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A Rare Ovarian Tumour Presenting as a Pathologist's Dilemma and a Surgeon's Nightmare

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ABSTRACT

Gynandroblastoma with juvenile Granulosa cell and Sertoli Leydig cell as components is an extremely rare tumor of sex-cord stromal variety. It's occurrence as a recurrent poorly differentiated Sertoli Leydig cell tumor with heterologous rhabdomyosarcoma component is even a rarer event. A 14 years old girl presented to us in our outpatient department with complaints of a pelvic mass and masculinizing symptoms. She underwent primary cytoreductive surgery at our hospital. The histopathology of the ovarian mass showed both juvenile granulosa cell and Sertoli Leydig differentiation and hence diagnosed as gynandroblastoma. She was given adjuvant chemotherapy and was on regular follow up. The patient developed first recurrence after a disease free interval of 23 months, with an abdominopelvic mass. She underwent secondary cytoreductive surgery for the same. The second histopathology report was suggestive of rhabdomyosarcoma-embryonal type (heterologous differentiation in sex cord stromal tumor). She had multiple relapses and each time the complexity of surgery further increased, and pathology varied. The case presented as a pathologist's dilemma as adjuvant chemotherapeutic regimen changed based on the dominant component of the tumor. Also, as intra-operatively the tumor was extremely vascular, its surgical management proved to be a nightmare for the operating surgeon.

Keywords: Gynandroblastoma; Juvenile granulosa cell tumor; Sertoli-Leydig cell tumor with heterologous component; Sex cord cell stromal tumor; Recurrence

INTRODUCTION

Gynandroblastoma is a rare subtype of ovarian tumor with a combination of granulosa cells and Sertoli Leydig cell tumor [1]. The tumor is usually seen in young females and presents with unique manifestation of estrogenic and androgenic components [2]. For diagnosis of this rare disease the minor tumour component should constitute at least 10% of the entire tumor [3]. Majority of the cases reported till date have shown a benign course with only one recurrence reported, in which the tumour recurred 10 years after the diagnosis [1]. Due to rarity of the disease the biological behaviour of this tumour is unpredictable and there are no clearcut guidelines regarding its management. Here, we present a rare case report of Gynandroblastoma of the ovary with Granulosa and Sertoli Leydig cell component with rhabdomyosarcomatous differentiation that presented with multiple recurrences and variable histopathology at each episode of relapse making the treatment challenging successively.

DESCRIPTION

A 14 years old girl presented in our outpatient department with the complaints of pain and lump in abdomen for two months. She had history of acne over face and back, accompanied by hoarseness of voice. She attained menarche at 13 years of age and had irregular menstrual cycles since then. On examination, she had a large abdominopelvic mass corresponding to 24 weeks size uterus with restricted mobility and clitoromegaly. CECT scan of whole abdomen showed a heterogenous attenu-

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ating mass in the left side of abdomen of size 18 cm × 8.25 cm, indicating the possibility of origin from left ovary. X-ray chest was normal.

DISCUSSION

The value of various tumor markers were as follows: Alpha feto protein (AFP)=167.42 ng/ml (0-10 ng/ml), Beta human chorionic gonadotrophin (BHCG)=<2IU/ml (<5 IU/ml), Lactate dehydrogenase (LDH)=1128 U/L (5-450 U/L), Inhibin B=333.15 pg/ml (22-85 pg/ml) and CA125=510 U/ml (0-30 U/ml). Tumor markers and clinical picture led to a provisional diagnosis of germ cell tumor or sex-cord stromal tumor. Patient was planned for fertility preserving surgery. Intraoperatively there was mild ascites with a left ovarian mass of size 20 cm × 15 cm × 10 cm that was adherent to the omentum. The uterus, right fallopian tube and ovary were normal. The upper abdomen including the liver, spleen and undersurface of diaphragm, peritoneal surfaces were free of any disease. The mass was excised and sent for frozen section. The frozen report suggested possibility of granulosa cell tumor of the left ovary. Staging surgery was completed along with left pelvic and para-aortic lymph node dissection, multiple peritoneal biopsies and omentectomy. On final histopathology along with immunohistochemistry (CK, CD56, CD99 and WT1 positive), diagnosis of Stage 1a Gynandroblastoma i.e., juvenile Granulosa cell tumor with moderate differentiation of Sertoli Leydig cell component was made. In view of rare histopathology, pathologically aggressive looking tumor, and lack of clear evidence in literature, decision was taken in multidisciplinary tumor board (MDT) for adjuvant chemotherapy. Patient received 4 cycles of adjuvant bleomycin, etoposide, and cisplatin chemotherapy regimen. She was kept on regular follow up post chemo.

