**Primitive Neuroectodermal Tumor of the Kidney Associated with Budd-Chiari Syndrome in a 17-Year-Old Girl**


Abstract: Renal primitive neuroectodermal tumor (PNET) is a rare and highly malignant neoplasm of the kidney. We report the case of a 17-year-old girl with renal PNET that was complicated by Budd-Chiari syndrome. She was admitted due to abrupt left flank pain and gross hematuria. Abdominal sonography and computerized tomography (CT) disclosed a large hemorrhagic left renal mass and thrombus in the inferior vena cava (IVC). Left radical nephrectomy was performed and renal PNET with tumor rupture and tumor invasion into the IVC was diagnosed based on operative findings and histologic features. Tumor cells were positive for neuron-specific enolase, chromogranin-A, and vimentin but negative for cytokeratin, leukocyte common antigen, CD3, and CD20. The thrombus in the IVC extended into the right atrium and caused obstruction of the right and middle hepatic venous outflow, which was evident on follow-up CT scan 5 months later. The patient died due to hepatic failure and progressive cardiovascular compromise 6 months after surgery. This case demonstrates that renal PNET can be life threatening when the tumor thrombus extends into the IVC and causes hepatic outflow obstruction.

Case Report

This 17-year-old girl had a 1-month history of left flank soreness and was admitted due to abrupt left flank pain and gross hematuria. Physical examination revealed a tender left abdomen and flank. Urinalysis revealed numerous red blood cells per high-power field. Roentgenographic examination of the kidneys, ureter, and bladder disclosed a soft-tissue opacity of the left abdomen, obliterating the margin of the left psoas muscle and displacing the bowel loops medially. On sonography, a large left renal mass measuring 14.4 x 11.7 x 7 cm³ was identified. Axial non-enhanced computerized tomography (CT) revealed intratumoral and perirenal hemorrhage. The tumor was heterogeneously and poorly enhanced after intravenous administration of contrast medium (Fig. A). It occupied most of the left kidney, invaded the perirenal fat, and extended into the inferior vena cava via the left renal vein.

Left radical nephrectomy was performed and revealed tumor rupture at the lower pole of the left kidney with 200 mL of blood accumulated in the perirenal space. Thrombi were found in the left renal vein and the inferior vena cava during surgery. Postoperative inferior venacavogram showed an elongated filling defect in the inferior vena cava from the level of the left renal vein to the retrohepatic segment (Fig. B). Skeletal survey and chest CT found no metastasis.
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Microscopically, the tumor was composed of numerous malignant small blue round cells (Fig. C). In focal areas, rosette-like structures were observed. Most tumor cells were positive for neuron-specific enolase, chromogranin-A, and vimentin, but negative for cytokeratin, leukocyte common antigen, CD3, and CD20. A diagnosis of PNET of the left kidney with rupture and invasion of the inferior vena cava was made. The patient was discharged after surgery, leaving the caval tumor thrombus untreated. A follow-up contrast-enhanced CT scan obtained 5 months later revealed further extension of the tumor thrombus into the right atrium and the right and middle hepatic veins, causing obstruction. She died at home due to hepatic failure and progressive cardiovascular compromise 6 months and 10 days after diagnosis.

Discussion

Renal PNETs are rare malignant tumors: only 20 cases have been reported since 1994 [2–17]. We reviewed the data of these reported cases as well as the present case to determine their clinical characteristics. There was no gender predominance for renal PNET. The right kidney was more commonly involved, with a right: left ratio of 4:3. The average age at the time of diagnosis was 27.8 years, ranging from 4 to 62 years. Most (62%) PNETs were diagnosed in the second and third decades of life [2, 4–7, 12–14, 16]. The size of tumors ranged from 4 to 24 cm, averaging 14.3 cm. Pain, palpable mass, and hematuria were the most common symptoms. Tumor necrosis was noted in 11 cases, and intratumoral hemorrhage in 10 cases [2, 4, 5, 7, 8, 10, 12–15]. Intravascular tumor extension developed in six cases, including the renal vein, inferior vena cava, right atrium, and hepatic veins [2, 5–8]. Local recurrence occurred in three cases [4, 13, 16], while distant metastasis developed in nine cases [2, 5, 6, 13, 15, 17]. The overall 1- and 2-year survival rates were 37.5% and 18.75%, respectively. Ten patients died, with a median survival time of 11 months [2–6, 8, 11]. Seven patients survived, with a median follow-up of 19.5 months [2, 9–13, 17]. In the nine cases with distant metastasis, the overall 1- and 2-year survival rates were 43% and 14.3%, respectively. Among the six cases with intravascular tumor invasion [2, 5–8], the 1-year survival rate...
was zero, with a median survival time of only 3.2 months. Chemotherapy (in 6 cases) and radiotherapy (in 3 cases) did not improve patients’ overall survival time. Among the four patients who received chemotherapy after nephrectomy (two of whom were also treated using radiotherapy), the overall survival time was 8.5 months [2–4]. In one patient who received chemotherapy and radiotherapy after biopsy only, the survival time was only 3 weeks [2].

Renal cell carcinomas are the most common renal neoplasms causing an inferior vena cava tumor thrombus and Budd-Chiari syndrome [18]. Although a renal PNET does occasionally invade the renal vein or the inferior vena cava [2, 5–7], this condition has not been previously reported in association with Budd-Chiari syndrome. Our patient had tumor invasion of the left renal vein and the inferior vena cava at the time of diagnosis. Tumor thrombectomy was suggested but was refused by the patient and her family. Continuous extension of the tumor thrombus along the inferior vena cava finally led to hepatic venous outflow obstruction and right atrial invasion, which led, in turn, to progressive deterioration of her condition.

In conclusion, failure to consider the flank pain led to delayed diagnosis of this aggressive and highly malignant tumor in our young patient. The untreated tumor thrombus in the inferior vena cava further contributed to the development of Budd-Chiari syndrome and the rapid decline of her clinical condition. Since intravascular extension of a renal PNET is an indicator of extremely poor prognosis [2, 5–8], imaging studies should be initiated to obtain an accurate diagnosis before intravascular invasion. Sonography should be performed whenever a renal tumor is suspected. CT study is superior to sonography in disclosing the extent of the tumor and its anatomic relationship with regional structures, as well as showing the presence of inferior vena cava thrombus and hepatic venous outflow occlusion. Cava tumor thrombectomy, in addition to a radical nephrectomy, is indicated in order to prevent the rapid development of hepatic outflow obstruction and pulmonary embolism [6, 7].

References

9. Grouls V: Primary, primitive (peripheral) neuroectodermal tumor (PNET) of the kidney. Pathologe 1999;15:246. [In German; English abstract]