Case Report

Primitive Neuroectodermal Tumor of the Kidney

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ABSTRACT

Primitive neuroectodermal tumor (PNET) is an uncommon malignancy of bone and soft tissue witch rarely occurs in the kidney. In more than 90% of the cases, the tumor cells relieves a balanced translocation (11; 22) (q24; q12). Immunohistochemical staining may be required for diagnosis of PNET. The cells of tumor express CD99, vimentin, NSE, FL1 but do not express Ck, LCA, myogenin, and WT1. We present a 36-year –old female with left –side tender abdominal swelling, and history of trauma to abdominal. CT imaging confirmed a huge solid mass of kidney, also extending into renal pelvis. Histological section of the lesion showed a malignant proliferation of small round cells in rosette-like pattern with foci of necrosis area. Tumor cells expressed high level of CD 99 antigen. The diagnosis of the lesion was primitive neuroectodermal tumors (PNET). Following-up after 6 months showed no recurrence.

Key words: **Primitive Neuroectodermal Tumor, Kidney, Iran**

Introduction

The most common renal tumor is renal cell carcinoma which has an accuracy of more than 85% (1). Primitive neuroectodermal tumor of the kidney is a rare, aggressive neoplasm although the incidence is increasing. Primitive neuroectodermal tumor (PNET) of the kidney was first reported by Mor in 1994 (2). This tumor is a small round cell tumor that has neuroepithelial differentiation, which typically occurs in young children and adolescents. PNET is derived from neuroectodermal cells and can occur in bone and soft tissue, visceral organ such as urogential,

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intra-abdominal and intratheracis organ (3).
We report here a PNET in the kidney and discuss relevant issues about the clinicopathological finding, origin, diagnosis and treatment of the present tumor.

Case Report

A 36 year old female was referred to Ghaem Hospital Mashhad Iran. She had 3- year history of trauma in the left abdominal region. Urine analysis was normal. The Axial CT image showed an extensively mass with foci of necrotic area in the left side of kidney which destroyed upper middle part of kidney and evading upper middle part of renal pelvis left renal vein were intact (Fig.1).

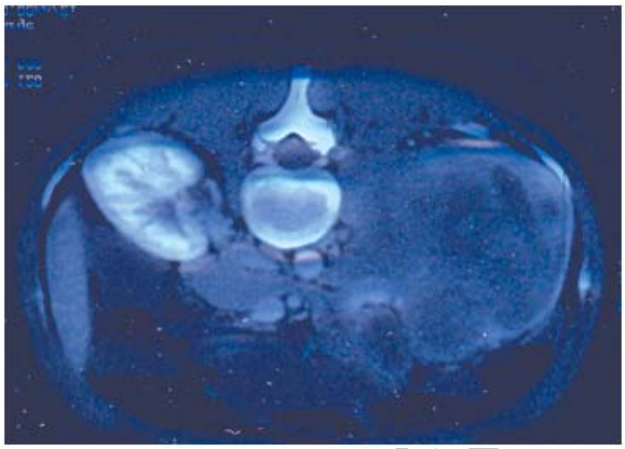

Fig.1- Axial abdomen revealed a mass with foci of necrosis area in the left kidney.

The physical examination which revealed evidence of a tangible abdominal mass and the laboratory results were normal. Under general anesthesia the patient subsequently underwent left radical nephrectomy. There was no need for blood transfusion. The sample was sent for evaluation.The gross specimen was a huge mass composed of a white-gray mass that was firm and the dimension was 44 ×31×18cm (Fig. 2). Microscopically features of the tissue revealed malignant neoplastis proliferation of small round cells which grew in sheet-like pattern. In some areas, tumor cells arranged in rosette-like structure, with scanty eosinophilic cytoplasm and round to ovoid open nuclei with dusty chromatin pattern, also geographic necrosis and mitotic figure of cells was presented (Fig.3). Through IHC, the cells of tumor expressed CD99, vimentin, NSE, FL1 but did not express Ck, LCA, myogenin, and WT1 (Fig.4).The diagnosis of primitive neuroectodermal tumors (PNET) was made. The patient referred for adjutant chemotherapy. Six-month following up after lesion excision showed no recurrence.

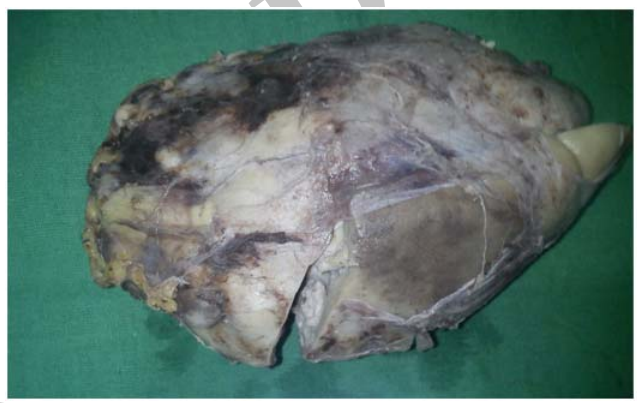

Fig.2- The gross specimen was a huge mass consisting a white-gray brown mass that was firm of dimension 44 × 31 × 18cm.