After almost 2 years, patient presented with complaints of sudden heaviness and pain in the abdomen. She was evaluated for her symptoms and a whole body PET CT scan was done that revealed metabolically active complex mass lesion of size 19.9 cm × 16.4 cm × 7.8 cm in the pelvis encasing the uterus and displacing the bowel loops posteriorly. The mass was extending and abutting the retroperitoneum with resultant right hydroureteronephrosis. In view of localized relapse and a good disease free interval of 23 months, patient was planned for secondary surgery. Intra operatively a large, encapsulated mass of size 20 cm × 20 cm was seen adherent to the anterior abdominal wall and mesentery. The tumor was highly vascular and there were multiple small feeding vessels from mesentery as well as anterior abdominal wall. Utmost care had to be taken in cauterizing the feeding vessels, separating the mass, and not compromising the vascularity of the mesentery. In view of normal uterus and right ovary, no deposits or disease seen elsewhere, and considering the young age of the patient, decision for fertility preservation was taken in the best interest of the patient. The patient required multiple blood transfusions and ionotropic support in intra as well as post-operative period.

The histopathology this time was reported as myxoid tumor with uniform short spindle cells seen in whorls and fascicles with rich vascularity, favoring poorly differentiated Sertoli-Leydig cell tumor with predominant sarcomatoid differentiation. IHC markers were reported as: Desmin, Myogenin-patchy; CD56, Myod1-weak focal; and Calretinin, Inhibin, Cytokeratin, EMA-negative. The final impression of Rhabdomyosarcoma (RMS), embryonal type (heterologous differentiation in sex cord cell stromal tumor) was made. The slides were reviewed at two more oncological centers and diagnosis reconfirmed. Patient then received chemotherapy 6 cycles of vincristine, actinomycin and cyclophosphamide (VAC) chemotherapy considering RMS histology.

After 5 months of completion of VAC regimen she presented in emergency with acute abdominal pain. CECT scan of whole abdomen revealed a well-defined lobulated abdomin-pelvic mass lesion of size 18 cm × 20 cm, insinuating along the small bowel loops and along right lateral wall of urinary bladder that was inseparable from the right adnexa, suggestive of recurrent neoplastic lesion. Patient was reassessed and case was discussed again in tumor board and expert opinion also sorted. The final decision was taken to proceed with surgery without fertility consideration. So, patient was taken up for tertiary cytoreductive surgery that included abdominopelvic mass excision with total abdominal hysterectomy and right salpingo-oophorectomy with right pelvic lymph node dissection. Intra-operatively, there was hemoperitoneum and a highly vascular mass that was already ruptured and adherent to all bowel loops with feeding vessels arising from the bowel mesentery. However, uterus and right ovary were normal. Removing the mass, regulating bleeding from the feeding vessels and simultaneously trying to achieve complete resection was a nightmare for us. Meticulous dissection, careful cauterization, packing and praying helped. Swiftness of the surgery had to be maintained hand in hand as patient was hemodynamically compromised. She again needed multiple transfusions and ionotropic support challenging the anesthesia team too. Histopathology examination was processed along with IHC markers. Inhibin and calretinin, CD99, Myogenin, SF1 were immunoreactive (score 2+), Desmin was immunoreactive to 3+ and CK (AE1+AE3) and WT1 were non immunoreactive. Final diagnosis of recurrent poorly differentiated sex cord stromal tumor-favoring Sertoli-Leydig cell tumor with some rhabdomyosarcomatous element was made. She was planned for adjuvant chemotherapy using paclitaxel and carboplatin regimen.

However, one month post-surgery, she again presented with a solid cystic mass in left hypochondrium of size 13 cm × 9 cm with moderate hemoperitoneum. In view of hemodynamic stability and refusal for surgical intervention by immediate family, she was managed with multiple blood transfusions and supportive care. She was started on adjuvant chemotherapy two weeks later. A fortnight after the first cycle of chemotherapy, she presented in emergency with dizziness and nausea. Hemoglobin was 6 and ultrasound showed gross hemoperitoneum. In view of recurrent hemoperitoneum, chemotherapy could not be continued. Thus, patient was recommended palliative care. After one month, patient expired at home.

