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Case study

Adult extrarenal Wilms tumor of the uterus with teratoid features

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Summary The present article reports for the first time a case of an extrarenal teratoid Wilms tumor in the uterus of a 62-year-old woman. It had triphasic histology with epithelial areas composed of metanephric tubules harboring glomerular structures, adamantine patterns, neural type rosettes, blastema, and a primitive, myxoid type stroma. Abundant heterologous elements such as cartilage, striated muscle, squamous epithelium, and an alpha fetoprotein and TTF1-positive early endodermal epithelium were also present. Immunohistochemistry in Wilms tumor areas showed positivity for markers also indicative of peripheral primitive neuroectodermal tumors such as neuron-specific enolase, CD99, and CD56. However, nuclear positivity for Wilms tumor antigen together with the presence of glomeruli and the absence of endometrioid tumor areas and the organoid arrangement of tissues excluded peripheral primitive neuroectodermal tumors, carcinosarcoma, and teratoma, respectively. Although the diagnosis of female genital tract Wilms tumors is difficult in cases where glomerular structures are lacking, it should be considered because these neoplasms have a better therapeutic response than peripheral primitive neuroectodermal tumors and carcinosarcoma.

Müllerian carcinosarcomas are the most frequent uterine mixed tumors. Histologically, they exhibit various malignant epithelial and mesenchymal mesodermal tissue components, although on rare occasions, they may acquire teratoid features and differentiate various neuroecto- or endodermal components [1,2].

Wilms tumors (WTs) are characteristic mixed neoplasms that may infrequently arise in the female genital tract [3,4], where they can reproduce every histologic pattern present in eutopic nephroblastomas. When renal WTs differentiate large amounts of heterologous components, they are called teratoid WTs (TWTs) [5], and to date, they have been described in children in 16 instances [6-8].

This article reports for the first time a case of an extrarenal location of a TWT. It occurred in the uterus of a 62-year-old woman and had extensive mesodermal and embryonal-type endodermal components. Morphology and immunohistochemistry were crucial in the differential diagnosis with various mixed uterine tumors such as primitive neuroectodermal tumors, carcinosarcoma, and teratoma.

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1. Case report

This nulliparous 62-year-old woman had an episode of profuse vaginal bleeding. She was referred to a regional hospital where she was seen to have a large uterine hemorrhagic polypoid mass protruding through the cervix. Surgery revealed an enlarged globular uterus without pelvic adhesions or ascites, and a total abdominal hysterectomy with salpingo-oophorectomy was performed. An initial histopathologic diagnosis of peripheral primitive neuroectodermal tumor (pPNET) was considered. The case was sent to us for consultation (FNF), and a diagnosis of extrarenal WT was made. Postoperatively, nuclear magnetic resonance (NMR) and positron emission tomography (PET) scan studies failed to show any pelvic or distant metastases. The patient underwent chemotherapy with 4 cycles of Cisplatin (20 mg/m^2) and Ifosfamide (4.5g/m^2) followed by radiotherapy with total dose of 50 Gy and is alive and well without evidence of disease 14 months later.

2. Materials and methods

Tissues from 18 samples of tumor were fixed in 10% neutral buffered formalin and paraffin embedded. Five-microgram-thick sections were stained with hematoxylin and eosin. Immunohistochemical studies with an avidin-biotin peroxidase complex method used antibodies directed against the following: CD56 (Master Diagnostica [MD], Granada, Spain; monoclonal 56C04, diluted), CD57 -Leu-7- (MD, monoclonal HNK-1, diluted, CD99 MD, monoclonal 013 diluted), neu-n (Chemicon International, Temecula, CA, USA; monoclonal MsX Neu-N, dilution 1:200), synaptophysin (MD, monoclonal SP11, diluted), neuron-specific enolase (NSE; MD, monoclonal E27, diluted), glial fibrillary acidic protein (GFAP; MD, polyclonal, diluted), chromogranin (MD, monoclonal LK2H10, diluted), alpha fetoprotein (AFP; MD, monoclonal ZSA06, diluted), TTF1 (MD, monoclonal 8G7G3, diluted), Wilms tumor antigen (WT1; Dako Cytomation, Denmark; monoclonal 6F-H2 1:50), CAM5.2 (MD, monoclonal CAM 5.2, diluted), cytokeratin 20 (MD, monoclonal Ks20.8, diluted), cytokeratin 7 (MD, monoclonal OVTL-12/30 diluted), α-actin (MD, monoclonal HHF35, diluted), desmin (MD, monoclonal D33, diluted), and myoglobin (MD, polyclonal, diluted).

