during follow up computerized tomography after 4 cycles of chemotherapy. It showed gravid uterus with 6 weeks intrauterine pregnancy and right adnexal cystic lesion measuring 2.9 x 2.0 cm. Patient developed leaking per vaginum and aborted at 16 weeks of gestation. She was advised hormonal contraception for 6 months.

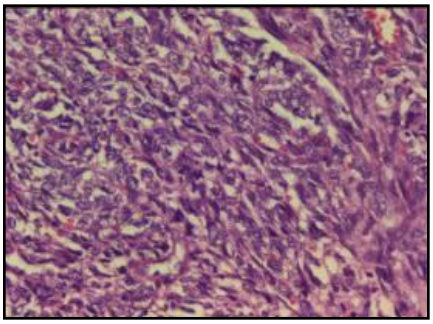
After one year of chemotherapy, patient conceived for the second time after receiving oral ovulation induction drugs. (Tab. Clomiphene citrate.) Ultrasound performed at 14 weeks showed a live fetus of 14 weeks without any evidence of ovarian tumour. Ultrasound examination performed at 18 weeks for anomaly scan revealed 18 weeks viable intrauterine pregnancy with 16.5x7.3x7.7cm soft cystic mass in right adnexa suggestive of recurrence of ovarian tumour. The tumour was hypoechoic in nature, reaching to lateral pelvic wall. Tumour was vascular on colour Doppler examination. It was displacing the uterus to left side. Patient and her relatives were counseled about the need for

removal of the tumour and possible risk of abortion in peri-operative period. She was admitted and investigated further. Her laboratory investigations revealed Haemoglobin of 10.7grams %, Total leucocyte count of 10,000/ cu mm, Liver and renal function tests were within normal limits. Her platelet count was 2.33 lacs, Plasma glucose was 80mg%, PT, APTT and INR values were within normal limits. Serological tests like HIV, VDRL, HbsAg, HCV were normal. Her blood group was B positive. Serum Testosteron level was 59.32ng/dl and Serum alpha- feto protein level was 29.4IU/ml.

Laparotomy carried out under general anaesthesia revealed a tumour of 15cm x 10 cm x 8 cms size,occupying right adnexal region. Peritoneal fluid was collected for cytological examination. Portion of right ovary was looking normal. Tumour was fleshy in consistency, non capsular. There were cystic areas in the tumour. (Fig 1 and 2) It was adherent to pouch of Douglas and anterior rectal wall. Tumour along with right ovary could be removed completely. Pregnant uterus had to be delivered out of abdomen to deal with the bleeding from raw area on anterior rectal wall. Left ovary and tube were normal. There was no other mass or evidence of metastasis in pelvic region or in upper abdomen Omental biopsy was taken. Patient was transfused with one unit of fresh blood during surgery. Prophylactic tocolysis was administered in the form of inj.isoxuprine hydrochloride 10 mg intramuscular every 8 hours for two days. She was given prophylactic antibiotics and analgesics for five days. Histopathology and immuno- histo chemistry confirmed the diagnosis of poorly differentiated Sertoli- Leydig cell tumour. (Fig 3) Onco-surgeon and physician were consulted for tumor surveillance and further management during pregnancy. It was decided to start cancer chemotherapy after the delivery of baby. After the laparotomy, patient had uneventful antenatal period. She was delivered by an elective caesarean section at the completion of 38 weeks. A healthy male baby with 3 kg birth weight was born with good Apgar score and without any congenital malformation. Peritoneal metastatic deposit of 3 to 4 cms in size was found in the pelvic region during caesarean section. This tumour deposit was removed during caesarean section and sent for histopathology. Histopathology report was suggestive of poorly differentiated Sertoli- Leydig cell tumour. Patient was treated with Paclitaxel and Carboplatin regimen in the postpartum period.

