A Primitive Neuroectodermal Tumor of the Ovary

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A peripheral primitive neuroectodermal tumor (PNET) is a small round tumor belonging to the PNET/Ewing's sarcoma family classified based on location in the body. Few cases of PNET arising in the ovary had been reported.

We present a rare case of PNET of the ovary occurring in a thirty-eight-year-old pregnant woman, who presented with lower abdominal pain. The MRI revealed a left ovarian tumor suspicious to be cystadenoma. Left salpingo-oophorectomy was performed as frozen section was inconclusive. The small bowel was resected and re-anastomosed during laparotomy as the mass was attached to it. The positron emission tomography revealed that the PNET had metastasized to the peritoneum and the lymph nodes. Vincristine, doxorubicin, cyclophosphamide, and etoposide/ifosfamide 4 cycles were prescribed. There was local tumor recurrence around umbilicus and biopsy was advised. The patient left for Egypt to carry out treatment in home country.

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A primitive neuroectodermal tumor (PNET) belongs to Ewings' sarcoma group. It is classified according to the location in the body which can be peripheral or central. In the literature, few cases of PNET involving the ovary were reported[1,2,3,4]. The PNET of ovary consists of neuroectodermal tissues. These tumors arising in the ovary are similar to the tumors in children arising in brain[5]. Histologically they consist of neuroblasts which are small undifferentiated cells. They are highly aggressive tumors and rapidly lead to metastasis and death.

The aim of this report is to present an extremely rare case of PNET of the ovary occurring in a pregnant woman.

THE CASE

A thirty-eight-year-old female G4P3+1 with a past history of ectopic pregnancy removed laparoscopically and one previous C-section was admitted with pain for three weeks which was not relieved by analgesia. She had a regular menstrual cycle and had Intrauterine Contraceptive Device (IUCD) removed a year ago. Her uncle died of colonic cancer.

Her pain score was 8/10. There was a history of reduced appetite. She was vitally stable; on abdominal palpation, there was some tenderness in the lower abdomen. The scan showed pelvic-abdominal mass with mixed echogenicity, solid and cystic cystadenoma/cystadenocarcinoma. The uterus was average sized; measuring 9x5 cm and no obvious myometrial focal lesion was seen. The endometrial thickness was 0.8mm, see figure 1 (A and B).

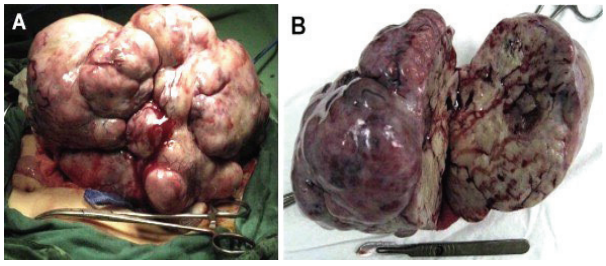

Figure 1 (A and B): PNET Tumor of the Ovary

The Hb was 11.2gms/dl. The CA 125 was 49. LDH was 248, CEA and bHCG were normal. MRI result was similar to ultrasound.

Intraoperatively, the ascitic fluid was bloodstained and a sample was sent for cytology. The left ovary was enlarged, looked cystic, soft and gelatinous occupying the whole left side of the pelvis, part of the right side of the pelvis and the Pouch of Douglas (POD). On the right side, it was found to be attached to the small intestine with some serosal and mesenteric involvement. The left adnexa were sent for frozen section and biopsies from serosa, omentum, undersurface of diaphragm and liver were taken. Bowel resection and anastomosis were performed, removing the diseased part and the lymph nodes were explored.

The frozen section diagnosis was inconclusive; therefore, a left salpingo-oophorectomy was performed. The recovery was uneventful and LDH dropped to 248. The patient was discharged on the 5th day.

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The histopathology report was peripheral neuroectodermal tumor (PNET) and the case was discussed at the Tumor Board meeting. The plan of management was chemotherapy after verifying the sensitivity of the tumor to chemotherapy. The patient wanted hysterectomy, however, the oncologist explained that it could be done later and the prognosis would not be affected.

A PET scan revealed lymph node involvement. She had her 4th course of chemotherapy and an MRI in September 2015 proved to be was normal. The patient noticed a mass close to the umbilicus one month later. A biopsy was advised by the oncologist. The patient planned to return to Egypt.

DISCUSSION

PNET of the ovary is a rare monophasic tumor. They are poorly differentiated tumors divided for histopathological purposes[6,7,8]. Immunohistochemical, karyotypic and reverse transcription-polymerase chain reaction analysis could diagnose the tumor. Diagnosing PNET of the ovary is straight forward when the tumor shows variable reactivity with antibodies to CD99, neuron-specific enolase and vimentin.

Most cells are negative for neurofilaments and synaptophysin or GFAP and S-100; however, scattered cells showing neural or glial differentiation will be positive. No cells react with antibodies for cytokeratin, desmin, chromogranin or inhibin. PNET of the ovary is highly cellular and composed of small cells with hyperchromatic nuclei and scanty cytoplasm. These cells form lobules, separated by fibromuscular septa. Areas of necrosis could be seen.

PNET of the ovary are rare and aggressive tumors associated with high morbidity and mortality. Many studies suggested that they are highly aggressive tumors and rapidly lead to metastasis and death.

The survival rate is 10.8 months to 3 years. The prognosis for PNET/Ewing's sarcoma family is improved as most of the patients with localized tumors are now cured with surgery, multiagent chemotherapy, radiotherapy or both. Two women got pregnant after treatment of PNET tumor[9]. A case of PNET arising from mature cystic teratoma in pregnancy has been reported in literature[9,10]. The prognosis is dependent on the sensitivity of the tumor to chemotherapeutic agents.

CONCLUSION

Primitive neuroectodermal tumor of the ovary is a rare entity with aggressive behavior. It must be considered as a differential diagnosis of ovarian or pelvic masses especially in young women.

Peripheral PNET found in adults is an uncommon malignancy. Further studies are required to evaluate the prognosis with multi-agent chemotherapy as optimal therapy for all patients with metastatic ovarian PNET.

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