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Primitive neuroectodermal tumor with inferior vena cava thrombosis in a young man: A Case Report

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Article Info

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Abstract

PNETs are neural crest malignancies that arise from the neuroectoderm and are classified as central or peripheral depending on where the tumors originate in thenervous system. Genitourinary PNETs are very rare. Compared with RCCs, renal PNETs are more commonly associated with patients younger than 20 years and with large tumors at presentation.

Keywords: Primitive neuroectodermal tumors, Immunohistochemistry, Thrombosis, sarcoma.

Introduction

PNET is related to the Ewing sarcoma family of tumors typically manifesting in the bone or soft tissues of the extremities, trunk, and head and neck and only rarely in the viscera or kidneys. These tumors are derived from primitive neural crest cells and account for less than 2 % of renal malignancies. Compared with RCCs, renal PNETs are more commonly associated with patients younger than 20 years and with large tumors at presentation. With the advancing IHC studies, we can diagnose these tumors better than the past.

Case Report

A 44-year-old man presented with a 10 month history of right flank pain and hematuria. He did not report any constitutional symptoms including fever, weight loss. He had no family history of renal disease or nephrolithiasis. On physical examination, he was afebrile.On palpation a firm mass was palpated in RUQ below the costal margin, which was non-tender with ill-defined margins. Both lower extremities had no edema. Laboratory analysis showed haemoglobin 10.1 g/dl. Urinalysis was unremarkable and urine culture was negative. Blood urea was 29 mg/dl and serum creatinine was 1.18 mg/dl .liver function tests were normal.

Renal sonography demonstrated a large hypoechoic mass in right kidney with 146*99mm and hypoechoic mass with 155*57 mm in para aortic .scrotal ultrasonography was normal. Liver and gall bladder in sonography were normal. . Computed tomography (CT) scan with intravenous contrast showed heterogeneous mass in right kidney associated with para aortic lymphadenopathy. Abdominopelvic MRI discovered a 213*145*190 mm lobulated enhancing solid mass in upper pole of right kidney.Right renal vessels and IVC were encased (Figure 1).

CT-guided biopsy demonstrated that therenal tumor definitely is not lymphoma .we decided to performright radical nephrectomy along with IVC thrombectomy.

A midline incision was given and huge mass was seen in the mid and upper pole of right kidney with multiple enlarged aortocaval lymph nodes. Diffuse desmoplastic reaction was seen around the area of tumor and in aortocaval region. There was tumor in renal vein extending in IVC until infrahepatic part. The radical nephrectomy performed and IVC removed because the thrombotic IVC was encased within tumor .Para aortic and inter aortocaval lymphadenectomy performed. (Figure 2)

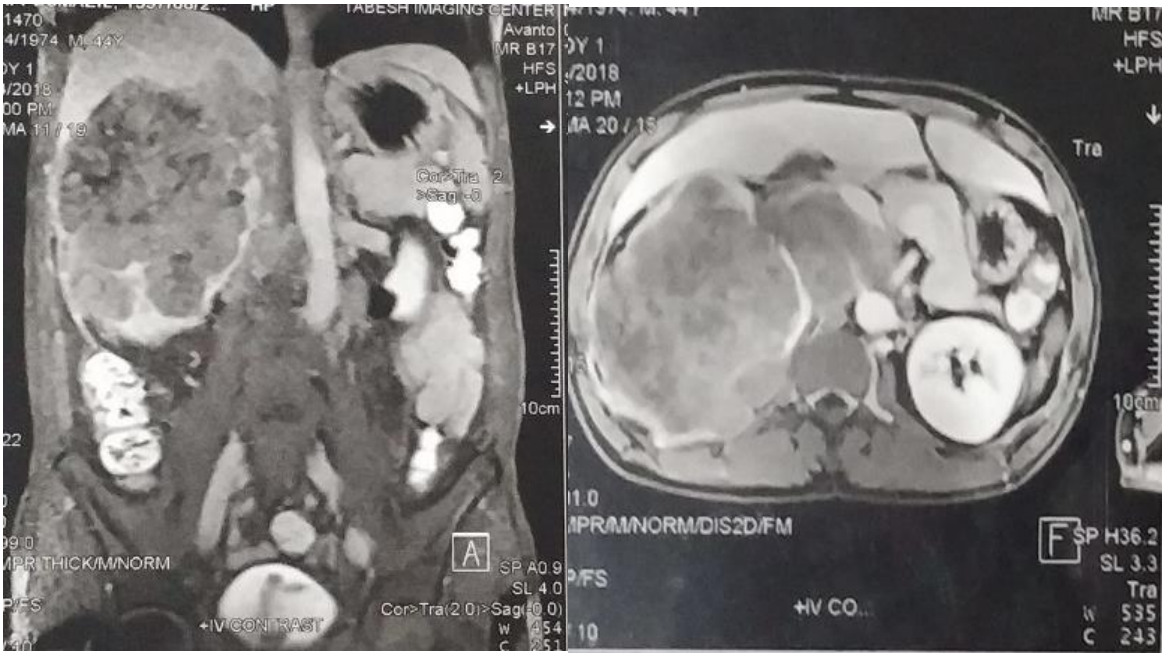


Figure 1. Abdominal MRI of the patient with IV and oral contrast showed large right kidney mass with heterogeneous enhancement and encasement of the ivc and displacement of aorta