Fig 1 and 2 -Tumour specimen after laparotomy

Fig.3 -Histopathology-Undifferentiated Sertoli - Leydig cell Tumour



3. Discussion

Majority of the Sertoli –Leydig cell tumors (SLCT) are unilateral, limited to the ovaries, and are seen below thirty years of age. These tumors are characterized by the presence of testicular structures that produce androgens. As a result, many patients have androgenic features depending on the quantity of androgen production.[2] However; hormonal disturbances in Leydig tumours are present in only 2/3 of cases. Serum testosterone level is high. A conclusive diagnosis is made via histology, as part of a pathology report made during or after surgery. Immuno-histochemical markers of Leydig cell tumours include inhibin-alpha, calretinin, and melan-A.[3]

Another characteristic feature of these tumors is the degree of differentiation of tissue in them. The nature of the tissue determines whether the tumors are benign or malignant.[4] Twenty percent of the tumors show heterologous contents represented by endodermal elements such as cysts and glands and mesenchymal elements such as bone, cartilage or skeletal muscle. A gastrointestinal structure is rarely reported in these tumors.[5,6] The SLCT with heterologous mesenchymal elements are usually poorly differentiated in contrast to neoplasms with endodermal elements, which typically are of intermediate differentiation.[5]

The primary modality of treatment is surgery. Considering the young age of the patients, the surgery usually is a fertility-sparing in the form of unilateral salpingo- oophorectomy. For malignant tumours, the surgery may be radical and usually is followed by adjuvant chemotherapy, sometimes by radiation therapy. The prognosis is generally good as the tumour tends to grow slowly and usually is benign: 10% are malignant. For malignant tumours with undifferentiated histology, prognosis is poor.[7] Adjuvant chemotherapy in stage I is given to those patients who have poorly differentiated SLCT or SLCT with heterologous elements or a metastatic tumor of any histologic type. Chemotherapeutic regimens that are used in the management of these tumours are Cisplatin, doxorubicin, cyclophosphamide (PAC); [8],Vincristine, actinomycin-D, cyclophosphamide (VAC);[9] and Bleomycin, etoposide, and cisplatin (BEP).[10] The BEP regimen is a comparatively safe chemotherapeutic regimen because it does not affect the fertility status of the patient.[11]

Recurrence is known to occur in Sertoli -Leydig cell tumours after initial treatment. Ovarian stimulation by gonadotrophins or other drugs have been associated with recurrence of these tumours. Present case had received clomiphene citrate from a private practitioner for preceding two cycles before conception. There was recurrence and then rapid growth of the tumour in forth month of pregnancy, that co relate well with highest levels of human chorionic gonadotrophins during pregnancy.[7] Pregnancy is a physiological state of rapid endocrine change.

However, the reported case clearly shows that, under certain circumstances, pregnancy-related endocrinological changes can lead to a dramatic increase in tumour growth. Increasing levels of endogenous hCG are thought to be responsible for extremely rapid growth of tumour during pregnancy.

The present case had undergone primary surgery at some other medical college hospital, where it was diagnosed as ruptured ovarian tumour during emergency laparotomy done with the suspicion of ruptured ectopic pregnancy. The discharge card notes suggest that she had weakly positive urine pregnancy test and there was hemoperitoneum with some pelvic mass on ultrasound examination. Diagnostic laparoscopy was done, which revealed a ruptured pelvic tumour with hemoperitoneum. Ovariotomy was done in view of her nulliparity, she was given chemotherapy after histo-pathological diagnosis of Sertoli-Leydig cell tumour. She received three cycles of chemotherapy after the histo-pathlogical diagnosis of poorly differentiated Sertoli- Leldig cell tumour. Second time recurrence was observed during pregnancy in fifth month. Complete tumour was removed at laparotomy during pregnancy. Considering the fetal toxicity of the drugs and nulliparity of the patient, onco-physician advised to give chemotherapy after delivery. Patient and relatives were counselled about it. During caesarean section, whole abdomen was inspected for evidence of metastasis. There was no other lesion except three centimeter mass in right adnexal region and it was removed completely. The histopathology of tumour revealed poorly differentiated Sertoli-Leydig cell tumour. Patient was put on chemotherapy (Paclitaxel plus Carboplatin) after stitch removal and had completed three cycles of chemotherapy without any evidence of tumour recurrence.