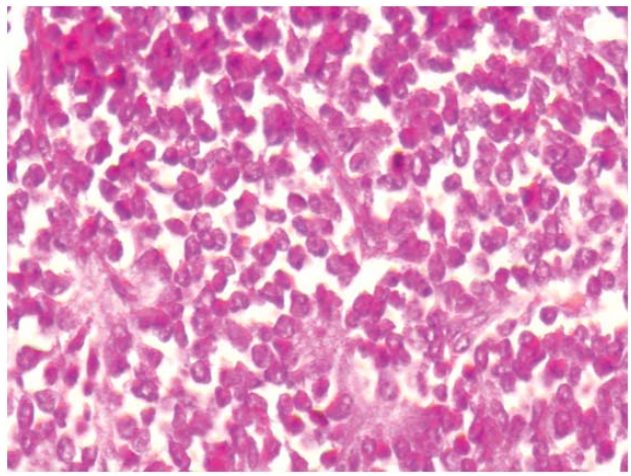

Fig.3- Malignant neoplastic proliferation of kidney small round cells in a sheet like pattern with scanty cytoplasm and round to oval nuclei with sharp borders also abnormal mitotic figure is present. (H&E, original magnification × 100).

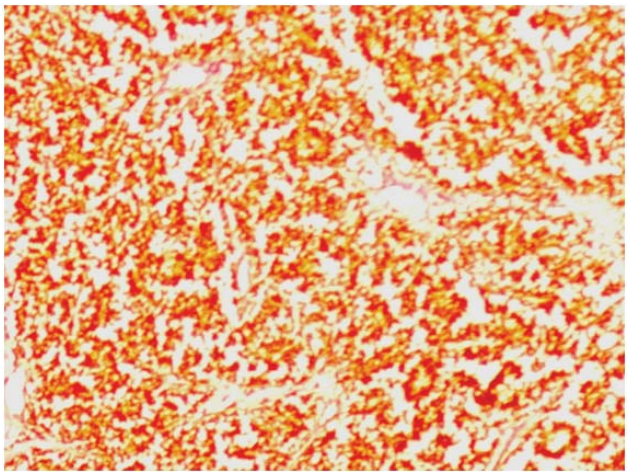

Fig.4 - With the use of immunohistochemistry, all cells of tumor expressed CD99 antigen (MIC-2 gen product).

Discussion

PNET is a round cell sarcoma first described by Stout in 1918 (4). Microscopically, the neoplasm is characterized by proliferation of small round – shaped mesenchymal cells and Homer-Wright rosettes, which occur commonly in bone and soft tissue. PNET may occur in any age, but most commonly involve young adult with a mean age of 28 years similar as the presented case. The most common symptoms of this tumor are pain and swelling as like as our case (1). More *et al*. for the first time described the first renal PNET. According to previous case repot renal PNET is more common in young male, but a few renal PNET has been reported in young female as the presented case.

Histologically, most of the cited cases describe the same histological pattern, consist of a sheet-like proliferation of small round cells with small amounts of clear to eosinophilic cytoplasm which formed in rosette-like structures in some areas as well as tumor cells are usually uniform and small with finely dispersed chromatin and small nucleoli (5). Geographic necrosis and mitotic figures are frequently present.

Variable histologic patterns designated as large cell (atypical), adamantinoma-like, scelorosing, and extensively spindle tumors (6-8). So PNET may be confused with other round cell tumor such as lymphoblastic lymphoma, poorly differentiated synovial sarcoma, small cell osteosarcoma, mesenchymal chondrosarcoma, small cell carcinoma and small variants of melanoma in both bone and soft tissue location (1). It is not easy to recognize high-grade tumor without IHC-staining (1), so we used CD-99, Fll-1 and Wt-1 to diagnose of PNET. Our case had a tumor composed of small round tumor which stained positive for CD 99.

In genetic findings less than 95% of PNET shows karyotyp translocations [11; 22] (q24; q12) and nearly 5% of them indicate t [21; 22] (q22; q12) (9). Renal PNET is a rare tumor that until 2005 have reported a huge series of renal PNET including 91 cases considered to be PNET according to the histopathology and immunohistochemistry (2) .

Definitive diagnosis can be obtained by fine needle aspiration cytology (FNAC), FISH or R T-PCR techniques. Kumar *et al*., introduced RT-CPR on FNAC method for diagnosis of the renal PNET (10). The high malignant nature of this tumor is reflected in its tendency for metastasis, especially to lungs, bone, and lymph nodes.

A primitive neuroectodermal tumor of the kidney is an aggressive tumor with a poor prognosis. A 5-year disease free survival is 45-55% (11-15). Jiminez *et al*. discussed 11 patients with follow up information, that one of them was alive for 64 months; it seems the longest survival of the patients (16). In a study, from 14 patients with diagnosis of kidney PNET, 4 died of metastasis 1-35 months after diagnosis and 10 survived (17). Therefore PNET of the kidney may be managed with surgery dissection plus postoperative radiotherapy and chemotherapy. Finally, in this case, we reported a case of renal PNET, which is

extremely rare.

The combination of morphological finding, immunohistochemical and genetic changes analyzed together form the base of diagnosing of the PNET. Patients should be evaluated for recurrent of tumor in long term.

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The authors declare that there is no conflict of interest.

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