3. Results

3.1. Pathology findings

Macroscopically, the uterus revealed an 8-cm sessile, friable, hemorrhagic mass in its fundus that protruded into the endometrial cavity and filled the cervical canal but on cut section was seen to invade only superficially a markedly dilated and thinned myometrium. The adnexa were involutive but otherwise unremarkable.

Microscopically, myometrial invasion was reduced to 3 mm. No areas of endometrioid differentiation were seen other than the normal endometrial glands embedded within the neoplasm, which had extensive necrosis and hemorrhage but had areas of viable tissue both in the intracavitary mass and endometrium, revealing an architecture of a triphasic WT with abundant admixed heterologous components.

Blastematous areas were characterized by masses of closely packed cells with indistinct basophilic cytoplasm (Fig. 1), which were surrounded by a loose myxoid stroma and arranged in nests or sheets (Fig. 1). Epithelial areas were frequently arranged in clusters of narrow metanephric-type tubules, some of which were associated with microcystic structures containing primitive, irregular renal glomerular tufts (Fig. 2). Often, they were surrounded by a condensation of blastemal or mesenchymal cells arranged in concentric layers. An adamantine trabecular pattern was also prominent (Fig. 2). In some areas, primitive tubules formed rosettes analogous to those found in neuroblastic tumors (Fig. 2). Only in the cervical canal were there mucosal implants that corresponded solely to blastema.

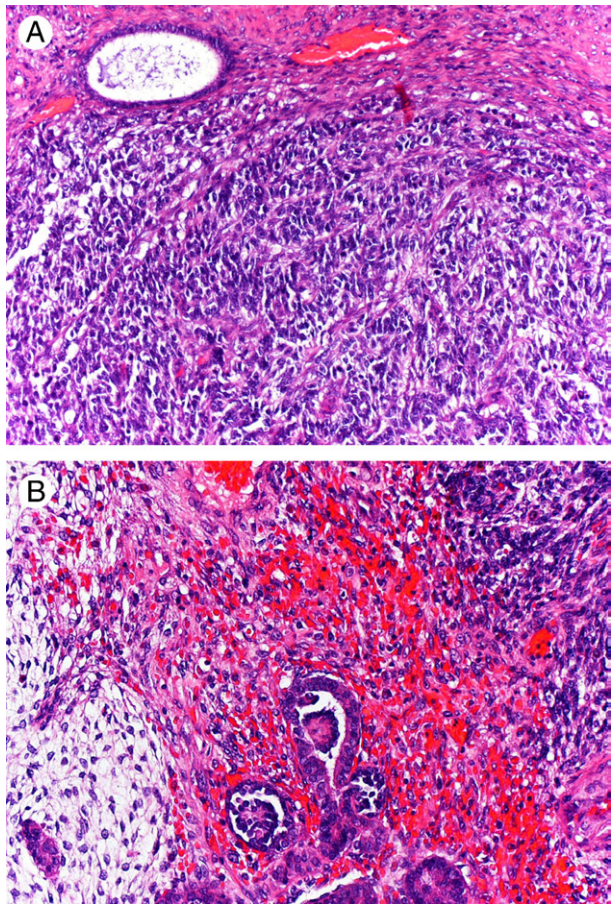

Fig. 1 Area composed of blastema in uterine cavity. A, An endometrial gland is seen at the periphery. B, Blastema, occasional tubules with glomeruli and myxoid stroma. (Magnification ×200.)