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PRIMITIVE NEUROECTODERMAL TUMOR (PNET) OF THE KIDNEY WITH LEVEL IV INFERIOR VENA CAVAL THROMBUS: A CASE REPORT

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Abstract

Primitive neuroectodermal tumor (PNET) is classified as a malignant tumor of the kidney. It was first recognized by Arthur Purdy Stout in 1918, as a member of the family of "small round-cell tumors". Primitive renal localization is very rare. We report a case of Primitive neuroectodermal tumor of the kidney with level IV inferior vena caval (IVC) thrombus. Histologically, the tumor consisted of small round cells with round nuclei and scant cytoplasm.

Keywords: Kidney, Neuroectodermal, PNET, Tumor, Venous thrombus

Introduction

The most comprehensive classification of Renal tumors is that offered by Deming and Harvard (1970)[1]. Barbaric (1994)[2] approached renal masses on the basis of pathologic features (malignant, benign or inflammatory) or radiographic features (simple cysts, complex cysts, fatty tumors and others). This classification appears practical and should assist in differential diagnosis of renal masses. Primitive neuroectodermal tumor (PNET) is classified as a malignant tumor of the kidney. It was first recognized by Arthur Purdy Stout in 1918, as a member of the family of "small round-cell tumors"[3]. Primitive renal localization is very rare. There are almost 50 cases reported in the literature, although it is difficult to estimate the exact number since often it has not been differentiated from Ewing's Sarcoma[4]. Renal PNET is more aggressive than in the other sites. It frequently arises during childhood or adolescence, having an aggressive clinical course towards metastatic disease and death. It often recurs locally and metastases early to regional lymph nodes, lungs, bone and bone marrow, resulting in a poor prognosis. The 5 year disease free survival rate for patients presenting with well confined extra-skeletal PNET, is around 45-55% and cases with advanced disease at presentation have a median relapse free survival of only 2 years[4].

We report a case of Primitive neuroectodermal tumor of the kidney with level IV inferior vena caval (IVC) thrombus.

Case Report

A 26 year old male presented to the hospital with symptoms of abdominal distension, bilateral pedal oedema, loss of appetite and haematuria. Physical examination revealed a left renal mass with minimal ascites and engorged veins over lower abdomen. Abdominal ultrasonography revealed a huge 23×13 cm left renal mass of mixed echotexture. Abdominal computerized tomography (CT) confirmed the mass with level IV inferior vena cava (IVC) thrombus. Echocardiography similarly revealed the thrombus in the right atrium of the heart. A clinical diagnosis of Left RCC with level IV IVC thrombus was made. Bone scan and chest x-ray revealed no evidence of metastases.

The patient underwent an abdominal exploration through a bilateral chevron incision. A well vascularised giant left renal mass was seen. The IVC was explored by making an incision along the right Line of Toldt and extending the incision along the mesentry of the small bowel. The left renal mass was dissected from its sorroundings. The left renal artery was identified, ligated and cut. The incision was extended through a median sternotomy. The patient was prepared for hypothermic complete cardiopulmonary bypass. The left renal vein, IVC were dissected so as to apply clamps. Once the patient was shifted to cardiopulmonary bypass, the IVC and the right atrium was opened. The IVC thrombus was gradually delivered through the IVC in toto. The left radical nephrectomy along with IVC thrombectomy was performed. The right atrium and IVC were closed, cardiopulmonary bypass reversed and the patient warmed.

The left renal mass was 1138 gms of weight and measured 23×13 cm in size. The tumor was multilobular, grey, glistening, focally haemorrhagic, surrounded by a capsule and with a sharp demarcation from the involved kidney. Histologically, the tumor consisted of small round cells with round nuclei and scant cytoplasm. It presented different patterns with

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cohesive lobules or rosettes and perivascular pseudo-rosettes or, in some areas, spindle cellular elements. The tumor expressed Vimentin, Mic-2 and Fli-1 which was considered diagnostic of Primitive Neuroectodermal tumor (PNET).

