A Case of Inflammatory Myofibroblastic Tumor of the Urethra with Overactive Bladder

Phil Hyun Song, Hwa Su Lim, Mi Jin Kim, Hee Chang Jung

From the Department of Urology and 1Pathology, Yeungnam University Hospital, Daegu, Korea

Inflammatory myofibroblastic tumor (IMT) of the urethra is a tumor composed of myofibroblasts and a mixed inflammatory infiltrate that rarely undergoes malignant transformation. The etiology and the biologic behaviors of IMTs are still unknown. Extensive pathologic examination is important to prevent misdiagnosis and the need for long-term follow up is emphasized. Recently, we experienced a case of an inflammatory myofibroblastic tumor of the urethra in a young female. To our knowledge, this is the first documentation of such an entity in published reports. So we present our experience with a review of literature. (J Korean Continence Soc 2009;13:80-82)

Key Words: Urethra, Inflammatory myofibroblastic tumor

Inflammatory myofibroblastic tumor (IMT) is a benign lesion of proliferating myofibroblasts with background inflammatory cell and hemangiofibrous tissue of unknown origin. IMT with a characteristic fibroinflammatory appearance, has been described in most organs and anatomic sites with few exceptions (1). IMT is a rare tumor and difficult to be distinguished from malignant neoplasm. Also preoperative diagnosis is difficult because of a large amount of specimen is needed for precise pathologic diagnosis. IMT is a tumor of low malignant potential that has limited clinical behavior in most cases but has the potential for aggressive local recurrence and even metastatic spread in a minority of cases (2).

Recently, we experienced a case of IMT of the urethra in a young female. So we present our experience with a review of literature. To our knowledge, this is the first documentation of such an entity in published reports.

CASE REPORT

A 24-yr-old woman was presented with a five-month history of abnormal protruding mass through urethral orifice.

On admission, she had intermittent painless hematuria, frequency and urgency. She had no specific past history except hyperthyroidism, which she had been taking a propylthiouracil daily for the past 1 year.

Physical findings were normal except for about 2×1cm sized reddish protruding mass at urethral opening. (Figure 1) Routine laboratory data, chest radiograph, intravenous
A Case of Inflammatory Myofibroblastic Tumor of the Urethra with Overactive Bladder

Figure 1. Preoperative photograph shows about 2×1cm sized, reddish protruding mass of urethral opening.

Figure 2. The tumor is composed of fascicles of spindle cells. Numerous lymphocytes and plasma cells are intermixed with tumor cells (H&E stain, ×200).

Figure 3. A. The tumor cells are strongly and diffusely positive for smooth muscle actin. B. Those are negative for desmin (Actin and Desmin stain, ×200).

pyelography (IVP) and cystoscopic findings showed no specific abnormality. So, we performed surgical resection under the impression of urethral prolapse or urethral benign tumor.

Grossly, the tumor was about 2×1cm sized, relatively well circumscribed and yellow-tan colored in appearance. Histologically, the tumor was composed of fascicles of spindle cells and numerous lymphocytes and plasma cells were intermixed with the tumor cells. (Figure 2) The tumor cells were strongly and diffusely positive for smooth muscle actin. (Figure 3A) Those were negative for desmin. (Figure 3B) Based on the above findings, the pathological diagnosis was inflammatory myofibroblastic tumor of urethra.

There was no abnormal finding on computed tomographic scan (CT) of abdomen at 1 month after surgery. We have been performing a follow-up check and there was neither intermittent hematuria nor voiding difficulty.

DISCUSSION

IMT with characteristic fibroinflammatory appearance, has now been described in most organs and anatomic sites with few exceptions (1).

Since IMT was first reported by Symmers on 1921, IMT has been found at various organs (3). Although the lung was the best known and most common site, it has subsequently been recognized involving a wide variety of anatomic sites. In the genitourinary tract IMT has been reported in the kidney, urethra, prostate, ureter and rete testis but is most frequently observed in the bladder (4).

While pulmonary IMT occurs more commonly in mid-adulthood, extrapulmonary IMT occurs more often in the soft tissues and viscera for children and young adults (5).

The clinical features of IMT were unexplained fever, weight loss, iron deficiency anemia, thromboctysis and
elevated erythrocyte sedimentation rate accompanying site-specific symptoms (5). However, such constitutional symptoms and serological findings are not common in cases of IMT of the genitourinary tract (6). The most common symptoms is painless hematuria. Less often patients present with dysuria, pelvic pain, or symptoms of urinary tract obstructions or infection (6).

Although there were reports of postsurgical, post-traumatic, and postinfectious (especially Ebstein-Barr virus) cases, the cause and pathogenesis of the IMT are unknown for the most part (7). Recently there are reports of constitutional systemic symptoms in a minority of cases that suggest a role for cytokines, especially interleukin-6, in the pathogenesis (8). And a provocative finding is the implication that human herpes virus 8 DNA may have some role in the cause of IMTs (9).

The microscopic features of the IMT are a myxoid, spindle cell dominant and less compact appearance (1). In all of the lesions, the mixed inflammatory infiltration was a distinctive microscopic feature. The spindle cells were reactive with antibodies to vimentin, smooth muscle actin, and muscle-specific actin in a majority of cases (5).

Three basic histological patterns are recognized, which are myxoid/vascular, compact spindle cells, and hypocellular fibrous patterns (1). The three patterns might be equally represented with one blending into another, or one or two patterns might predominate. And none of which had any discernible relationship to prognosis.

The most popular therapy for IMT is complete surgical resection. And radiation therapy and/or chemotherapy have been reported to have benefit in the treatment of incompletely resected or locally invasive IMTs (2).

Conclusively most IMTs are successfully managed by surgical resection. But the IMT may have locally aggressive growth, the capacity for vascular invasion, and may undergo malignant transformation in a small number of cases (2). So the characteristics of IMT were as obscure.

Most reported cases were benign, nonmetastasizing lesions. However, based on some cases of recurrence, local invasion and metastasis, For practical purposes IMT of the genitourinary tract should be considered a neoplasm of uncertain malignant potential, and routine surveillance and close clinical followup are recommended.

REFERENCES