



Original contribution

Pseudosarcomatous and sarcomatous proliferations of the bladder

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Received 26 June 2006; revised 7 November 2006; accepted 9 November 2006

Keywords:

Pseudosarcomatous
fibromyxoid tumor;
Postoperative spindle cell
nodule;
Sarcoma;
Sarcomatoid carcinoma;
Bladder

Summary Pseudosarcomatous fibromyxoid tumor (PFT), postoperative spindle cell nodule (PSN), sarcoma, and sarcomatoid carcinoma of the bladder are frequently difficult to distinguish histopathologically with significant differences in disease-related outcomes. A retrospective review of our pathology registry over the last 25 years identified 7 PFT, 10 PSN, 18 primary bladder sarcomas, and 17 sarcomatoid carcinomas. Most patients with PFT, PSN, sarcoma, and sarcomatoid carcinoma were diagnosed between the ages of 50 to 60 years with PFT and PSN most commonly detected in women. A previous history of urological instrumentation and bladder cancer was present in all patients with PSN but none of the patients with PFT. Pseudosarcomatous fibromyxoid tumors were characterized by a tissue culture-like proliferation of myofibroblastic cells with focal atypia and overall cytoarchitectural features mimicking nodular fasciitis. Sarcomas and sarcomatoid carcinomas exhibited cellular atypia, mitotic activity with atypical mitosis, and the presence of necrosis. Transurethral resection was sufficient to control all PFT and PSN with no evidence of distant metastatic spread. In contrast, local recurrences and distant metastases frequently occurred in patients with primary sarcoma and sarcomatoid carcinoma despite aggressive surgical management, which was often combined with neoadjuvant chemotherapy (50% and 65% disease-specific mortality, respectively). Pseudosarcomatous fibromyxoid tumor and PSN have unique clinical and pathologic features that allow their distinction from primary bladder sarcoma and sarcomatoid carcinoma.

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1. Introduction

Pseudosarcomatous spindle cell proliferations of the bladder comprise 2 groups of lesions with overlapping microscopic features, referred to as pseudosarcomatous fibromyxoid tumor (PFT) and postoperative spindle cell

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nodule (PSN). Both of these lesions represent florid proliferations of spindle cells that may be confused with sarcomas. These lesions have overlapping microscopic features but are believed to represent pathogenetically distinct lesions.

Pseudosarcomatous fibromyxoid tumor of the urinary bladder is a rare reactive proliferation of myofibroblasts [1]. This lesion was first described by Roth [2] as a pseudosarcomatous myofibroblastic tumor, and numerous designations have since been used to describe it, including inflammatory pseudotumor [3,4], nodular fasciitis [5], and pseudosarcomatous myofibroblastic proliferation [6]. Postoperative spindle cell nodule has a very similar gross and histologic appearance consisting of a reactive proliferation of spindle cells, which usually occurs several months following a lower genitourinary surgical procedure such as a transurethral resection (TUR) or a biopsy [7-10]. Despite these previous studies, it remains unclear whether these lesions are reactive or neoplastic in origin [11-13]. Both PFT and PSN of the urinary bladder portend an excellent prognosis, with some of these tumors exhibiting spontaneous regression [11,14-16]. However, these benign lesions are frequently composed of atypical spindle cells, making their potential misdiagnosis as a bladder sarcoma or sarcomatoid carcinoma an important concern for the pathologist and treating physician [6,16]. We previously described 2 cases of PFT that were initially diagnosed as bladder rhabdomyosarcoma [17].

The purpose of the present study was to define the clinical and pathologic features that distinguish these lesions to aid pathologists and treating physicians in establishing the correct diagnosis and provide appropriate management.

2. Materials and methods

2.1. Pathologic specimens

We searched our tumor registry over the last 25 years (January 1980 to January 2005) and identified 7 patients with PFT, 10 patients with PSN, 18 patients with primary sarcoma, and 17 patients with sarcomatoid carcinoma of the urinary bladder. The pathologic slides and pathology reports were reviewed for all cases. Cases of PFT and PSN treated at our institution and previously reported [15,17] were not included in this report. Pseudosarcomatous proliferations, primary bladder sarcomas, and sarcomatoid carcinomas were classified and graded, when appropriate, according to the World Health Organization Classification and Grading Systems (1999) [18]. The associated urothelial carcinomas were classified according to the World Health Organization/International Society of Urological Pathology Consensus Grading Scheme [19]. Of those patients with primary sarcoma, the sarcoma subtypes included leiomyosarcoma (n = 11), unclassified sarcoma (n = 4), and angiosarcoma (n = 3).

2.2. Clinical data

We reviewed patient medical records to determine their clinical presentation, treatment, and disease-related outcomes. The follow-up information for PFT and PSN was available for all cases, with median follow-ups of 3 years (8 months to 13.1 years) and 3.1 years (2 months to 5.6 years), respectively. The median lengths of follow-up for primary bladder sarcoma and sarcomatoid carcinoma were 2.6 years (2 months to 11.9 years) and 1.4 years (2 months to 5.6 years), respectively. These data were used to calculate disease-specific mortalities defined as the number of years between the date of diagnosis and the date of death from disease. All pathologic specimens and clinical data were analyzed under institutional review board–approved protocols. All patient identifiers were removed so that patient confidentiality could be maintained throughout the study.

3. Results

3.1. Microscopic features

3.1.1. Pseudosarcomatous fibromyxoid tumor

All 7 PFTs were characterized microscopically by proliferation of tissue culture–like spindled myofibroblastic cells within a myxoid stroma (Fig. 1). Scattered cells with enlarged atypical nuclei were nearly always present. The myofibroblastic cells were admixed with lymphocytic inflammatory cells. Typically, spindle cells were loosely arranged within the myxoid stroma. In some cases, a more compact arrangement of interlacing bundles mimicking the herringbone pattern, typically seen in fibrosarcoma, was present. A network of radiating capillary channels was prominent in peripheral parts of the lesion. Scattered mitoses (2-4 per 10 high-power fields) were present, but no atypical mitotic figures could be identified. In 3 PFT cases, paraffin blocks or unstained slides were available for immunohistochemical studies, with the spindle cells staining positive for smooth muscle actin. Scattered positive staining for cytokeratin was present within myofibroblastic cells in all 3 cases; however, some of the areas showed diffuse, strong positive staining for cytokeratin (AE1/AE3). Pseudosarcomatous fibromyxoid tumors were typically located within the submucosal stromal tissue. The involvement of the muscularis propria could be documented in all cases in which the biopsy material involved the deeper portion of the bladder wall.

3.1.2. Postoperative spindle cell nodule

Some degree of reactive spindle cell proliferation follows every biopsy or TUR of the bladder; however, PSN is distinct in that it consists of a florid spindle cell proliferation forming a cystoscopically identifiable lesion or constituting a major component of the surgical specimen. Typically, PSNs are identified on follow-up cystoscopy as nodular mucosal protrusions measuring 1 cm or less. Consistent with a prior

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