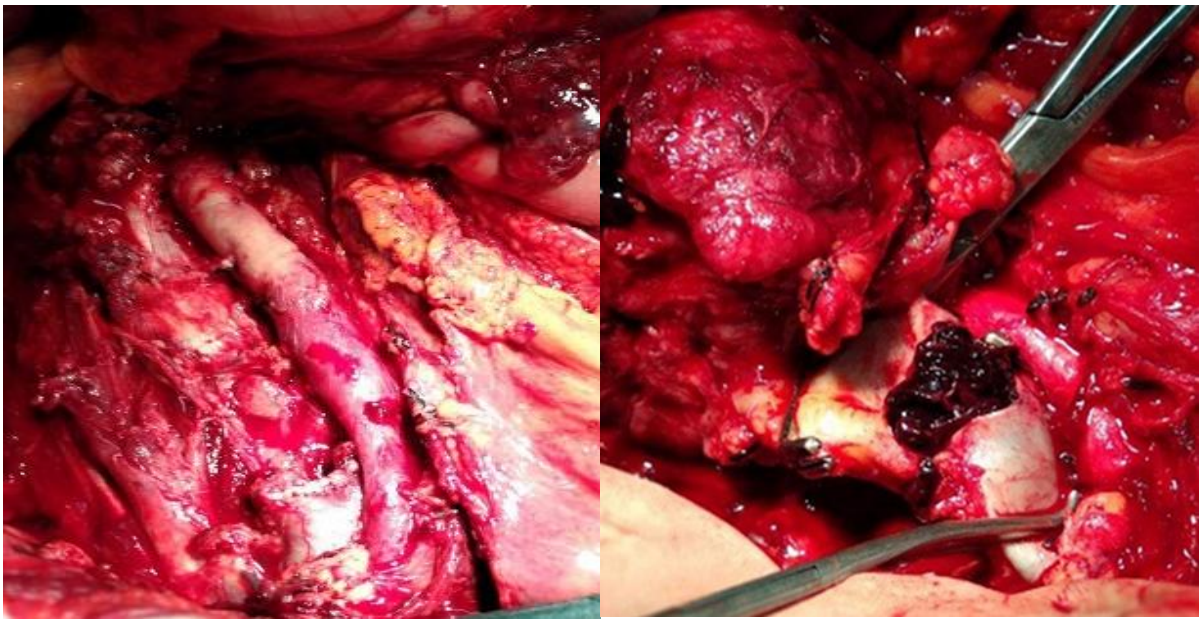


Figure 2. Aorta after Lymphadenectomy and resection of IVC with thrombus within IVC

Pathology

Macroscopically nephrectomy specimen measured 30 x 27 x 13 cm and weighed 2670 gm. Ureter measured 1.5 cm in length. Renal vessels were identified. Cut-section showed a tumor involving almost entire native renal parenchyma. The cut-surface showed a poorly circumscribed, grey, tan multilobular tumour, with multiple foci of haemorrhage and necrosis.The right adrenal was grossly involved.A tumor thrombus entered the renal vein. Haematoxylin and eosin staining showed the

tumor composed of cohesive sheets of small, uniform, primitive cells separated by fibrous bands (Figure 3). The tumor cells showed variable chromatin distribution pattern from hyperchromatic to vesicular nuclei with inconspicuous nucleoli,scant cytoplasm, high nuclear cytoplasmic ratio and round nuclei .Homer-Wright pseudorosettes were present and perivascular rosetting was noted(Figure 4). Prominent areas of necrosis were seen. Mitotic figures were readily apparent and numerous. Tumour cells infiltrated renal capsule, renal vessels, Gerota fascia and adrenal gland. Immunohistochemical evaluation revealed a diffuse FLI-1 positivity in the nuclei of neoplastic cells (Figure 5). The tumour cells were also positive for Vimentin.In contrast WT1, Synaptophysin, Chromogranin, LCA, CD10, Cytokeratin were negative. Taken with histomorphological pattern and immunohistochemical markers, a diagnosis of renal PNET was made.

