Case Report

Botryoid-type of embryonal rhabdomyosarcoma of renal pelvis in a young woman

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Abstract

A 22-year-old woman presented with three weeks history of intermittent left loin pain, on radiological evaluation by U\S and MRI revealed left renal pelvic mass, ureterorenoscopy and biopsy taken, but couldn't reveal definitive diagnosis other than presence of a malignant process. Left nephroureterctomy was performed. Grossly there is a polypoid mass attached to the upper pole of the kidney by stalk. Light microscopic examination and immunohistochemical staining confirm a diagnosis of Botryoid-type of embryonal rhabdomyosarcoma. Treatment and follow up to 1 year is mentioned. Reviewing the literature the presented case is the second of this tumor in adult renal pelvis.

Key Words: Botryoid, nephroureterctomy, rhabdomyosarcoma

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INTRODUCTION

Rhabdomyosarcoma (RMS) is extremely rare malignancy in adult.[1-3] It usually occurs in the head, neck, genitourinary tract and extremities. Botryoid-type of embryonal rhabdomyosarcoma in the kidney is so rare that only one case is documented in literature.[2] Depending on histological examination RMS is classified into three types, embryonal, alveolar and pleomorphic type.[3,4] A nonspecific radiological appearance precludes the pre-operative diagnosis mandating surgical intervention for a definitive management.

CASE REPORT

A 22-year-old female patient complaining of left loin pain which was colicky in nature without other urinary tract symptoms or history of trauma. Physical examination was unremarkable except for mild left loin tenderness, her hematocrit and kidney function laboratory test were normal and urine analysis showed microscopic hematuria. Ultrasonographic examination reveals left renal pelvic soft tissue mass around 3 × 4 cm [Figure 1], MRI confirms this finding and a tumor is confined to the left renal pelvis without evidence of abdominal lymphadenopathy [Figure 2a and b]. Left retrograde pyelography showed renal pelvic filling defect and lower group calyx hydronephrosis [Figure 3]. Ureterorenoscopy could confirm tumor and biopsies were taken, but histopathology report did not give definitive diagnosis other than the presence of a malignant process.

A left nephureterectomy with transurethral ureter stripping and ureter ostium resection on a presumption of transitional cell carcinoma is carried out. The surgical resection specimen showed a polypoid tumor with a grape-like appearance in the left renal pelvis [Figure 4].

Microscopically there was a condensation of epitheloid to spindle cells underneath the basement membrane, forming the cambium layer. Immunohistochemical examination stains for

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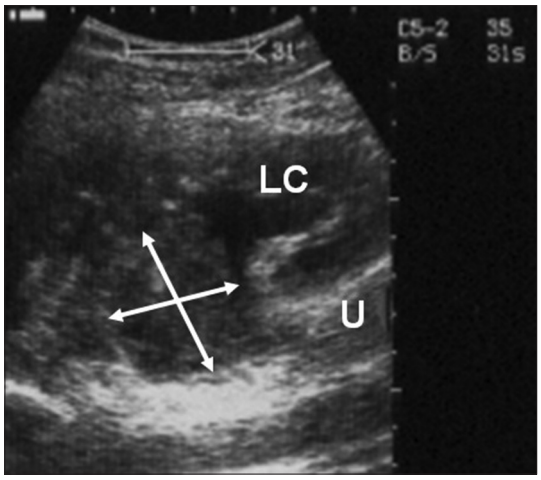

Figure 1: Ultrasound of renalpelvic mass (arrows). LC = Lower calices, U = Ureter

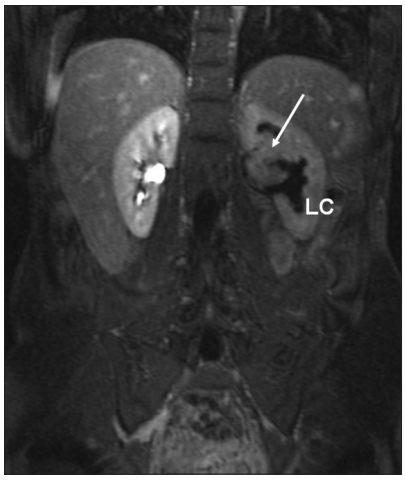

Figure 2a: MRI IVP coronal view mass in left renal pelvis (arrow), dilatation of lower calices (LC) without excretion