In view of this rare diagnosis the patient was counselled and put on multidrug chemotherapy. Patient has put on weight after surgery and the bilateral pedal oedema disappeared. Patient is on close follow-up.

Figure 1: CT Scan Images showing a large tumor arising from and replacing whole of left kidney with tumor thrombus in the inferior venacava extending into the heart.

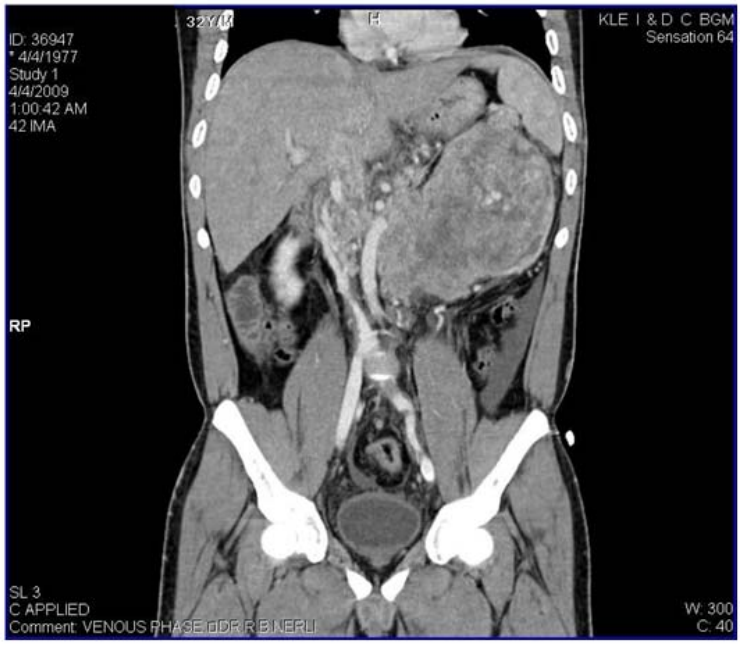


Figure 2: Round oval cells with centrally placed nuclei. Nuclear pleomorphism is not remarkable. Tumor is vascular. X40 (H& E)

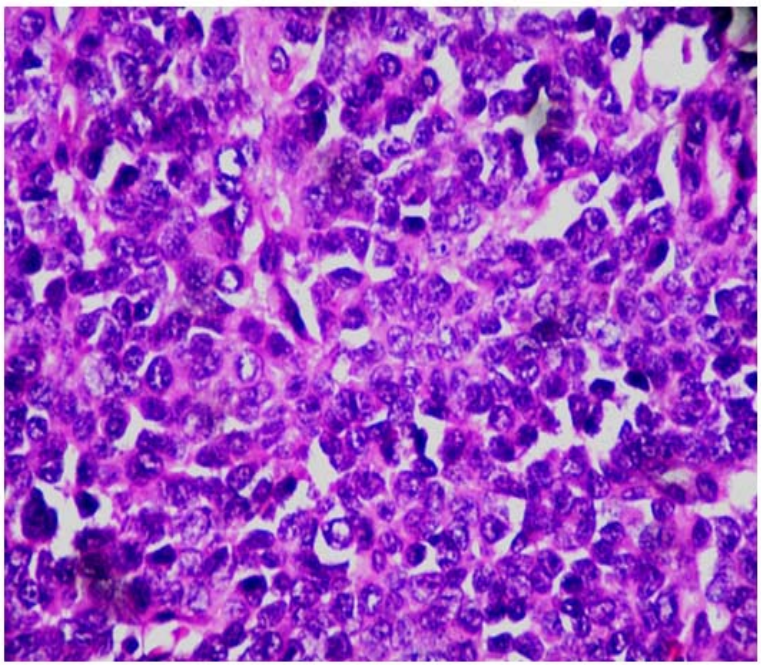
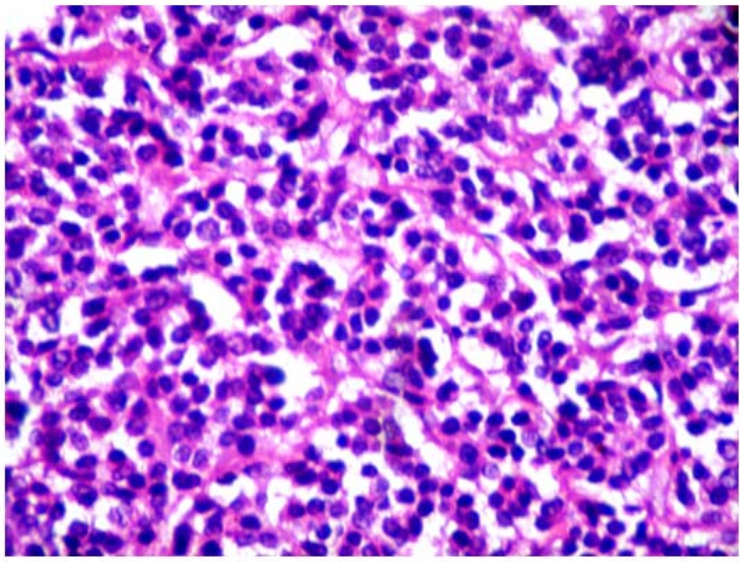


