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Current topics

Soft tissue tumors of the urinary bladder, part I: myofibroblastic proliferations, benign neoplasms, and tumors of uncertain malignant potential

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Summary Most bladder tumors arise from the urothelium. However, there are several uncommon but significant bladder lesions that must be differentiated from urothelial carcinomas. These include both benign and malignant spindle cell lesions. The first half of this 2-part review will describe benign myofibroblastic proliferations including inflammatory myofibroblastic tumor and postoperative spindle cell nodule; benign neoplasms including leiomyoma, hemangioma, neurofibroma, and schwannoma; and tumors of uncertain malignant potential including paraganglioma, granular cell tumor, and perivascular epithelioid cell tumor. Common clinical presentations, morphological characteristics, and immunohistochemical features are described to aid the practicing pathologist in the identification of these entities. This review also describes current theories as to the pathogenesis of inflammatory myofibroblastic tumor and postoperative spindle cell nodule and details the current molecular markers identifying several of these lesions.

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. The rest of bladder cancers are all rare and have been described in small series and isolated case reports. Benign spindle cell lesions of the bladder are interesting and enigmatic entities, and the exact nature of some of these lesions is still undetermined. Two tumors in this category of supposed myofibroblastic origin are the postoperative spindle cell nodule (PSCN) and the inflammatory myofibroblastic tumor (IMT). Other benign lesions of the bladder include leiomyomas, hemangiomas, and neurofibromas, with other examples of benign soft tissue tumors very rarely described. Recognizing these

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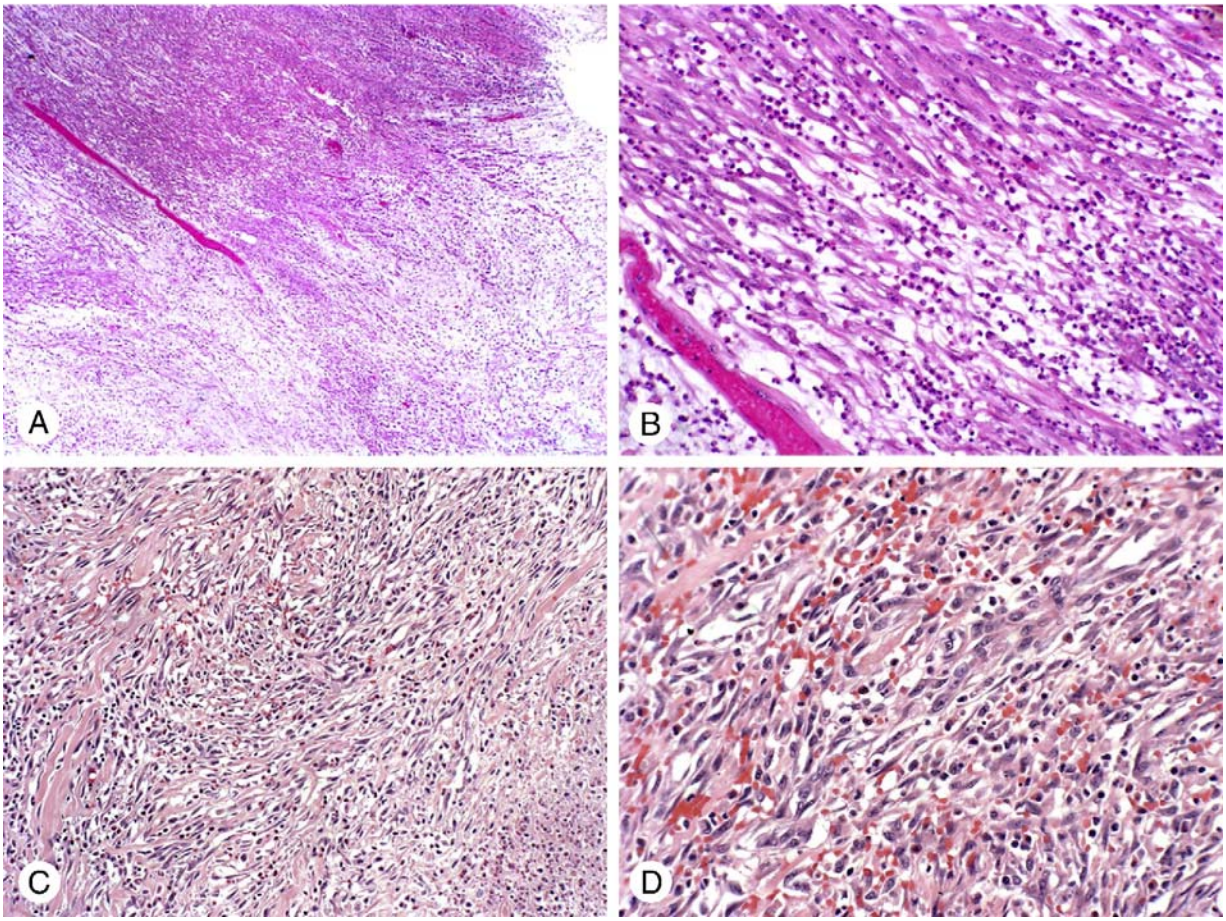

Fig. 1 IMT of the bladder is shown. A, The tumor displays spindle cells and a loose, edematous stroma. B, Scattered chronic inflammatory cells are seen. C and D, Mild cellular atypia, prominent blood vessels, and extravasated red blood cells are seen.

spindle cell lesions and differentiating them from sarcomatoid carcinoma are important, as they have differing prognostic as well as therapeutic implications.

The purpose of this review is to describe various benign mesenchymal lesions of the bladder, to describe the clinical setting in which these lesions typically occur, and to present the differential diagnosis and distinguishing features, including immunohistochemical staining patterns, of these rare but fascinating lesions. In next month's issue of Human Pathology, we will present part II of this 2-part review that describes various malignant mesenchymal lesions of the bladder [1].

2. Myofibroblastic proliferations

2.1. Inflammatory myofibroblastic tumor

IMT of the bladder has had many designations, including inflammatory pseudotumor, inflammatory pseudosarcomatous fibromyxoid tumor, nodular fasciitis, pseudosarcomatous myofibroblastic tumor, and fibromyxoid pseudotumor [2-20]. Initially reported by Roth in a 32-year-old woman, the lesion was described as being composed of spindle cells in a myxoid stroma, with scattered chronic inflammatory cells [21]. Typical mitotic figures were present, and the lesion infiltrated the muscle. There was no recurrence after resection (Figs. 1 and 2) [21].

The most frequent presenting symptom is hematuria; other symptoms include irritative and/or obstructive voiding symptoms, abdominal pain, or the discovery of a mass lesion. Rarely, constitutional symptoms including fever and weight loss have been reported, possibly due to the release of cytokines [18]. Grossly, the lesion is either a polypoid mass or a submucosal nodule. The tumor may or may not be associated with surface ulceration. The cut surface is often pale, firm, and gelatinous. Microscopically, the tumor is classically described as a spindle cell proliferation with elongated, eosinophilic cytoplasmic processes in a loose and edematous, or myxoid,

Fig. 2 IMT of the bladder. A, Low-power view of hypocellular variant is shown. B, High-power view of same tumor seen in A. C, Hypercellular variant of IMT is shown. D, Myxoid variant of IMT is seen extending into muscle. E, IMT with arrows identifying mitotic figures. F, Another interesting myxoid variant of IMT is shown. G, IMT extending into muscle is shown. H, End stage of sclerotic/fibrotic variant of IMT is shown. I, Positive ALK staining in an IMT is shown.