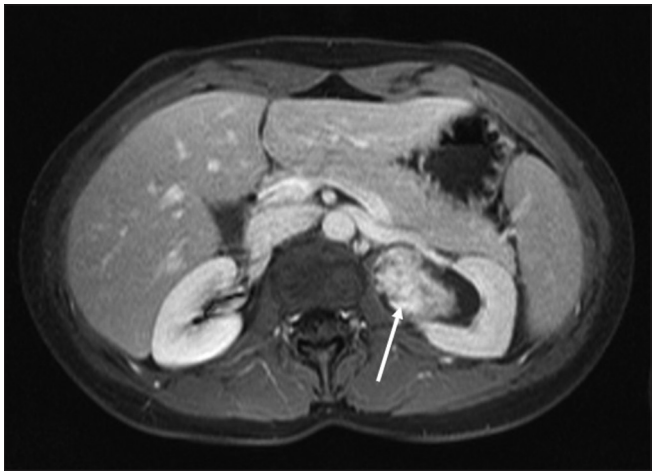

Figure 2b: MRI cross section view left renal pelvic mass (arrow)

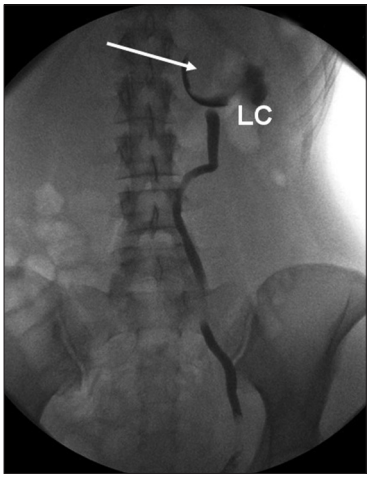

Figure 3: Left retrograde ureteropyelography filling defect (arrow) of renal pelvis, LC = Lower calices

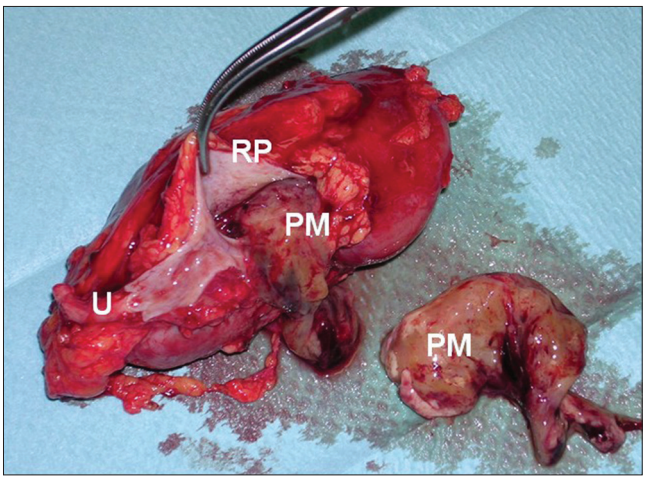

Figure 4: Nephrectomy - specimen with opend renal pelvis. U = ureter, RP = Renal pelvis, PM = Renal pelvis mass

vimentin, WT1, CD117, CD99, HMB45, CEA, CK7, and S100 were negative, stains for desmin, MYO D1 and actin, confirming the diagnosis of Botryoid-type of embryonal rhabdomyosarcoma.

DISCUSSION

Rhabdomyosarcoma is the fourth most common solid tumor and the most common soft tissue sarcoma in children constituting more than 50% of cases. In contrast, the tumor is exceedingly rare in adult.[1,3,4] It can occur in a variety of organs and tissue, including those that lack striated muscle.[1,4]

Nearly 40% of all RMS occur in the head and neck region, one-third occurs in the genitourinary region, while the remaining of tumors appears in the trunk and extremities.[5,6] Genitourinary tract RMS arise most commonly in the urinary bladder, prostate, vagina, uterus, and as paratesticular disease.[1,4] Its appearance in the renal pelvis has only reported twice, one shows botryoidal-embryonal differentiation and the other a pleomorphic type.[2,6]

Tumors generally arising from renal pelvis are mainly transitional cell carcinoma (85-90%).[7] Others are squamous cell carcinoma

and adenocarcinoma. In expectation of a common tumor of the renal pelvis a classical management for this patient including ureter and its ostium resection is chosen. Only the final histology could confirm the diagnoses of botryoid RMS. In adult the presented case is the second descript in the Literature.

