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Primitive Neuroectodermal Tumor of the Ovary: A Case Report

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ABSTRACT

Due to its rarity, the standard clinical presentation, treatment protocol, and prognosis of primitive neuroectodermal tumor (PNET) have not been clearly described yet. Hence, herein we reported a case of ovarian peripheral PNET whose histomorphology reports caused her certain diagnoses at Imam Khomeini Hospital, affiliated to Tehran University of Medical Sciences, Tehran, Iran, in 2020.

Considering different clinical presentations, and poor prognosis of PNET compared to other ovarian malignancies, to on-time diagnosis and treatment, the patient's histomorphology and immunohistochemical (IHC) profile assessment, particularly in younger women, seems beneficial.

Keywords: Chemotherapy, Ovary mass, Primary neuroectodermal tumor, Prognosis

Introduction

A primitive neuroectodermal tumor (PNET), which can be detected in different sites in the female genital system, is scary and malignant. This tumor's most prevalent involvement site is the ovary, and the uterine corpus, respectively **(1, 2)**.

This tumor was classified into two subgroups (peripheral and central PENT) according to clinical characteristics, immunohistochemical (IHC) profiles, and genetics **(3)**. However, it has the same pathology features in different cases, its clinical manifestation broadly varied **(4)**.

In addition, the standard clinical presentation, treatment protocol, and prognosis of PENT, because of its rarity, have not been clearly described yet **(5)**. Hence, herein we reported a case of ovarian peripheral PNET that her histomorphology reports caused to her certain diagnoses at Imam Khomeini Hospital, affiliated to Tehran University of Medical Sciences, in 2020.

Case Report

A 16-year-old virgin girl with three months experience of amenorrhea followed by spotting presented at the Vali-e-Asr Hospital hospital. She had a history of surgery due to an inguinal and umbilical hernia when she was eight. Her lab tests showed an elevated serum CA125 concentration up to 432.7 U/mL and CA19-9 up to 82 U/mL. AFP, βhCG, and CA15-3 had normal concentrations.

Magnetic resonance imaging (MRI) showed bicornate uterine features and a septated vagina. A solid and complex cyst measured 42*58 mm was reported in the left ovary. The right ovary was normal, with a little amount of fluid in the pelvic cavity.

During the laparotomy, the ovaries were an elongated and striped feature, and the uterine was septated. An exophytic mass measuring 8 cm in the left ovary

and a dermoid mass measuring 3 cm in the right ovary were found. Finally, bilateral cystectomy (due to large ovarian masses) and liver biopsy were conducted.

Cytology revealed atypical cells. Microscopic evaluation showed neoplastic tissue with small cells and a high proportion of nuclear-cytoplasmic with mild pleomorphism in nuclei and several mitotic activities mainly ordered in sheets with the occasional organoid design. Furthermore, MIC2 (CD99), NSE, Synaptophysin, and Vimentin were positive in IHC analysis (Figure 1).

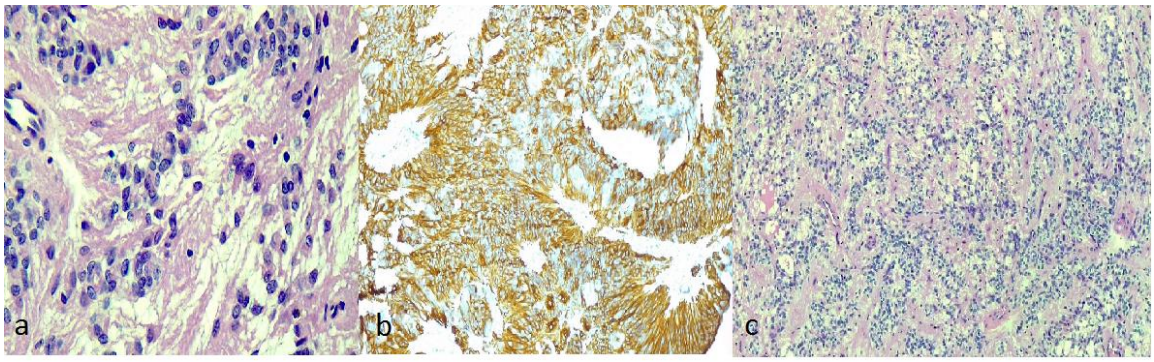

Figure 1. Immunohistochemical staining results in the study case

According to the above histopathological findings, the diagnosis of peripheral PENT was made. Immediately, six cycles of chemotherapy with Vincristine, Doxorbosin, and Cyclophosphamide were initiated. Six months after the disease detection, the patient was alive without evidence of recurrence or metastasis.

Discussion

PNETs are an uncommon and poor prognosis tumor, typically affecting young people, children, and adolescents with a median age of 25 years. However, seldom studies indicated PNET in postmenopausal women, too **(6,9)**.

The early diagnosis of pPNET is so hard because of its non-specific presentation, sign, and para-clinic data. As mentioned, there are no clear and specific para-clinic data such as imaging findings and tumor markers for PNET; however, in some patients (similar to our study case), serum CA-125 elevated was reported **(10)**.

The key point in pPNET diagnosis is pathology reports and IHC staining analysis for specific tumor markers, including CD99, vimentin, synaptophysin **(11)**. Furthermore, reverse transcription-polymerase chain reaction (RT-PCR) or fluorescence in situ hybridization (FISH) can be helpful in unusual tumor sites or patients with unspecific pathology and IHC findings **(12)**.

Considering limited reported cases with ovarian PNET, the most prevalent clinical manifestation were abdominal pain, abdominal distension, and pelvic masses. Microscopic, IHC, and FISH data were the gold diagnostic method, and no confirmed therapy exists yet, and most treatment usages are empirical.

Conclusion

Considering different clinical presentation, and poor prognosis of PNET compared to other ovarian malignancies, to on-time diagnosis and treatment, the patient's histomorphology and immunohistochemical (IHC) profile assessment, particularly in younger women seems beneficial.

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Conflict of Interest

Authors declared no conflict of interests.

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