Case Report

Urologia Internationalis

Urol Int 2000;65:57–59

Received: November 26, 1999
Accepted: March 7, 2000

Recurrent Aggressive Angiomyxoma of the Urinary Bladder

Case Report and Review of the Literature

F. May[a] A. Luther[a] W. Mohr[b] R. Bachor[a] R.E. Hautmann[a]

Departments of [a]Urology and [b]Pathology, University of Ulm, Germany

Key Words
Angiomyxoma aggressive · Urinary bladder · Pathology

Abstract
Aggressive angiomyxoma is a rare neoplasm which predominantly involves the female pelvis and perineum. Forty-four cases have been reported in the world literature, including 10 cases in men. To the best of our knowledge, the first case of recurrent aggressive angiomyxoma of the urinary bladder is presented here. Operative management, radiologic features and pathological findings are discussed.

Case Report

In January 1991, a 38-year-old man presented with a large intrapelvic painless mass. Surgical exploration in May 1987 and in June 1990 had revealed a large myxoid tumor of the posterior wall of the bladder. Wide resection with tumor-free margins was performed in conjunction with partial bladder resection. The histological diagnosis was recurrent aggressive angiomyxoma.

Upon presentation in January 1991, excretory urography (IVP) showed a big intrapelvic mass displacing the bladder to the right (fig. 1). Sonography and computed tomography revealed a large paravesical cystic tumor (fig. 2). Staging procedures including ultrasound of the liver, chest radiography and bone scan were found to be normal. The patient underwent radical cystoprostatectomy with ileal neobladder urinary diversion. The intraoperative situs revealed a large paravesical gelatinous tumor of homogeneous consistency originating from the left bladder wall. In the long-term follow-up of 9 years, the patient has shown no evidence of disease up to date.

Pathological Findings

The specimen consisted of the urinary bladder with an adherent greyish-white tumor tissue containing cystic spaces filled with gelatinous material (fig. 3a). The diameter was approximately 15 × 12 × 10 cm. The tumor was composed of variable-sized blood vessels with a surrounding loose tissue of preferentially spindle-shaped cells (fig. 3b–d). Neither the spindle-shaped cells nor endothelial cells of the blood vessels exhibited nuclear atypia and mitotic activity. Some areas consisted of foamy tumor cells with slightly alcianophilic material in the cytoplasm (fig. 3e), in other areas cystic spaces were present also filled with slightly alcianophilic material (fig. 3f, g). Lymph nodes were negative for metastasis and surgical resection margins were not involved. The histological diagnosis was recurrent angiomyxoma.

KARGER

Fax + 41 61 306 12 34
E-Mail karger@karger.ch
www.karger.com

0042–1138/00/0651–0057$17.50/0

Accessible online at:
www.karger.com/journals/uin

Dr. med. F. May
Department of Urology, Klinikum rechts der Isar, Technische Universität
Ismaningerstrasse 22
D–81675 Munich (Germany)
Tel. +49 89 4140 2521, Fax +49 89 4140 4843, E-Mail tb811aa@mail.lrz-muenchen.de

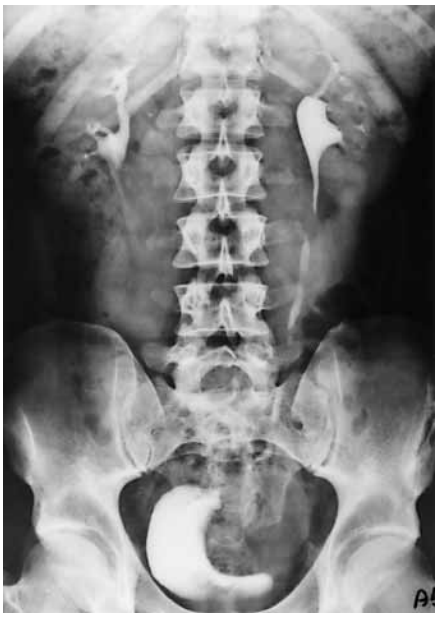

Fig. 1. Excretory urography showing a big mass displacing the bladder to the right.

