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Ovarian Primitive Neuroectodermal Tumor (PNET) – a case report

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SUMMARY

The Primitive neuroectodermal tumor (PNET) is a malignant tumor which arises from neural crest cells. It is a rare tumor, usually occurring in children and young adults. PNET Categorized in the same tumor family as Ewing sarcoma which occur in bones and soft tissues. A small number of PNET cases arising in the pelvic organs like as uterine corpus, ovaries, cervix, and vulva in the broad ligament have been reported. We present a case of pelvic PNET in the left ovary with adhesion to the surrounding organs associated with para aortic lymphadenopathy. The site and volume of tumor, and presence or absence of metastasis are prognostic factors of PNET. PNET is a malignant tumor which has lymphatic and haematogenous metastases. Although the local tumor irradiation could be a radical treatment even in some cases but some of them requires surgical resection and chemotherapy.

Key words: Primitive Neuroectodermal Tumors; Ewing's Sarcoma; Pelvic Neoplasm; Ovary Cancer

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INTRODUCTION

The Primitive neuroectodermal tumor (PNET) is a malignant tumor which arises from neural crest cells. It is a rare tumor, usually occurring in children and young adults. The overall 5 year survival rate is about 53% [1]. The majority of the cells in PNET are derived from neuroectoderm, but have not differentiated in a normal course. The (PNET) was first used by Hart and Earle in 1973 to introduce a kind of tumors with fetal Neuroectodermal cells origin [2]. According to the tissue of origin the PNET family of tumors divided into the 3 groups: CNS primitive neuroectodermal tumors (PNETs) – Tumors derived from the central nervous system [3]. Neuroblastoma and Tumors derived from the autonomic nervous system. Peripheral primitive neuroectodermal tumors (pPNETs) - Tumors derived from tissues outside the nervous system. Tumors which often involve the sympathetic nervous system or bones and soft tissues are described as the peripheral PNET; this group arises from the neural crest and primitive neuroendocrine cells [4]. Pelvic PNETs are often detected in the uterine corpus, ovaries, cervix, and vulva and even in the broad ligament [5]. The diagnosis of the PNET can be made based on histological examinations. Abdomen and Pelvic ultrasonography computed Tomography (CT) scan with IV contrast and in some cases Magnetic Resonance Imaging (MRI) are essential in the diagnosis of tumor size, position, surrounding involvement and ruling out of lymphadenopathy and metastasis. A review of imaging findings in PNET suggests that no characteristic finding aids in the preoperative diagnosis of this tumor. The PNET often occur in bones and soft tissues around the bones PNET also can arises in the pelvis especially in the ovary and even in the broad ligament. The PNET is aggressive and like to involve other organs and has poor prognosis than other small round cell tumors.

PNET treatment protocol includes local control that followed by chemotherapy and in some cases radiotherapy [6].

CASE PRESENTATION

The patient was a 30-year-old woman with a history of pelvic pain and distention for less than two weeks. The patient provided written informed consent, and her anonymity was preserved. After physical examinations the patient referred to Pelvic ultrasonography and ovarian torsion in the left ovary was diagnosed. For better assessment she refers to imaging center of haft Tir Hospital. Abdominopelvic CT scan with IV contrast revealed an irregular heterogeneous mass in the left adnex with adhesion to the surrounding organs associated with para aortic lymphadenopathy (Fig.1.). She is referred to surgeon and underwent laparotomy and pelvic surgery for resection of tumor. In laparotomy a mass (greatest diameter 10 cm) and, Para aortic lymphadenopathy were observed. The tumor infiltrated most of the left broad ligament and ovary. Complete pelvic and Para aortic lymphadenectomy was impossible. S O the tumor and one of lymph node resected for frozen-section analysis which reportedan undifferentiated malignant tumor with 70 % proliferative index. Histopathologic examination showed malignant neoplastic tissue composed sheet and nests of medium to large pleomorphic tumoral cells with vesicular nuclei prominent nucleoli and scant eosinophilic cytoplasm diffuse area of necrosis and brisk mitotic figures are identified .probably ovarian dysgerminoma of the pelvis and the tumor sample pathology lab in early assessment the pathologic revealed ovarian dysgerminoma (Fig.2.). Complete pelvic and Para aortic lymphadenectomy was impossible. For definite diagnosis, immunohistochemistry (IHC) was done and revealed strong diffuse reaction for CD99 (Fig.3.), weakly positive for CD10, and negative reaction for CD117, CD10, CD20, CD34, CK, EMA, Tdt and myiogenin and LCA. Which supports the

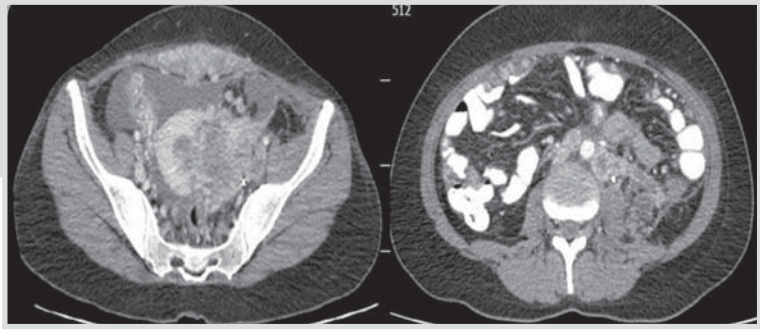

Fig.1. Abdominal CT scan with contrast that shows an irregular heterogeneous mass in the left adnex with adhesion to the surrounding organs associated with para aortic lymphadenopathy

