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Renal medullary carcinoma case presenting with abdominal mass

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ABSTRACT

Renal medullary carcinoma is a highly aggressive neoplasm of the kidney that originates from renal medulla and affects most commonly young adults and can also occur in children. The presentation is usually with hematuria, abdominal pain and less commonly with weight loss, fever and abdominal mass. The prognosis is dismal. The mean survival from time of the diagnosis is 3 months. A child with renal medullary carcinoma presenting with abdominal mass is described because this tumor is very rarely encountered in childhood and can be confused with yolk sac tumor histopathologically.[Turk J Cancer 2005;35(2):96-98].

KEY WORDS:

Renal medullary carcinoma, abdominal mass

INTRODUCTION

Renal medullary carcinoma (RMC) is a very rare tumor that originates from renal medulla. It is frequently reported together with sickle cell anemia and is mostly defined in black race (1). The age of diagnosis is mostly between 10-33 years and the average is 20 years. The typical features of the tumor are mucin positivity and a tubulopapillary growth pattern that infiltrates the desmoplastic stroma although it resembles yolk sac tumor microscopically (2). The tumor grows rapidly and extends to the vascular and lymphatic structures by filling the renal pelvis. At the time of diagnosis metastases are detected in 40% of cases. After diagnosis, the survival is, on average, 3 months (3). In this article, a case of unilateral RMC which is seen rarely and may be confused with yolk sac tumor is reported.

CASE REPORT

A seven-year old boy was admitted to an outpatient clinic because of abdominal pain where an abdominal tomography revealing a solid mass of 13x12.5x11 cm in the right kidney and right nephrectomy was performed. He was sent to our Pediatric Hematology-Oncology outpatient clinic of Cerrahpaşa Medical Faculty with the diagnosis of yolk sac tumor after pathological evaluation. There was

nothing special in the patient and family histories. His physical examination was normal except for bilateral axillary lymphadenomegalies of 1 cm diameter and an incision mark at the right inferior region of the abdomen. The laboratory results were unremarkable. No sickle cell was found in the blood smear. In hemoglobin electrophoresis, abnormal hemoglobin or HbS were not detected. The alphafetoprotein and ß-hCG were in normal range. The kidney function tests, the serum electrolytes and the liver enzymes were normal. Also the whole body scintigraphy was normal. The abdominal tomography, performed before surgery revealed a solid mass detected in the right kidney that was 13x12.5x11 cm in dimensions, partially well-limited and consisting of multiloculated cystic compartments (Figure 1). Extension into the abdominal wall was also present. In the pelvic tomography multiple lymphadenomegalies, 2 cm in greatest dimension were present in the mesenteric and paraaortic regions. A soft tissue mass causing destruction and structure depression at the right half of the body and arch of the L4 vertebra was interpreted as bone metastasis (Figure 2). In the chest CT, metastatic nodules were detected in the lower lobes of both lungs (Figure 3). 'Yolk sac tumor' was diagnosed at the first pathological examination performed in the center where surgery was done. After the tumor material was reevaluated histopathologically in our center, it was diagnosed as renal medullary carcinoma with microcystic, papillary and reticular patterns. It was evaluated as stage 4 due to the presence of lung and bone metastases. Chemotherapy with vincristin 1.4 mg/m^2 once a week, actinomycine-D 15 mgr/kg/day for 5 days and concomitant radiotherapy were started. Radiotherapy was applied for 29 days with a dose of 2160 cGy to the operation region and 1080 cGy to the tumor region as boost. During the radiotherapy period, actinomycine-D was omitted, vincristine was given. While under chemotherapy since 4.5 months, the patient died due to a febrile neutropenia attack.