Gynandroblastoma is an extremely rare ovarian tumor seen in young women. Sex cord stromal tumors accounts for 5%-10% of all ovarian malignancies and gynandroblastoma accounting for less than 1% of ovarian sex cord cell stromal tumors [4,5]. In 1930, Robert Meyer was the first to bring into knowledge this type of tumor in his studies on arrhenoblastoma. He suggested that ovarian tumors can have indifferent elements and these constituents can lead to feminization and masculinization

characterstics [6]. Clinically, the female presents with variable spectrum of symptoms depicting the hormonal activity of this tumour. The symptoms can vary due to feminizing or masculinizing effects of the tumour depending on the predominance of the ovarian element (granulosa or Sertoli-leydig). Our patient also presented with lump in abdomen, amenorrhea, virilizing features like clitoromegaly, hirsutism and acne.

Gynandroblastoma's biological behaviour is less studied due to its lower incidence and usually follows a benign course as depicted in past studies [7]. In all the cases, the tumour is usually in stage I as was seen in our patient. However, on reviewing the literature, no malignant conversion has been reported in the past. Our patient presented with a recurrence after a disease free interval of 23 months but the histopathology at recurrence was Sertoli Leydig cell tumor with rhabdomyosarcoma differentiation. James Nef et al. conducted a systematic review of relapsed cases of ovarian Sertoli-leydig cell tumors from January 1998 to January 2021. They studied 85 patients from 33 articles with a median age of 20 years and the median time to relapse was 14 months. It was observed in their study, that young patients mostly presented with a poor prognosis in contrast to gynandroblastoma.

CONCLUSION

As compared to previous case reports, our patient presented at a relatively younger age and her initial diagnosis was of gynandroblastoma which on recurrence presented as Sertoli Leydig cell tumor with rhabdomyosarcoma differentiation. Also, our patient had multiple recurrence over a short span of 34 months. These factors depict the unique nature of presentation of this case report. It was a diagnostic dilemma for the pathologist each time and patients further course of treatment entirely dependent on the final words of pathologist. Also, extremely vascular nature of the tumor, and hemodynamic instability of the patient was truly a surgeon's nightmare as the surgical procedure needed to be performed as quickly as possible with the need to control the bleeding meticulously.

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CONFLICT OF INTEREST

The author declares there is no conflict of interest in publishing this article has been read and approved by all named authors.

REFERENCES

1. Chivukula M, Hunt J, Carter G, Kelley J, Patel M, et al. (2007) Recurrent gynandroblastoma of ovary-A case report: A molecular and immunohistochemical analysis. Int J Gynecol Pathol. 26(1):30-33.
2. Young RH, Scully RE (1985) Ovarian Sertoli-Leydig cell tumors. A clinicopathological analysis of 207 cases. Am J Surg Pathol. 9(8):543-569.
3. Jang NR, Lee DH, Jang EJ, Bae YK, Baek J, et al. (2018) Ovarian Gynandroblastoma with a Juvenile Granulosa Cell Tumor Component in a Postmenopausal Woman: A Case Report and Literature Review. J Pathol Transl Med. 52(5):344-348.
4. Gressel GM, Buza N, Pal L (2017) Ovarian Sertoli-Leydig cell tumors: A single institution experience and review of the literature. Eur J Gynaecol Oncol. 38(2):214-220.
5. Nef J, Huber DE (2021) Ovarian Sertoli-Leydig cell tumours: A systematic review of relapsed cases. Eur J Obstet Gynecol Reprod Biol. 263:261-274.
6. Guérard MJ, Ferenczy A, Arguelles MA (1982) Ovarian Sertoli-Leydig cell tumor with rhabdomyosarcoma: An ultrastructural study. Ultrastruct Pathol. 3(4):347-358.
7. Takeda A, Watanabe K, Hayashi S, Imoto S, Nakamura H (2017) Gynandroblastoma with a Juvenile Granulosa Cell Component in an Adolescent: Case Report and Literature Review. J Pediatr Adolesc Gynecol. 30(2):251-255.