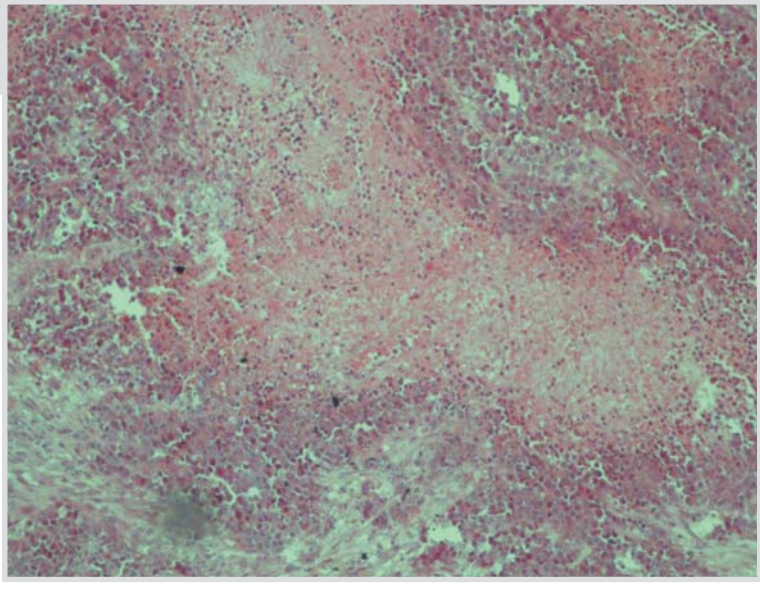

Fig. 2. Histopathologic examination showed a high grade tumor with extensive necrosis

diagnosis of PNET? Because of lymphadenopathy the patient was subsequently treated with chemotherapy BEP regimen (Bleomycin, Cisplatinum, and Etoposide). But she died due two mounts later to liver, adrenal and lung metastasis and pulmonary emboli before completing all the courses of chemotherapy.

DISCUSSION

The PNET is a malignant tumor which arises from neural crest cells. It is a rare tumor (about 1% of all soft tissue sarcomas), usually occurring in children and young [7]. Although PNET often occur in bone and soft tissue but rare cases have been reported in pelvis especially in ovary. Almost all of pelvic PNET had no specific sign and symptom. Some of PNET cases had the reciprocal translocation in cytogenetic examination. The most common sites of PNET metastases are the lung, bone, and bone marrow. In our case metastasis occurred in adrenal and lung. The main treatment for primitive neuroectodermal tumors (PNETs) is surgery to remove the whole tumor or as much of it as possible. Surgical approach of the PNET should be complete resection of the tumor and lymph node in near side of tumor. Although in some cases complete resection of PNET is not possible due to the aggressive behavior and dissemination of the neoplasm, in our patient, complete Para aortic lymphadenectomy was impossible. In PNET Chemotherapy regimens have significantly improved outcomes and survival. A postoperative chemotherapy program as an adjuvant therapy is recommended immediately following diagnosis. In the current treatment protocols, chemotherapy regimens include combination of Vincristine, Actinomycin D, Cyclophosphamide, and Doxorubicin. Moreover, a combination of Etoposide and Iosfamide appears to have the greatest survival advantage. Also combination of Taxol/carboplatin chemotherapy is reported [5]. Jones et al and Jürgen's et al reported somewhat better survival rates in patients with localized disease, with 2 large trials showing 2- and 3-years survival rates of 65% and 56%, respectively [8]. Kushner et al reported a similarly dismal 25% progression-free survival rate at 2 years in patients with only localized disease [9]. Radiotherapy for Local tumor control (LTC) by using a total dose of 60Gy in 30 fractions also reported to be effective [10]. The PNET must be considered in the differential diagnosis of ovary tumor in the pelvis in young females.

The PNET is an aggressive, malignant, immature embryonic tumor that tends to metastasize and should be considered in the differential diagnosis of pelvic masses.

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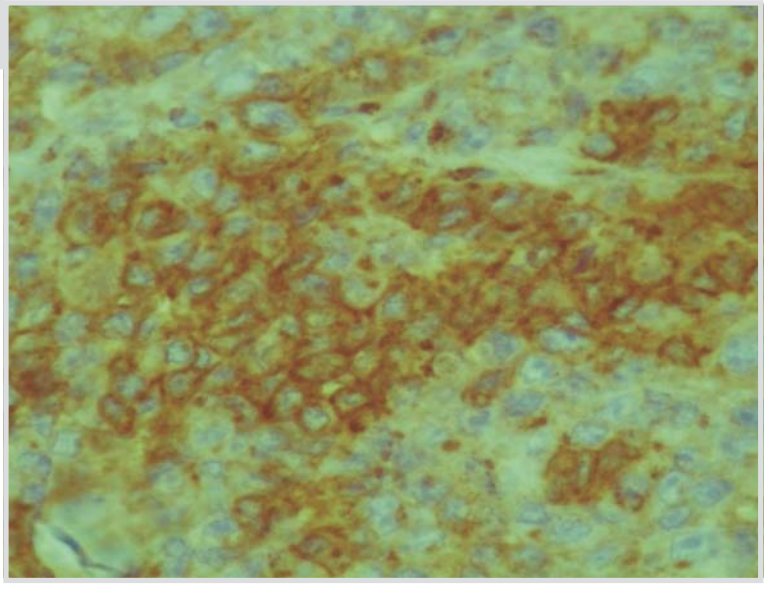

Fig. 3. Strong immunoreactivity for CD99 in tumoral cells

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