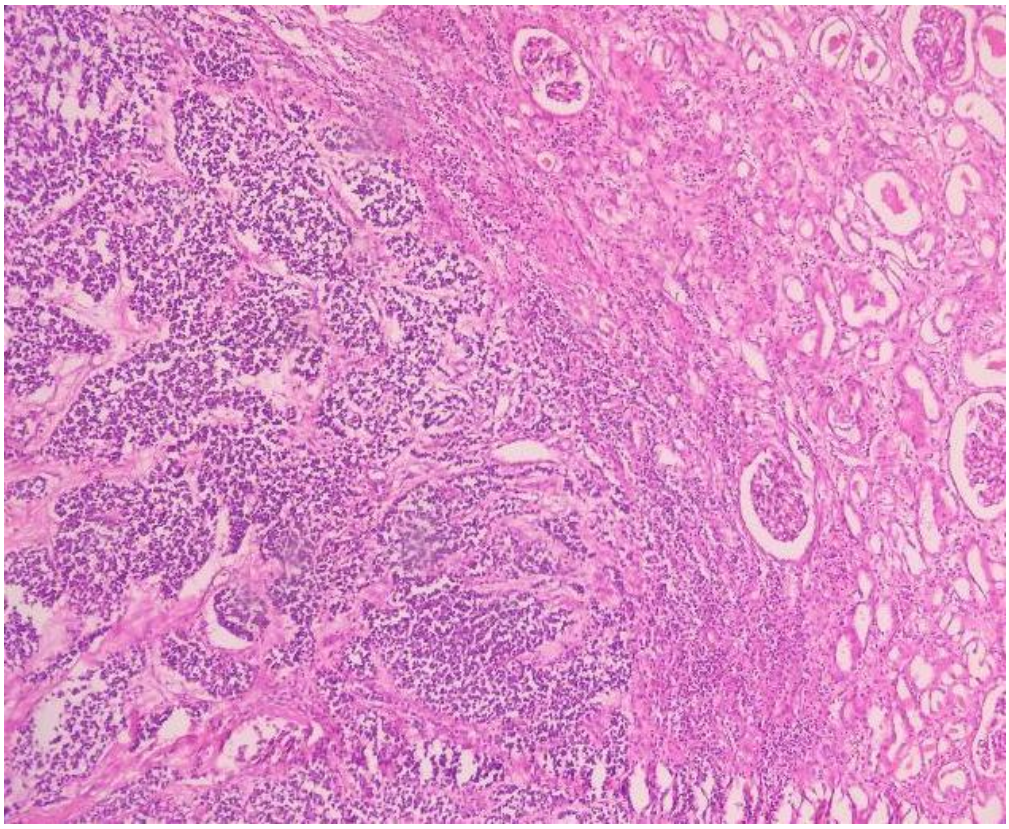
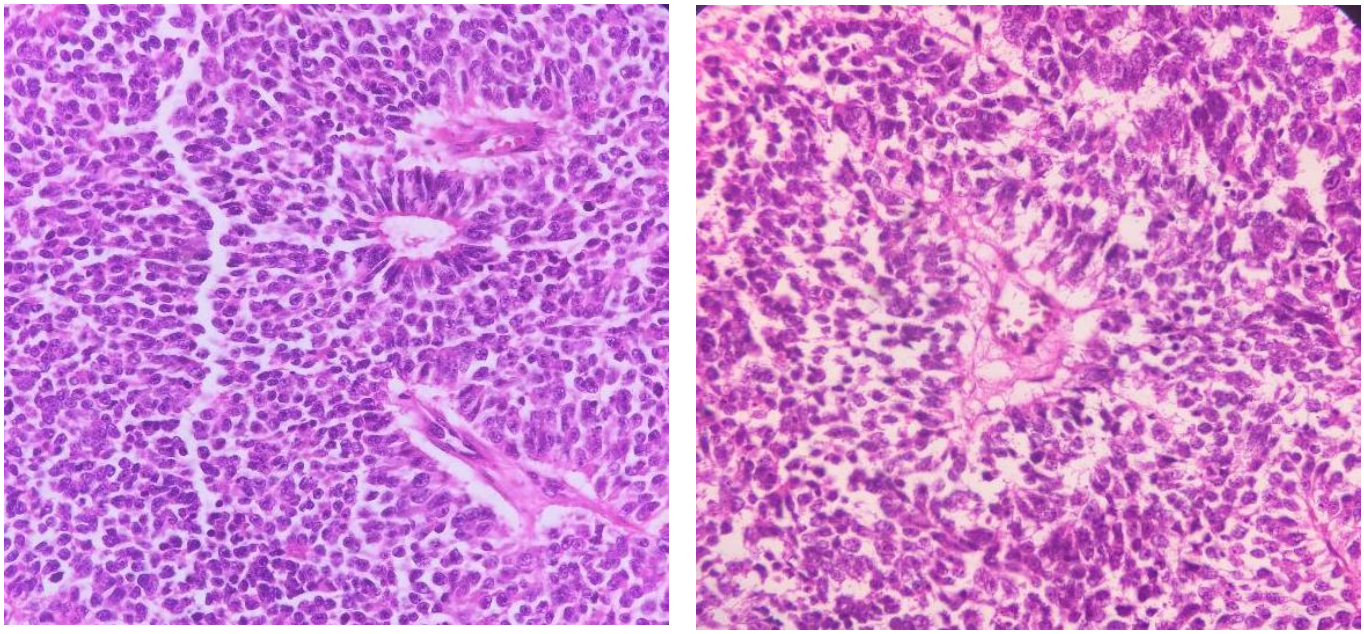


