MALIGNANT SERTOLI LEYDIG CELL TUMOR OF OVARY IN A YOUNG ADOLESCENT

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This article is available online at www.ssjournals.com

ABSTRACT

Malignant sertoli leydig cell tumors of ovary are rare gynecological malignancies. When they present in adolescent age group they pose a management problem with respect to conservative or radical treatment. These tumors are characterized by the presence of testicular elements that produce androgens. Hence, many patients have symptoms of virilization depending on the quantity of androgen production. The decision of management is usually based on surgical staging. Proper counselling is necessary to ensure compliance for chemotherapy. We are presenting a case of a 16 yrs old girl who presented with symptoms of virilization.

Keywords: Sertoli Leydig cell tumor, Ovary, Adolescent

1. Introduction

Ovarian Sertoli Leydig cell tumors are rare. These constitute less than 0.5% of ovarian tumors. They are known to produce various hormones like testosterone and androstenedione. About 1/3 rd cases may present with virilisation. They may be mimicked by many different tumors, some of them more common than Sertoli Leydig cell tumors. The main differential diagnosis is endometroid tumor, carcinoid tumor and granulosa cell tumor. Patients with Sertoli-Leydig cell tumors present with signs of defeminization followed by masculinization. Age of the patient, stage of the disease and degree of tumor differentiation based on morphology are the most important factors to consider in the management of the case. Poorly differentiated Sertoli Leydig cell tumor can present a management challenge in young adolescent girls. We are presenting one such case.

2. Case Details

A 16 year old unmarried girl presented with the history of amenorrhea since 1 year and excessive hair growth on face, chest, abdomen and limbs since 1 year. She also complained of change in voice. Her age of menarche was 13 years and previous menstrual cycles were 5-6/30 days, regular with average flow. Her general condition was fair with normal vital signs. She was pale, anicteric with no pedal odema or palpable lymph modes. Her systemic examination was within normal limits. However she had excessive facial hair growth especially on her upper lip and chin. Chest, abdomen and legs also had excessive hair. Her voice was husky with male type overtones. A mass of 14 weeks uterus size was palpable per abdominally with no ascites. Per rectal examination revealed a normal sized uterus with a right forniceal mobile mass of 10 X 10 cm size. Ultrasonography revealed a highly vascular right sided ovarian mass of 10.2 X 9.6 X 6.4 cm size with multiple septations. Her Ca-125 level was 9.30 ng/dl. However testosterone levels were raised (155.96 ng/dl) . Exploratory laparotomy was done. Operative findings showed replacement of the right ovary by a 10x10 cm solid, grey-white, smooth-surface mass with intact capsule. No remnant healthy ovarian tissue was seen. There was no spillage of tumor cells during surgery. The abdominal cavity was explored systematically but there were no deposits anywhere else in the cavity. The para-aortic lymph nodes were not enlarged. The left ovary was found to be normal. Right salpingo-oophorectomy was performed and biopsy was taken from left ovary. Peritoneal washings from all quadrants were sent for cytologic examination for malignant cells. An omental biopsy was also done and sample from parietal peritoneum was sent for histopathological examination. There were no palpable pelvic lymph nodes. Frozen section report was sex chord stromal tumour. Histopathology report was poorly differentiated malignat Sertoli Leydig cell tumor of right ovary. Other samples were unremarkable. Peritoneal fluid cytology showed a cellular protienaceous debris. The patient and her relatives were counseled about various treatment options and the patient was put on chemotherapy

in the form of bleomycin, etoposide and cisplatin (BEP). Six cycles of the multidrug chemotherapy regimen have been administered at 3 weeks interval. The patient has tolerated the chemotherapy well, with no major complications. During one year follow-up, the patient has resumed her periods and virilization symptoms have subsided.Hirsutism was managed with epilation cream and it has not recurred. Repeat testosterone levels on follow-up are within normal range. She is on regular follow up and is doing well.

3. Discussion

Ovarian Sertoli Leydig cell tumors are rare and their morphologic spectrum, behavior and factors influencing the latter are not clearly established[3]. Their incidence is less than 0.5% of all ovarian tumors with mean age of 25 years, 15% present before age of 30 years. They are usually unilateral, 1.5% are bilateral. Their average size is 13.5 cm, with a range from 5-15 cm. Their appearance is variable but usually they do not have much blood filled cysts and are almost never unilocular. Poorly differentiated tumors have more necrosis or hemorrhage. Serum testosterone and androstenedione may be elevated. Urinary 17 ketosteroids levels may be high[4]. Most Sertoli tumors are stage I, unilateral, cytologically bland and clinically benign but occasional examples are higher stage, and about 11% of stage I tumors have worrisome histologic features that may portend an adverse outcome. Histologically they are classified into 5 categories well differentiated, intermediately differentiated, poorly differentiated, heterologous and retiform[5]. Well differentiated tumors usually show a tubular pattern with nodular architecture and fibrous bands separating the lobules. Tubules are lined by round to oval nuclei with minimal nuclear atypia and rare mitotic figures. Stroma shows variable amounts of Leydig cells.

