Pelvic pleomorphic rhabdomyosarcoma presenting as oliguria in a 61-year-old woman

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Rhabdomyosarcomas (RMSs) are malignant soft-tissue tumors arising from skeletal muscle progenitor cells.[1] They are most commonly diagnosed in children and adolescents and are rare in adults. These tumors arise from a variety of anatomical sites, including the head and neck, urogenital tract, and extremities. Classification of RMSs depends on histopathologic and immunohistochemical features. Embryonal and alveolar subtypes are more common in children and adolescents, whereas the pleomorphic subtype is seen almost exclusively in adults. Adult RMS is associated with poor outcomes and high recurrence rate.[2,3] Because of the low incidence of adult RMS, most published reports of RMS in adults are either case series or retrospective analyses, and most treatment protocols are extrapolated from clinical trials performed in children. The present report describes a 61-year-old woman with RMS whose presentation included atypical symptoms.

Case presentation

A 61-year-old African American woman was transferred to Tulane Medical Center in New Orleans, LA, for care. She had presented to an outside hospital emergency department complaining of significantly decreased urine output for 3-4 days. On questioning at presentation, she also related progressively worsening pelvic pain that had started 1 year previously. Past medical history was significant for hypertension, type 2 diabetes mellitus, and hyperlipidemia. Past surgical history was significant for a total abdominal hysterectomy secondary to menorrhagia and a bilateral tubal ligation. Family history was significant for numerous malignancies including lung, thyroid, colon, and breast cancer. A computed-tomography (CT) scan of the abdomen and pelvis showed a large pelvic mass compressing the ureters bilaterally.

The patient was subsequently transferred to our facility for further care. Upon initial physical exam, the patient had diffuse abdominal tenderness and a large abdominal-pelvic mass. Bilateral ureteral stents were placed, and a needle biopsy of the mass was done. Pathologic exam revealed nondiagnostic necrotic tissue. Exploratory laparotomy was then performed to obtain tissue for diagnosis and possible excision. Intraoperative findings showed a pelvic mass of about 15 x 15 cm that seemed to be invading both the bladder and the sigmoid colon, which was displaced to the right side of the pelvis. Biopsy of the mass showed a pleomorphic rhabdomyosarcoma. No malignant epithelial component was identified. After discussion with the family, neoadjuvant chemotherapy was planned, followed by possible tumor resection.

About 3 weeks later, just before she was due to start chemotherapy, the patient developed colon obstruction secondary to tumor compression. She underwent emergent transverse loop colostomy, and it was noted that the tumor had increased in size, measuring about y 20 x 20 cm, and was now accompanied by malignant ascites. Postoperatively, the patient experienced relief from her obstructive symptoms, but developed abdominal distension secondary to the ascites, and her condition deteriorated. After extensive counseling regarding the poor prognosis of this condition, the patient and her family opted for outpatient hospice care.

Histopathology

Pleomorphic RMS is a high-grade sarcoma occurring

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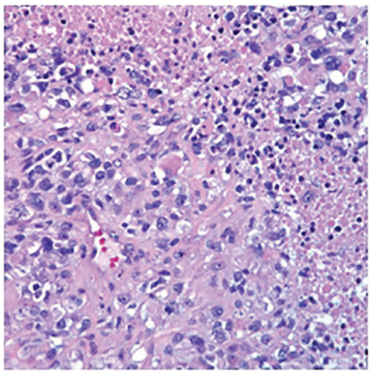

FIGURE 1 Histological features of pleomorphic rhabdomyosarcoma. Hematoxylin and eosin stain, 400x. Pleomorphic epithelioid and spindle cells in a necrotic background. Some cells display prominent acidophilic cytoplasm representative of rhabdoid or rhabdomyoblastic cell differentiation.

almost exclusively in adults consisting of bizarre polygonal, round, and spindle cells, which display evidence of skeletal muscle differentiation. The presence of pleomorphic polygonal rhabdomyoblast on routine hematoxylin and eosin stains coupled with immunohistochemical evidence of at least one skeletal muscle-specific marker by immunohistochemistry is required for diagnosis. Imunohistochemical markers for pleomorphic rhabdomyosarcomas include: myoglobin, MyoD1, skeletal muscle myogenin, fast (skeletal muscle) myosin, and desmin. These tumors show variable expression for muscle-specific actin, smooth-muscle actin, and myogenin and lack expression of epithelial markers (eg, AE1/AE3, CK7, CK20) and S-100 protein.