Figure 3.H&E section (x100), shows cohesive sheets of monotonous small, round blue cells with scant cytoplasm infiltrating through the renal parenchyma.



A B

Figure 4. (A)Tumor tissue showing Homer-Wright rosette and (B) perivascular pseudo rosettes. (H&E, 400X).

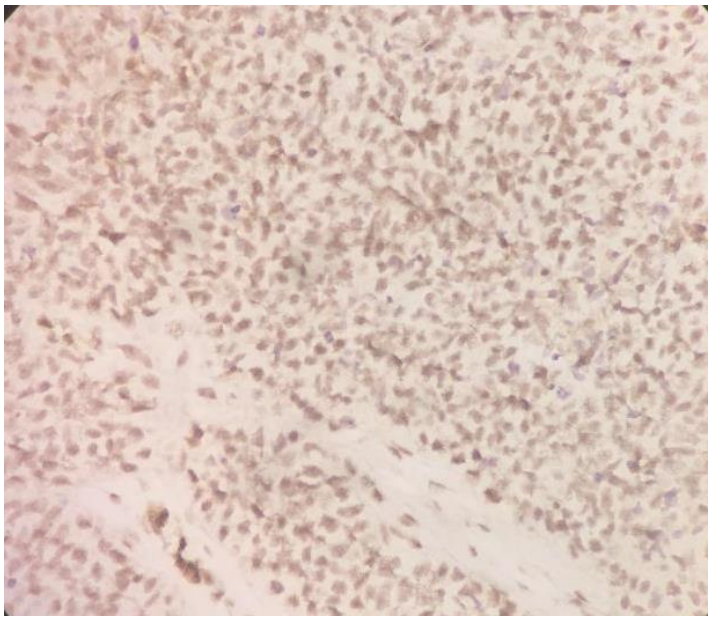


Figure 5. Immunohistochemical staining showing a diffuse expression of FLI-1 (100x).

Discussion

PNETs infrequently are seen as an organ-derived neoplasm. Less than 15 % of PNETs originate in the abdomen, retroperitoneum, and pelvis. PNET of the kidney is rare and in it there are only a handful of reported cases of PNET presenting with IVC thrombus. They belong to the Ewingsarcoma family of tumors (EFT), because of their similar histologic and immunohistochemical characteristics. The most common symptoms of renal PNETs at presentation are flank pain (67.5 %), hematuria (33.8 %), and a palpable abdominal mass (33.8 %) with fever and weight loss being infrequent symptoms.

Patients with renal PNETs tend to present with locally advanced or metastatic disease. Tumor thrombus extension has also been described in renal PNET.In our case thrombosis had extended into renal vein and ivc and seen multiple lymphadenopathies in Para aortic and interaortocaval regions.

Tumor thrombus is not a rare phenomenon in PNETs and highlighting the importance of suspecting renal PNETs.

Although these findings raise a suspicion of PNETs, no clinical characteristics pathognomonic of renal PNETs have been identified, and accurate diagnosis relies predominantly on the histopathological, IHC, and cytogenetic features of the disease.Carefully selected immunohistochemical panel is important for differentiating this tumor from other smallround cell tumors of the kidney such as rhabdomyosarcoma, neuroblastoma, clear cell sarcoma of the kidney,desmoplastic small round cell tumor (DSRCT), carcinoid tumor, nephroblastoma and Ewing's sarcoma. Imaging manifestations of peripheral PNETs include large soft tissue masses with cystic and necrotic areas and heterogeneous enhancement after intravenous administration of contrast materials.Althoughit is difficult to distinguish them from other types of soft tissue sarcomas only based on imaging, the possibility of peripheral PNETs should be considered when a large heterogeneous mass is detected in the abdomen especially in young patients with inferior vena cava involvement.

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There is no acknowledgement.

Conflict of Interest

The authors report no conflict of interest.