

Primary neuroblastoma of ovary in an adult: A case report and review of literature

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ABSTRACT

Primary neuroblastoma of ovary is an extremely rare case. We present a case of a 26-year-old female with pain abdomen and right ovarian mass for which surgery was done. Histopathology revealed primary neuroblastoma without other elements of teratoma/ neuroectodermal tissue. She later received adjuvant chemotherapy and is in follow-up for 13 months and is currently disease-free.

Key words: Neuroblastoma, neuroblastoma of adult, neuroblastoma of ovary

INTRODUCTION

Neuroblastoma is derived from neural crest cells and is the most common solid tumor in infancy, fourth most common in childhood and 90% cases occur in less than 10 years of age.^[1] It is extremely rare in adults and the usual sites of occurrence are retroperitoneum, adrenal gland, pelvis, and mediastinum.

Presentation

A 26-year-old nulliparous female presented with lower abdominal pain for 2 months with no other associated complaints. On examination, there was a mobile mass arising from pelvis. A CECT showed a well defined, smooth margined heterogenous soft tissue mass of size 6.3 × 9.7 cm close to uterus, with a small speck of calcification, with right ovary not visualized and normal left ovary. A serum analysis performed revealed normal beta HCG and AFP with mild elevation of serum LDH 221 m/l.

Patient underwent staging laparotomy and there was enlarged right ovary of with intact capsule, minimal pelvic

free fluid with 1 × 1 cm paraaortic lymphnodes, pelvic lymph nodes, right obturator 2 × 1 cm, external iliac 1 × 1 cm, and rest of the abdominal structures were normal [Figure 1]. She underwent right salpingo-oophorectomy, omentectomy, Paraaortic lymphnode dissection, right pelvic lymph node dissection, and peritoneal cytology. Grossly the tumor was 9 × 10 × 5.5 cm with smooth external surface and intact capsule. Cross section showed solid lobulated gray-white lesion occupying the entire ovary. Microscopically, the specimen showed ovarian tissue with a neoplasm composed of small round cells with fine nuclear chromatin and scanty

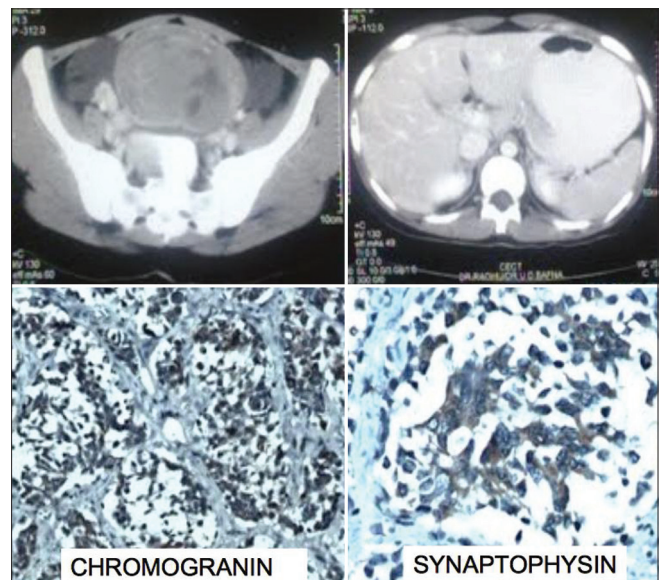


Figure 1: CT Scan: Ovarian tumor, adrenals IHC staining: chromogranin, synaptophysin

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10.4103/2278-0513.99577

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cytoplasm arranged in groups separated by thin stromal septa. Immunohistochemistry revealed the neoplastic cells positive for immunostatins, synaptophysin, chromogranin [Figure 1] and negative for cytokeratin, EMA, Inhibin and Mic 2. Peritoneal cytology, pelvic lymph nodes, paraaortic lymphnodes, and omentum did not show disease.

Later a CECT of thorax was done which showed no evidence of disease. She was started on adjuvant chemotherapy IE/ VACA and patient received 17 cycles of chemotherapy. She is on follow-up for 13 months and doing well with no evidence of recurrence.

DISCUSSION

The incidence of neuroblastoma in adults is 0.2 cases per million person years.^[2] Adrenal medulla is the most common site of neuroblastoma in infants and children. The cell of origin is neural crest cells and it can origin from any site in the sympathetic nervous system. The most common site in adults being retroperitoneum, followed by adrenal medulla, pelvis and posterior mediastinum. In patients with immature teratoma, the tumor may be associated with other components such as hair, cartilage, etc., and in patients with PNET they may be associated with other neuroectodermal elements.^[3,4]

Microscopic examination of neuroblastoma shows small dark round cells arranged in sheets and may show Homer- Wright rosettes. Neuroblastoma can be differentiated for PNET/Ewings sarcoma as they are negative for CD99, metastatic small cell carcinoma as they

are negative for CKs, EMA and from lymphoma as they are negative for LCA.

The treatment of NB in adults is surgery, chemotherapy and radiotherapy. Biological behavior of adult neuroblastomas are different and they are aggressive tumors and require aggressive management.^[5,6] Clinical data, management protocols in adults patients with neuroblastoma are not defined and need collection and evaluation of data to define guidelines for this group of patients.

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Cite this article as: Shobha K, Bafna UD, Devi UK, Amirtham U. Primary neuroblastoma of ovary in an adult: A case report and review of literature. *Clin Cancer Investig J* 2012;1:92-3.

Source of Support: Nil, **Conflict of Interest:** No.