Our case showed an extensively necrotic pleomorphic epithelioid and spindle cell malignant neoplasm. In areas the tumor cells had rominent acidophilic cytoplasm with focal apparent cytoplasmic filaments. Those features were consistent with rhabdoid or rhabdomyoblastic differentiation. Immunohistochemical stains showed positivity for myogenin, desmin, vimentin, muscle-specific actin, CD68 (variable intensity), and focal staining for EMA and CD99. The tumor was negative for pankeratin (AE1/AE3), CK7, CK20, HMB45, Melan-A, CA125, S100 protein, p63, smooth-muscle actin, CD117, CAM 5.2, and CD45. In addition, the Ki-67 shows a proliferative index of about 80%. The aforementioned histologic findings and immunohistochemical profile support the diagnosis of pleomorphic RMS in our patient.

Discussion

The differential diagnosis of subjectively decreased urine output in an ambulatory woman is broad and includes both primary nephropathies (decreased perfusion because of hypovolemia, myocardial or valvular dysfunction, drug reactions; or intrinsic kidney injuries such as acute tubular necrosis, small-vessel disease, acute interstitial nephritis), and obstructive uropathy (ureteral stones, retroperitoneal fibrosis, or external ureteral/bladder compression from a tumor). The diagnostic evaluation must be done carefully and promptly, because failure to alleviate oliguria in a timely fashion may result in renal failure and uremia.

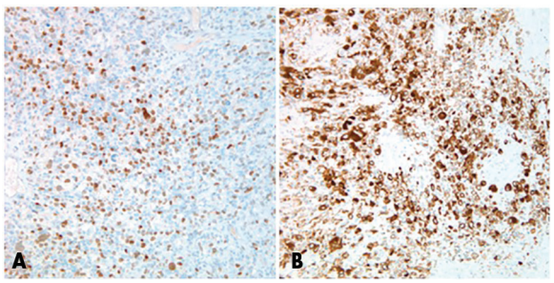

FIGURE 2 Immunohistochemical features of pleomorphic rhabdomyosarcoma. A, Myogen, 200x; and B, Desmin. Strong and diffuse staining with these markers support the diagnosis of pleomorphic rhabdomyosarcoma.

The initial workup should therefore include pelvic examination performed by an experienced clinician with confirmation of possible sources of external ureteral compression by radiologic evaluation. This is particularly important in patients with additional complaints of chronic or worsening pelvic or abdominal pain, as in the present case. Even though the patient's tumor was advanced and had a rapid rate of progression, she experienced symptomatic relief from the ureteral stent placements and transverse loop colostomy. Further, when a tumor mass is identified as the cause of an obstructive uropathy, prompt tissue diagnosis is also important. The presence of an advanced malignancy with a very poor prognosis, as in this case, will influence subsequent treatment decisions.

Rhabdomyosarcoma is a rare malignant neoplasm in adults, and the pleomorphic subtype is associated with the worst prognosis.[2] Here, a 61-year-old woman who was ultimately diagnosed with pleomorphic RMS presented to the emergency department with a chief complaint of subjectively decreased urination. The authors' literature review identified no other cases of adult RMS presenting with oliguria. Rare pelvic tumors such as RMS will occasionally be associated with subjective oliguria and should be considered during the initial evaluation, because prompt diagnosis can allow for both immediate symptomatic relief and a realistic discussion of prognosis.

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