Figure 3 CD99 staining of tumor and kidney (X200)



Discussion

Peripheral PNET is a malignant small cell neoplasm of neural origin arising outside the brain, spinal cord and sympathetic nervous system. This tumor most often arises in the chest wall and paraspinal region: however, less common location described in the literature are bones, limbs and genitourinary tract[5,6]. PNET of the kidney is rarely seen and highly aggressive malignant neoplasm. The disease occurs predominantly in older children and young adults, and often with metastatic disease[6]. Histologically, the tumor is composed of small dark cells arranged in cords, nests or clusters with or without rosettes and pseudo rosettes. The most important histological criterion is the formation of rosette like structures formed by the tumor cells [5,6]. Well formed rosettes can also be identified in neuroblastoma. Therefore carefully selected immunohistochemical panel is important for differentiating this tumor from other small round cell tumors of the kidney such as rhabdomyosarcoma, neuroblastoma, desmoplastic small round cell tumor, nephroblastoma and Ewing's sarcoma[6,7]. Immuno histochemically PNET cells express Vimentin, NSE and CD99.

PNET tumors of the kidney usually affect young individuals, are large at presentation and tend to be highly aggressive tumors. Sivaramkrishna et al[8] reported a case of PNET of the kidney with renal vein thrombus. This patient underwent radical nephrectomy with ligation of renal vein flush on the IVC, clear of the thrombus. The patient underwent postoperative chemotherapy, and was disease free at nine months follow-up. Karnes et al[9] similarly reported a case of PNET of the kidney with level II IVC thrombus. The patient underwent right radical nephrectomy and level II vena caval thrombectomy. Chemotherap, consisting of multiple drugs was initiated. At more than 1 year follow-up the patient had no evidence of recurrence after completion of chemotherapy. Thomas et al [10] reported on a level IV IVC thrombus. The patient underwent right radical nephrectomy and level IV inferior vena caval thrombectomy with cardiopulmonary bypass and deep hypothermic circulatory arrest. The patient remained disease free at limited follow-up of 5 months postoperatively. Wada et al [11] reported a 23 year old female who presented with a level II thrombus and pulmonary metastases. The patient underwent radical nephrectomy with IVC thrombectomy. The pulmonary metastatic lesions decreased in size and number about 14 days after surgery, and subsequently increasingly regressed with time. At 2 months after surgery they had completely regressed. The patient remained well 1 year after surgery. The resection of the primary lesion was probably responsible for the complete regression of the pulmonary metastases.

PNET tumors of the Kidney are rare and there has been no standard treatment protocol. Most authors recommend surgery followed by multi drug chemotherapy. PNET of the kidney needs to be kept in mind as a differential diagnosis in young adults presenting with a large kidney mass.

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