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Case Report

Primitive Neuroectodermal Tumor of the Kidney with Inferior Vena Caval Thrombus

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Abstract ***Background:*** **Primitive neuroectodermal tumor (PNET) of the kidney is extremely rare.**

Objective: **The objective is to report a case of neuroectodermal tumor of kidney, result of the treatment and review of the literatures.**

Case report: **We reported a case of young female presented with abdominal mass which computed tomographic scan showed a large inhomogeneous enhancing solid tumor of right kidney with retrohepatic vena caval thrombus. Radical nephrectomy with vena caval thrombectomy was performed. Histopathology demonstrated round tumor cells with rosette formation which immunohistochemistry stained positive for CD99 (MIC-2), the characteristic of this tumor. Despite undergoing adjuvant chemotherapy, the patient developed local recurrences and multiple distant metastases at only 8 months after operation.**

Conclusion: **The diagnosis of renal PNETs should be considered in young adult patients who presented with aggressive renal masses at initial presentations. Immunohistochemistry is of invaluable help for making a definite diagnosis. This particular type of renal tumor is highly aggressive in nature and has poor prognosis despite any adjuvant treatments.**

Keywords: **Kidney, Inferior vena cava, Primitive neuroectodermal, Thrombus**

Introduction

Primitive neuroectodermal tumor (PNET) is a group of small round cell tumors that may arise in central or peripheral nervous systems. Bones and soft tissues are the common extracranial locations whereas kidney is extremely rare primary location for these tumors.[1] Approximately 50 cases were reported in the literatures but only few cases developed tumor thrombus in the inferior vena cava (IVC).[2-5] Clinical features of PNET do not differ from renal cell carcinoma (RCC) which is the most common neoplasm of kidney. Herein we report a case of PNET of right kidney with IVC thrombus.

Case Report

A 36-year-old female was admitted to our department in February 2006. The patient presented with abdominal distension and a huge, firm, fixed mass on right upper quadrant of abdomen was found

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during examination. Complete laboratory values were normal except high erythrocyte sedimentation rate (60 mm. per hour). Chest and abdominal computed tomographic (CT) scan revealed a large, moderately enhanced, heterogeneous mass (12 × 15 × 15 cm) occupying anterior aspect of right kidney with multiple feeding arteries and some area of tumor necrosis (Fig. 1). No calcification was detected. Additionally, there was a large precaval mass (4×4×10 cm) extended from renal tumor down to sacral level. Tumor thrombus extended to retrohepatic portion of IVC. Echocardiography revealed no intracardiac mass and bone scan was unremarkable.

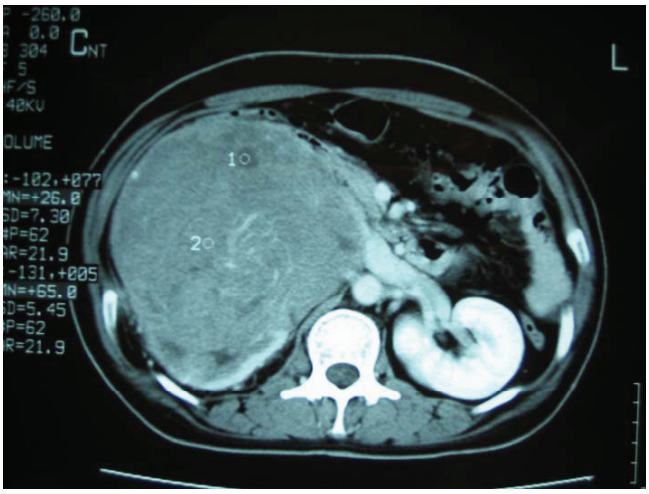

Figure 1 Computed tomographic scan shows a large heterogeneous enhancing mass of right kidney with inferior vena cava thrombus

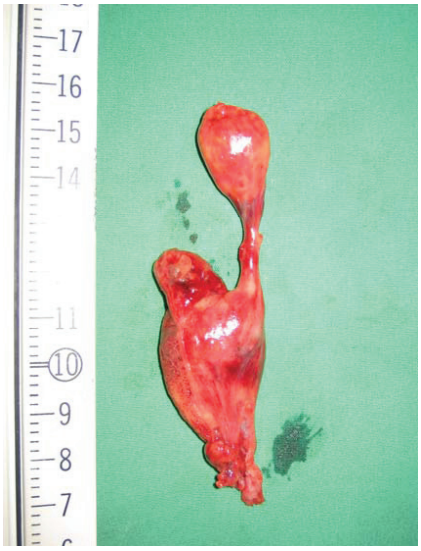

Figure 2 A gross appearance of IVC thrombus (retro-hepatic level)

The patient underwent standard radical nephrectomy and IVC tumor thrombectomy (Fig. 2). Intra-operative findings demonstrated the tumor adhered to liver and second part of duodenum. Gross specimen revealed the tumor mass invading IVC, angiolymphatic vessels, renal pelvis, renal capsule, perinephric fat and ureteral resection end.