RMS can be divided into three distinct types; pleomorphic or undifferentiated, alveolar and embryonal.[5] Pleomorphic RMS, the most frequently occurring tumors, affect the older age groups, this lesion usually occurs in males on their extremities, but may occur in any site. Alveolar RMS is an extremely malignant subgroup, it occurs mostly in adolescents and young adults. Embryonal RMS, primarily a lesion of the head and neck region, especially the orbit, is a type of tumor usually occurs in youngsters and at the age of 10 years. This tumor is characterized by a mixture of round cells and cells with an eccentric nucleus, so called "tadpole" or tennis "racquet".

Sarcoma botryoides is the only RMS features a grossly characteristic appearance and presents itself as a polypoidal mass. Primarily of the genital and urinary tract, this tumor is a macroscopic morphological characterization of the embryonal type.[5]

The tumor cells composing embryonal RMS variably exhibit all cellular phases of myogenesis, from stellate undifferentiated mesenchymal cells to elongated myoblast, multinucleated myotubes, and fully differentiated myofibers.[7]

In the 21st century, immunohistochemistery has attained popularity as the foremost method of diagnostic confirmation. A variety of muscle markers have been identified: Myoglobin, one of the first markers used, suffers from lack of sensitivity while only in well differentiated cells expressed. Desmin and Actin have been used most extensively used, MyoD and myogenin are the most sensitive and specific groups of protein useful in immunohistochemical diagnosis of RMS. Pattern of staining appears to predict classification, as a heterogeneous pattern is more typical of embryonal RMS, whereas strong diffuse staining is a feature of alveolar RMS.[7]

Treatment approaches to RMS include surgery, radiation therapy and chemotherapy. Depending on the extreme seldom appearance of this tumor entity it is difficult to give a recommendation about best treatment options in adults.

In the presented case suspected malignancy uncertain of specification, flank pain, hematuria and a kidney without excretion of contrast in MRI leads to perform radical surgery.

Other treatment options in RMS are, in addition to radical surgery, radiotherapy and chemotherapy. To avoid relapse it is proposed to use adjuvant prophylactic chemotherapies and local radiotherapy. In children, surgery alone resulted in survival rates less than 20%, associated to suspected micrometastatic disease. The development of adjuvant chemotherapy has shown increased survival rates in patients with localized disease up to 60%. The golden standard of chemotherapy in children is vincristine (V), actinomycin (A) and cyclophosphomide (C), so called VAC. Radiotherapy is suggested for local recurrence or incomplete resection as an eradication using a dosage of 4,000 to 4,500 cGy with possibility of escalation to 5,000 cGy in residual masses >5 cm. Patients with completely resected RMC receives no radiotherapy.[8]

The prognosis is highly associated with the tumor location, local extension, size, regional nodal involvement, and metastatic condition. The main metastatic sites are pulmonary and skeleton, recurrences in RMS mainly appeared during the first 2 years.[9]

In the presented case clinical and radiological (ultrasound and MRI) investigation could not show any suspicion of metastasis. In consideration of resection without regional lymph node involvement, without local recurrence and a metastasis free follow-up for 6 months, up to now we do not see an indication for adjuvant therapies. Bearing in mind the aggressive behavior of these tumors, a strong and close follow-up is necessary.

We present the history and radiological finding of a Botryoid-type embryonal RMS which managed by nephroureterectomy and surveillance up to 1 year. The possibility of cure for this young patient with localized disease encourage to delay toxic therapies having the potential of development of long-term morbidity due to side effects of specific chemotherapy agents and avoiding the risk of secondary neoplasms.

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