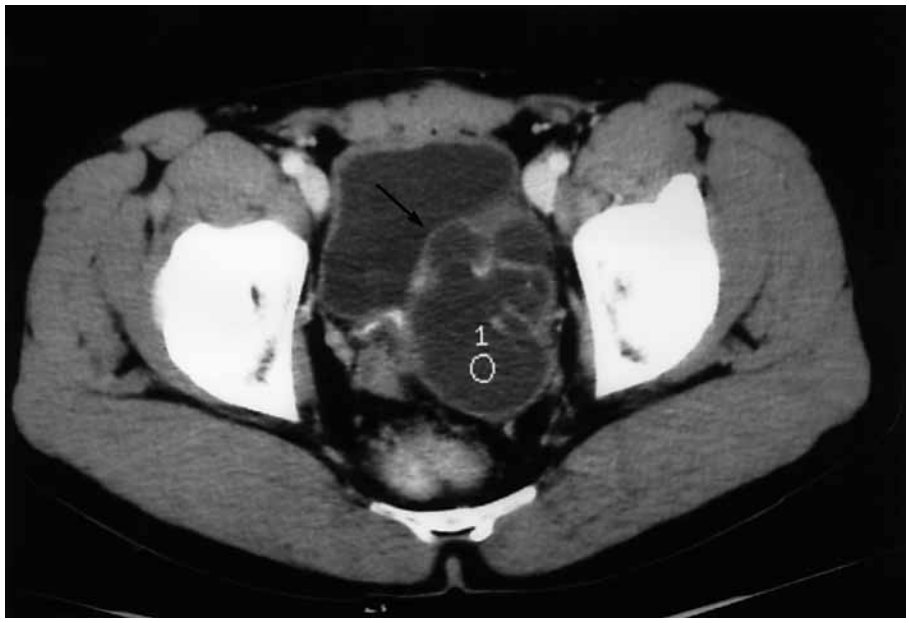

Fig. 2. CT revealing a big intrapelvic cystic tumor (numbers) without contrast medium uptake in most areas.

Discussion

Originally described in 1983 as a distinct tumor derived from myofibroblasts [1], these neoplasms tend to be firm, poorly encapsulated myxoid masses that locally infiltrate the soft tissues of the pelvis and perineum. Characteristic histological features include loose myxoid stroma and prominent vascularity. In contrast to malignant tumors with myxoid components, the nuclei have neither atypical features nor mitotic activity. A close resemblance between the fibroblasts of the pelvic soft parts and those of aggressive angiomyxoma both at the histological and the ultrastructural level was found [2] providing additional support for a pathogenic relationship with the pelvic soft tissue compartment. Treatment typically consists of local excision with tumor-free margins. About 70% of the patients develop local recurrences. While by some authors recurrences have been attributed to inadequate excision, even wide resection with tumor-free margins did not prevent local recurrence [3, 4]. No deaths or distant metastases have been reported.

In conclusion, aggressive angiomyxoma should be considered in the differential diagnosis of genitourinary tumors. The second local recurrence of a locally advanced neoplasm made us choose a radical surgical approach resulting in a 9-year interval without evidence of disease up to date. The value of CT scan and ultrasound to identify any invasion into adjacent organs has been emphasized [5]. Additional studies including excretory urography, bone scan, or barium enema may provide further information regarding the extent of disease. Reviewing the literature, a careful primary surgical extirpation, as complete as technically feasible, should be the main primary surgical approach. Regarding the high local recurrence rate, wide resection of local recurrences or radical pelvic surgery in selected patients is necessary. Long-term careful postoperative monitoring for recurrent tumors is, of course, mandatory.