Intermediate or poorly differentiated immature Sertoli cells have small, round oval or angular nuclei arranged in ill defined masses, creating a lobulated appearance. Solid and hollow tubules resembling sex cords of embryonic testes with broad columns of sertoli cells may be seen. Stroma may be composed of immature mesenchymal cells resembling a sarcoma[3].

Because ovarian sex cord stromal tumors are morphologically heterogeneous neoplasms that are relatively infrequently encountered, their diagnosis can be difficult. Immunohistochemical staining may be useful for establishing diagnosis[6].

To determine the prognosis of a patient, stage of the tumor is very important. Stage I a i is found in 80% of tumors, tumor rupture or involvement of external surface of ovary in 12% and ascitis in 4%. Spread beyond the ovary within the pelvis or upper abdomen is found in 2.5%. Poorly differentiated were more often ruptured or presented at a higher stage. (Metastasis is usually found in lung, scalp, supraclavicular lymph nodes and liver). Well differentiated tumors were usually 0-1% malignant, intermediate 11%, poorly differentiated 59%, heterologous 19% and retiform 25%[7].

Unilateral salpingoophorectomy is preferred in young women with stage I tumors. If poorly differentiated elements or heterologous elements are present, adjuvant therapy may be indicated with radiation or combination chemotherapy. In stage II or higher total abdominal hysterectomy with bilateral salphingoophorectomy is indicated and adjuvant therapy may be considered. BEP (Bleomycin, Etoposide and Cisplatinum) is an active combination regimen for first line chemotherapy in such patients[8].

Our case was a young adolescent with malignant Sertoli Leydig cell tumor of right ovary, stage Iai. The most important point of consideration was the age of the patient but the histopathology of the tumour being highly de differentiated required a careful selection of chemotherapy and proper counseling of the relatives to ensure compliance in post operative period. Unilateral salphingoophorectomy with adjuvant chemotherapy consisting of BEP regimen was judged to be the best line of management in her case.

Conclusion

Poorly differentiated malignant sertoli leydig cell tumor of ovary represents a management challenge in a young adolescent. Staging of the disease on laparotomy directs the course of management. Keeping in mind future fertility and young age of the patient unilateral salphingoophorectomy and multidrug chemotherapy is the accepted line of management whereas in higher stages bilateral salphingo-oophorectomy and hysterectomy is required. Proper counseling of the patient and her relatives is mandatory.

References

1. Young RH, Scully RE. Ovarian sertoli-leydig cell tumours. A Clinicopathological analysis of 207 cases. Am j surg pathol1985; 9:543–56.
2. 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.
3. Oliva E, Alvarez T, Young RH. Sertoli cell tumors of the ovary; a clinicopathologic, and immunohistochemical study of 54 cases. Am J Surg. Pathol 2005 Feb; 29(2): 143-56.
4. Tsuji T, Catasus S, Prat J. Is loss of heterozygosity at qq 22.3 (PTCH gene) and 19 p 13.3 (STK 11 gene) involved in the pathogenesis of ovarian stromal tumor? . Hum Patho 2005 Jul; 36(7): 792-6.
5. Mayers K, Chan YF, Hamill J, Massey R.Ovarian Sertoli –Leydig cell tumor with retiform pattern. Pathology 1995 Oct; 27(4): 371-3.
6. Deavers MT, Malpica A, Liu J, et al. Ovarian sex cord stromal tumors: immunohistochemical study including a comparison of calretinin and inhibin Mod. Pathol 2003 Jan; 16(6): 584-90.
7. Homesley HD, Bandy BN, Hurteau JA, et al. Bleomycin; etoposide and cisplatin combination therapy of ovarian granulosa cell tumors and other stromal malignancies. A Gynecologic Oncology group study. Gynecol Oncol 1999 Feb, 72 (2): 131-7.
8. Henry JB.Clinical diagnosis and management by laboratory methods Twentieth Edition, WB Saunders 2001.

Figure 1 Poorly differentiated malignant Sertoli-Leydig cell tumor (40 X)

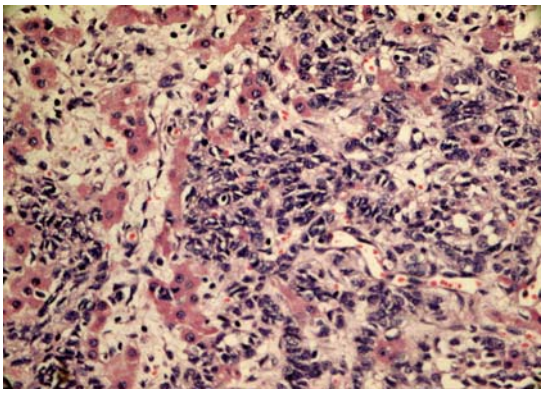

Figure 2 showing Poorly differentiated malignant Sertoli-Leydig cell tumor with cords of immature Sertoli cells and clusters of Leydig cells (100 X)