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Current topics

Soft tissue tumors of the urinary bladder Part II: malignant neoplasms

Sarah Lott MD[a], Antonio Lopez-Beltran MD[b,c], Rodolfo Montironi MD[d], Gregory T. MacLennan MD[e], Liang Cheng MD[a,b,c,*]

[a]*Department of Pathology and Laboratory Medicine, Indiana University School of Medicine, Indianapolis, IN 46202, USA*
[b]*Department of Urology, Indiana University School of Medicine, Indianapolis, IN 46202, USA*
[c]*Department of Pathology, Cordoba University, E-14004 Cordoba, Spain*
[d]*Institute of Pathological Anatomy and Histopathology, School of Medicine, Polytechnic University of the Marche Region (Ancona), United Hospitals, 60020 Ancona, Italy*
[e]*Department of Pathology, Case Western Reserve University, Cleveland, OH 44106, USA*

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Malignant peripheral nerve sheath tumor;
Hemangiopericytoma

Summary Most bladder tumors arise from the urothelium. However, there are several uncommon but significant malignant bladder lesions that must be differentiated from urothelial carcinomas and from benign lesions of the bladder. The second half of this two-part review will describe rare nonurothelial malignant tumors of the urinary bladder including leiomyosarcoma, rhabdomyosarcoma, angiosarcoma, malignant fibrous histiocytoma (undifferentiated sarcoma), primitive neuroectodermal tumor, malignant peripheral nerve sheath tumor, hemangiopericytoma, and alveolar soft-parts sarcoma. Common clinical presentations, morphologic characteristics, and immunohistochemical features are described to aid the practicing pathologist in the identification of these entities. Because the distinction between malignant and benign lesions has significant therapeutic and prognostic implications, key factors for differentiating them are presented.

1. Introduction

Most primary bladder cancers are transitional cell (urothelial) carcinomas; cases of squamous cell carcinoma, primary adenocarcinoma, or small cell carcinoma are encountered much less frequently. All other bladder cancers are rare and have been described in small series and isolated case reports. In last month's issue of *Human Pathology*, we

* Corresponding author. Department of Pathology and Laboratory Medicine, Indiana University School of Medicine, Indianapolis, IN 46202, USA.
E-mail address: liang_cheng@yahoo.com (L. Cheng).

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presented "Soft tissue tumors of the urinary bladder part I: myofibroblastic proliferations, benign neoplasms, and tumors of uncertain malignant potential" [1]. The purpose of Part II of this two-part review is to describe various malignant mesenchymal lesions of the bladder, describe the clinical setting in which these lesions typically occur, and present the differential diagnosis and distinguishing features, including immunohistochemical staining patterns, of these rare but fascinating lesions. Special emphasis is placed on differentiating malignant spindle cell lesions from myofibroblastic proliferations as well as from sarcomatoid carcinoma, as they have differing prognostic as well as therapeutic implications.

1.1. Leiomyosarcoma

Although leiomyosarcoma is the most common malignant mesenchymal tumor of the urinary bladder in adults, it is still relatively very rare, accounting for less than 1% of all bladder malignancies [2]. Patients range in age from 15 to 75 years, with most patients presenting in the sixth to eighth decades [3,4]. There is a male predominance of over 2:1 [5]. Some leiomyosarcomas have reportedly developed 5 to 20 years after the administration of cyclophophamide [6,7]. Acrolein, a degradation product of cyclophosphamide, is thought to be the causative agent in such cases. The most common presenting complaint (in 80%) is gross hematuria; less frequently, patients complain of dysuria or obstructive voiding symptoms or are noted to have an abdominal mass. Any part of the bladder may be involved by leiomyosarcoma, but the dome followed by the lateral walls are the most common tumor sites [3]. A recent review of 18 bladder leiomyosarcomas showed them to be "aggressive neoplasms" with more than 60% of patients developing metastases or dying of recurrent or metastatic tumor [2]. Higher-grade tumors had a worse prognosis. Most low-grade tumors have lower risk of recurrence or metastasis [2,3]. When feasible, treatment generally involves surgical excision of the lesion (Fig. 1).

Grossly, leiomyosarcomas are often large and polypoid; they are unencapsulated and usually exhibit full thickness involvement of the bladder wall. They are described variously as firm, fleshy, or fibrous masses that may have mucoid or myxoid consistency, hemorrhagic and/or focally necrotic areas, and often surface ulceration [2,8].