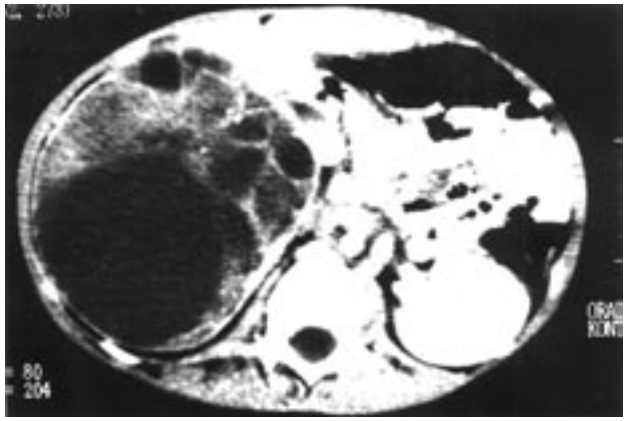

Fig 1. Large renal tumor with cystic components

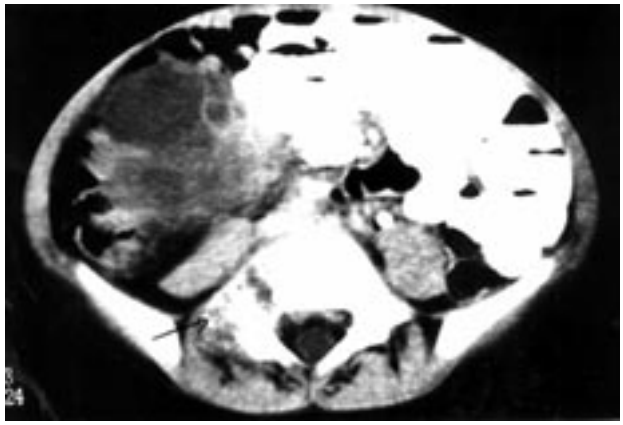

Fig 2. Bone metastasis detected in L4 vertebra

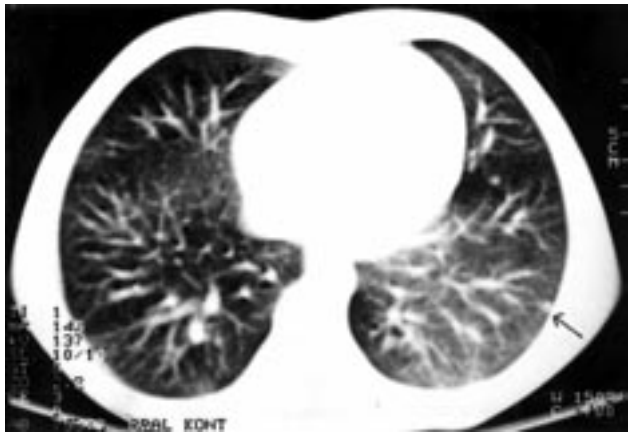

Fig 3. Multiple lung metastasis

DISCUSSION

Renal medullary carcinoma is rather an aggressive and a rare epithelial tumor that is thought to originate from the pelvic mucosa border of the kidney. Differential diagnosis

is important because this tumor is frequently confused with yolk sac tumor. The microscopic features of the yolk sac tumor are variable. Pseudopapillary and microcystic or reticular patterns are frequently detected and these patterns are characterised by eosinophilic, hyalinized intra- and extracytoplasmic globules. Tumor cells are cytokeratin, vimentin and ulex europaeus lectin positive (4). In RMC, an adenoid cystic appearance that contains reticular or dense fibroblastic stroma as yolk sac tumor does, may be present. Basic features are a tubulopapillary growth pattern that infiltrates desmoplastic stroma and mucin positivity (2). As a matter of fact our patient was also mucin positive. RMC is frequently reported with sickle cell heterozygote form. If sickle cell trait form and hematuria are present, RMC should be considered. But our patient's blood smear revealed no sickling and no abnormal hemoglobin was detected. In the literature, just like in our case, sickling is not detected in some cases (5,6). The tumor presents with macroscopic hematuria, abdominal or lateral pain and less commonly abdominal mass. In our case, abdominal swelling and lateral pain that persisted for a month were present. The interval until the diagnosis is reported is as 2-12 months, on average 4.7 months in the literature (7). Generally the tumor grows rapidly and shows regional and distant extension (8). Metastases are frequently present at diagnosis. In our case, similarly, extension to the lung, bones and pelvis were also present at diagnosis although the history was short. The prognosis of the disease is poor and the survival after diagnosis is given about 3 months (3). In published cases, the survival after diagnosis is 3-52 weeks (mean 12 weeks) (9). There is not a standard treatment regimen for this tumor. The response to chemotherapy and radiotherapy is satisfactory in some reports (5). Tumor burden was reduced by 80%, by using taxol/carboplatin in a study performed by Gollob et al (6). But there are also studies that show the ineffectiveness of the chemotherapy and radiotherapy (3). A protocol consisting of chemotherapy and radiotherapy simultaneously was started to our patient. The treatment of etoposide and actinomycin D were omitted during radiotherapy. Furthermore the wound healing after operation was not good enough. Etoposide was started after radiotherapy, at the 19^{th} day and actinomycin D at the 48^{th}day of treatment protocol, as we did not want to cause severe aplasia due to concomitant radiotherapy. The response to chemotherapy and radiotherapy were not good in our patient and we learnt that he had died at home 5 months after the diagnosis, due to an attack of febrile neutropenia. In conclusion RMC is a rare childhood tumor which should be considered in cases presenting with renal mass and pathologically diagnosed as yolk sac tumor, furthermore sickle cell anemia is not always present.

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