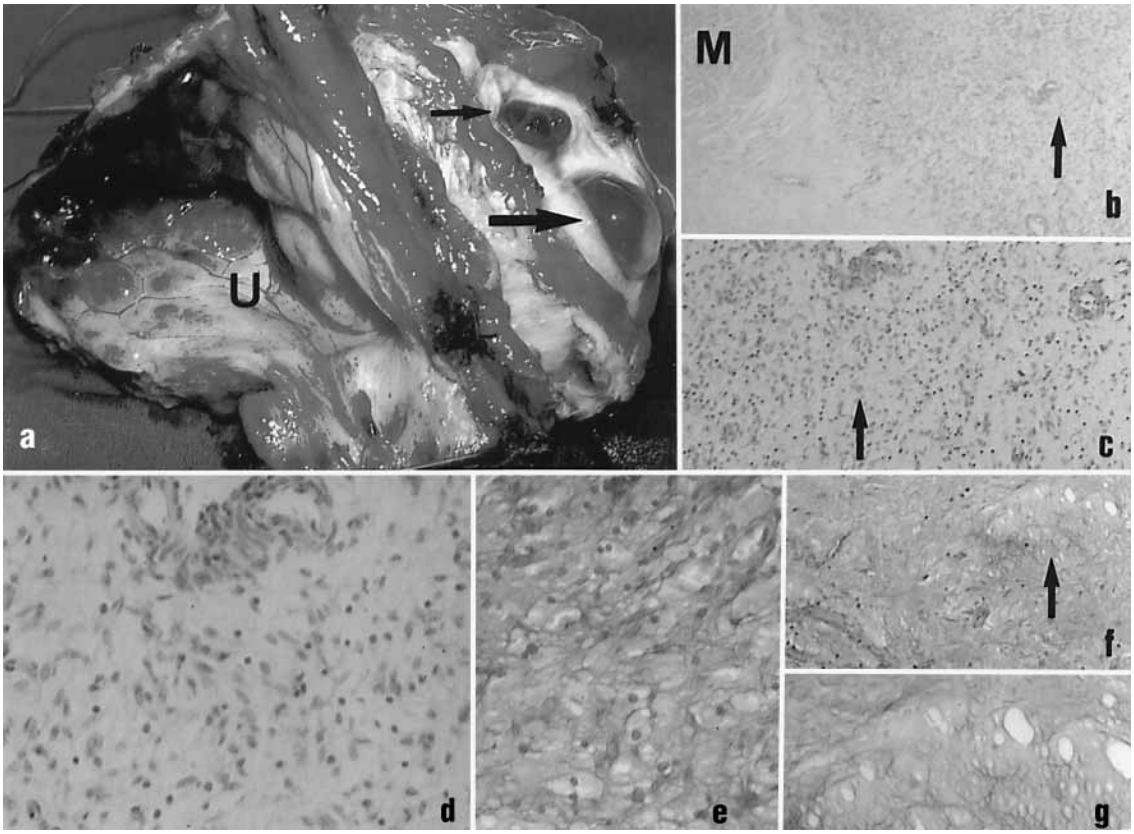

Fig. 3. a Operative specimen: Adjacent to the urinary bladder (U) a greyish-white tumor tissue with cystic spaces filled with gelatinous material (arrows). **b** Adjacent to the muscles of the urinary bladder (M) tumor tissue with prominent vessels. HE. × 35. **c** Higher magnification of the area marked with arrow in **b**: Tumor tissue of medium cellularity with prominent vessels. HE. × 85. **d** Higher magnification of the area marked with arrows in **c**: Blood vessels surrounded by tumor tissue of spindle-shaped loosely arranged cells and fibrillar intercellular matrix. HE. × 220. **e** Tumor tissue consisting of blood vessels and tumor cells with slightly alcianophilic cytoplasm. HE. × 220. **f** Small cystic spaces filled with foamy slightly alcianophilic material. Alcian blue and PAS stain. × 35. **g** Higher magnification of the area marked with arrow in **f**: Small cystic spaces filled with slightly alcianophilic foamy material. Alcian blue and PAS stain. × 85.

References

1 Steeper TA, Rosai J: Aggressive angiomyxoma of the female pelvis and perineum: Report of nine cases of a distinctive type of gynecologic soft-tissue neoplasm. Am J Surg Pathol 1983; 7(5):463–475.

2 Begin LR, Clement PB, Kirk ME, Jothy S, Caughey WTE, Ferenczy A: Aggressive angiomyxoma of pelvic soft parts: A clinicopathologic study of nine cases. Hum Pathol 1985; 16(6):621–628.

3 Stephenson BM, Williams EV, Sturdy DE, Vellacott KD: Aggressive angiomyxoma of the perineum and pelvis. Br J Surg 1992;79:1181.

4 Smith HO, Worrell RV, Smith AY, Dorin MH, Rosenberg RD, Bartow SA: Case report. Aggressive angiomyxoma of the female pelvis and perineum: Review of the literature. Gynecol Oncol 1991;42:79–85.

5 Llauger J, Perez C, Coscojuela P, Palmer J, Puig J: Aggressive angiomyxoma of pelvic soft tissue: CT appearance. Urol Radiol 1990;12: 25–26.