In the present case, there was very good compliance from the patient and the relatives. They were counseled regarding the essential investigations for confirmation of the diagnosis, need for continuous monitoring for the recurrence during pregnancy and necessity of chemotherapy after delivery of the baby. Baby was fed on animal milk as breast feeding was contraindicated during cancer chemotherapy.

4. Conclusion

Sertoli –Leydig cell tumour is a rare type of ovarian sex-cord tumor and is usually unilateral. It should be suspected when a young woman presents with menstrual symptoms, features of hyper-androgenism and the presence of ovarian tumour. Management mainly depends on histopathology of the tumor. Poorly differentiated tumors require aggressive management because the chances of them being malignant are high. Recurrence of the tumour during pregnancy is rare. It is managed by surgery. Adjuvant chemotherapy may be started after fetal viability is achieved. Early termination of pregnancy by induction of labour followed by chemotherapy is advocated by some workers. Long term surveillance is necessary.

References

1. Sachdeva P, Arora R, Dubey C, Sukhija A, Daga M, Singh DK. "Sertoli-Leydig cell tumor: a rare ovarian neoplasm. Case report and review of literature". *Gynecol. Endocrinol*.2008; 24 (4): 230–4.
2. Young RH, Scully RE. Ovarian Sertoli-Leydig cell tumours. A clinicopathological analysis of 207 cases. *Am J Surg Pathol*. 1985; 9:543–569.
3. Ulbright TM, Srigley JR, Hatzianastassiou DK, and Young RH "Leydig cell tumors of the testis with unusual features: adipose differentiation, calcification with ossification, and spindle-shaped tumor cells". *Am. J. Surg. Pathol*. 2002; 26 (11): 1424–33.
4. Dietrich JE, Kaplan A, Lopez H, Jaffee I. A case of poorly differentiated Sertoli-Leydig tumour of the ovary. J Pediatr Adolesc Gynecol. 2004; 17:49–52.
5. Lantzsch T, Stoerer S, Lawrenz K, Buchmann J, Strauss H-G, Koelbl H. Sertoli-Leydig cell tumour. *Arch Gynecol Obstet*. 2001; 264:206–208.
6. Mathur SR, Bhatia N, Rao IS, Singh MK. Sertoli Leydig cell tumour with heterologous gastrointestinal epithelium: a case report. *Indian J Pathol Microbiol*. 2003; 46:91–93.
7. Lenhard M, Kuemper C, Ditsch N, Diebold J, Stieber P, Friese K, Burges A "Use of novel serum markers in clinical follow-up of Sertoli-Leydig cell tumours". *Clin. Chem. Lab. Med.* 2007; 45 (5): 657–61.
8. Sood AK, Gershenson DM. Management of early-stage ovarian cancer. In: Bristow RE, Karlan BY, editors. Surgery for Ovarian Cancer: Principles and Practice. London, UK: Taylor and Francis; 2005. 57–86.
9. Gershenson DM, Copeland LJ, Kavanagh JJ, et al. Treatment of metastatic tumour of the ovary with cisplastin, doxorubicin and cyclophosphamide. *Obstet Gynecol*. 1987; 70:765–769.
10. Schwartz PE, Smith JP. Treatment of ovarian stromal tumours. *Am J Obstet Gynecol*. 1976; 125:402–411.
11. Gershenson DM, Morris M, Burke TW, et al. Treatment of poor-prognosis sex cord-stromal tumors of the ovary with the combination of bleomycin, etoposide, and cisplatin. *Obstet Gynecol*. 1996; 87:527–531.