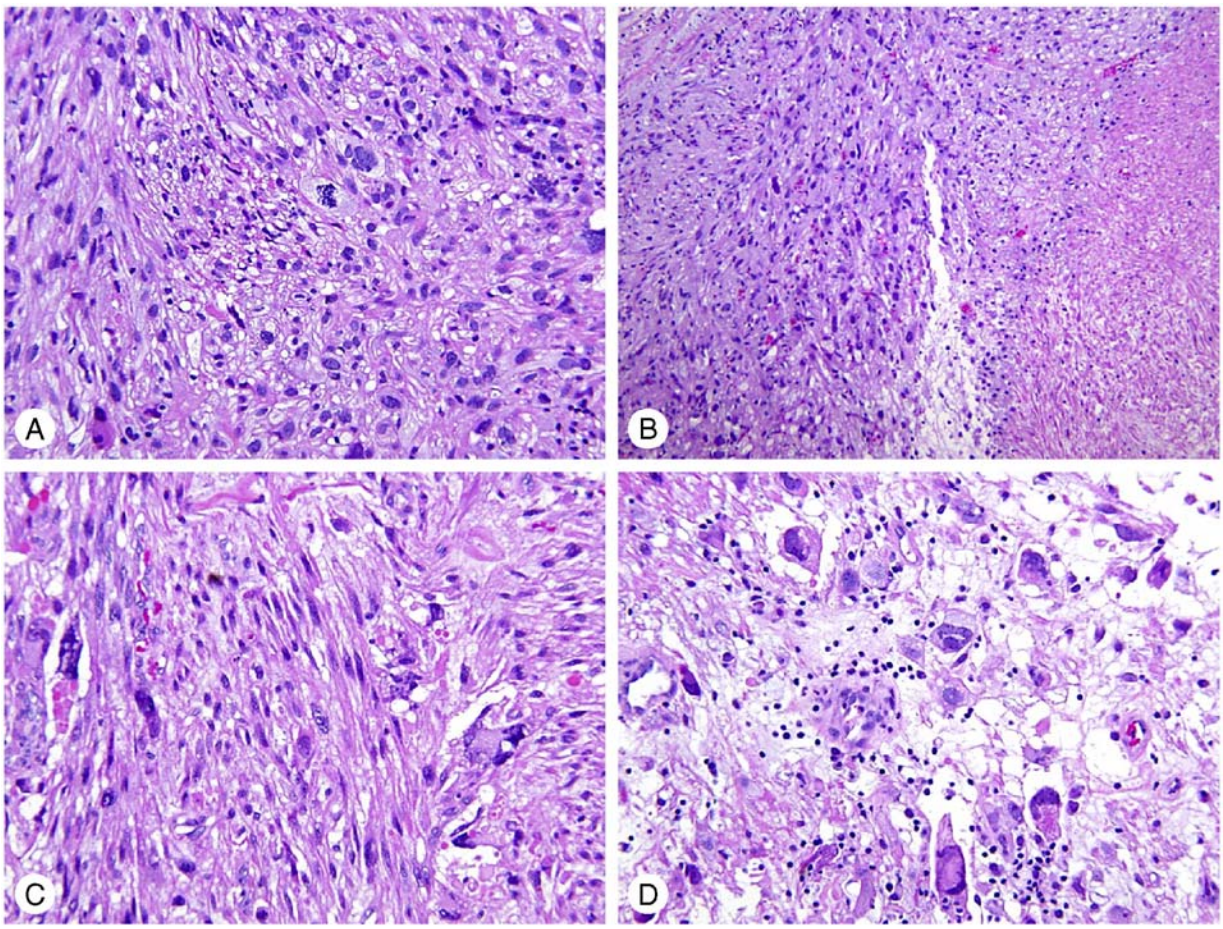

Fig. 1 Leiomyosarcoma. A, Leiomyosarcoma of bladder with significant nuclear pleomorphism, hyperchromasia, and bizarre mitosis. B, Leiomyosarcoma with area of necrosis. C, Tumor with significant nuclear atypia. D, Tumor with nuclear atypia, macronucleoli, inflammation, and multinucleated giant cells.