Histopathology showed malignant small round tumor cells with rosette formation (Fig. 3). No glycogen was detected. The tumor cell positively stained with CD99 (MIC-2) (Fig. 4) but negatively with AE1/AE3 cytokeratins, vimentin, CD45, TdT, or NSE. These findings confirmed the diagnosis of PNET of the kidney.

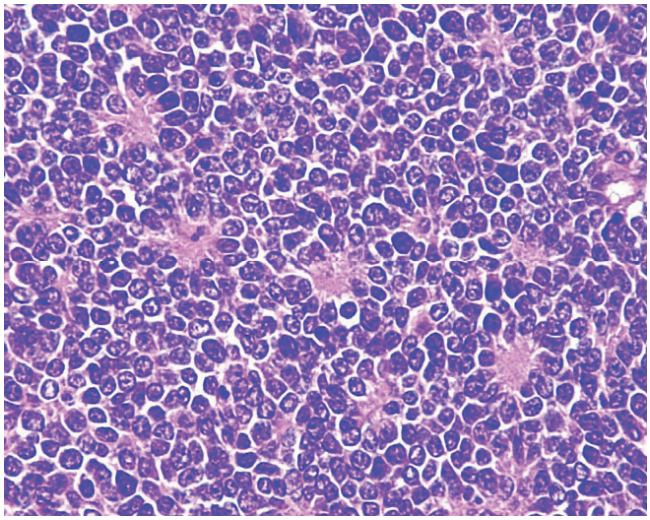

Figure 3 Rosette formation amidst the malignant round cells in PNET of the kidney (× 400).

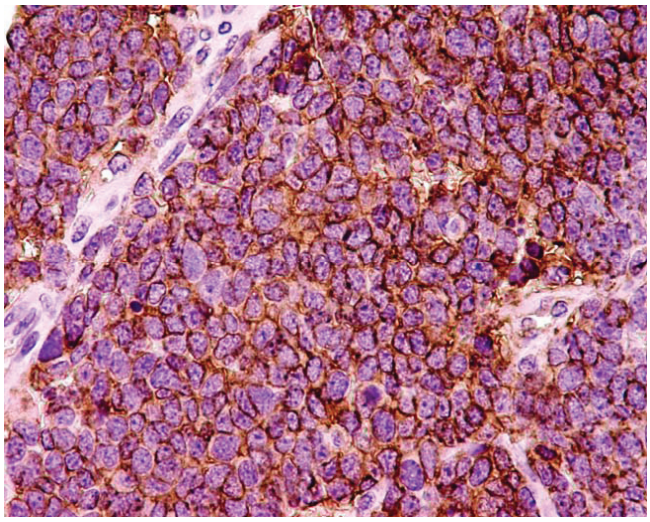

Figure 4 CD99 immunoreactivity (membrane staining) in PNET of the kidney (× 400).

The postoperative course was uneventful. The patient received four cycles of adjuvant chemotherapy (vincristine, adriamycin, cyclophosphamide). At 8 months post surgery, the patient experienced abdominal discomfort. Repeated CT scan discovered local recurrence, infrahepatic IVC thrombus, enlarged para-aortic and left supraclavicular lymph nodes and multiple liver metastases. Palliative external beam radiation was given to alleviate her intractable back pain and symptomatic left supraclavicular lymphadenopathy. The patient died 2 months later from multi-organ failure.

Discussion

PNET of the kidney is extremely rare and there are a few reported cases presenting with IVC thrombus. To the best of our knowledge, this is the first report case of primary PNET of the kidney with a level III (retro-hepatic IVC) thrombus. Only two cases of level II (infra- hepatic IVC) and two cases of level IV (supra-diaphragmatic IVC) have been reported in the literatures.[2-5] All cases were younger than 60 years old with female predominance. Most of them underwent radical nephrectomy concomitant with IVC tumor thrombectomy. Resection of pulmonary nodules was performed along with radical surgery in one case with multiple lung metastases recognized at initial presentation. Adjuvant chemotherapy was given in 3 cases. The prognosis depended on tumor staging at presentation and chemotherapy did not alter tumor progression. Multiple recurrences and metastases developed in a short period after treatment. Palliative radiotherapy was used only in our case. A unique finding in the present case was the large longitudinal ureteral tumor extension found intra-operatively. Such a behavior of tumor has not been previously described in renal PNETs and must be distinguished from transitional cell carcinoma (TCC) of the kidney with vena caval involvement, especially a rare hypervascular renal pelvis TCC.[6]

In general, all PNETs are positive for CD99 and some of them are positive for NSE, as well. Many authors use molecular studies, the characteristic translocation t(11;22) (q24;q12) demonstrating by polymerase chain reaction (PCR), to confirm the diagnosis of these tumors.[7,8] However, NSE immunostaining was negative in the present case and molecular analysis was not performed.

The diagnosis of renal PNETs should be considered in young adult patients who present with aggressive renal masses at initial presentations. Immunohistochemistry is of invaluable help for making a definite diagnosis. This particular type of renal tumor is highly aggressive in nature and has poor prognosis despite any adjuvant treatments. Radical surgery is the primary treatment, followed by combination chemotherapy and/or radiation in selected cases.

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