



Case Report

Retroperitoneal myofibroblastoma in an 88-year-old male

Shogo Watari,¹  Takaharu Ichikawa,¹  Hiromasa Shiraishi,¹ Takafumi Sakuma,¹ Risa Kubota,¹ Norihiro Kusumi,¹ Tomoyasu Tsushima,¹ Keina Nagakita² and Yoko Shinno²

Department of ¹Urology and ²Pathology, National Hospital Organization Okayama Medical Center, Okayama, Japan

Abbreviations & Acronyms

ALK = anaplastic lymphoma kinase

RB = retinoblastoma

STAT6 = signal transducer and activator of transcription

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Correspondence: Takaharu Ichikawa, M.D., Ph.D., Department of Urology, National Hospital Organization Okayama Medical Center, 1711-1 Tamasu, Kita-ku, Okayama 701-1192, Japan.
Email: takaichiaug02@yahoo.co.jp

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Introduction: Extramammary myofibroblastomas are extremely rare.

Case presentation: The patient was an 88-year-old male. He presented for evaluation of frequent urination and a “pushing up” sensation from the groin during defecation. Thorough physical and radiographic examinations revealed a retroperitoneal tumor on the right side of the rectum. The pathologic examination of the biopsy tissue showed that the tumor was unlikely to be malignant. Nevertheless, the patient was symptomatic and thus underwent a laparoscopic tumor resection through a transperitoneal approach. The tumor was circumscribed with a solid capsule. Based on the pathologic findings, which included immunostaining, the tumor was diagnosed as a myofibroblastoma. There was no evidence of a recurrence 6 months postoperatively.

Conclusion: We present this case with the clinical course and surgical findings, and discuss the possibility of establishing a preoperative pathologic diagnosis of a myofibroblastoma.

Key words: laparoscopy, myofibroblastoma, pathology, retroperitoneal neoplasms, surgical procedure.

Keynote message

Extramammary myofibroblastomas are extremely rare. The preoperative diagnosis of a myofibroblastoma is challenging; however, the loss of nuclear retinoblastoma 1 expression may serve as a diagnostic clue. Myofibroblastomas have clear demarcation lines and are easily resected along the capsule.

Introduction

Myofibroblastomas are rare, benign tumors that often occur in the breasts of postmenopausal females or elderly males.¹ Myofibroblastomas occurring in the breast, also known as mammary fibroblastomas, were first reported in 1987.² It is extremely rare for myofibroblastomas to occur in extramammary sites. We report a case involving an extramammary myofibroblastoma that developed in the retroperitoneum of an elderly male.

Case report

An 88-year-old male presented to our institution for evaluation of frequent urination and a “pushing up” sensation from the groin during defecation. He had no clinical or family history of malignancies. A digital rectal examination was significant for a hard and painless mass compressing the rectum from the right side. Computed tomography demonstrated a retroperitoneal tumor on the right side of the rectum, 13.7 × 6.4 × 12 cm in size, dorsal to the bladder. The differential diagnosis included a leiomyoma, leiomyosarcoma, or solitary fibrous tumor based on magnetic resonance imaging (Fig. 1).

Seventeen days after the initial visit a needle biopsy of the retroperitoneal tumor was performed under trans-abdominal ultrasound guidance. Hematoxylin and eosin staining showed that the biopsy cores were mainly composed of fibroblasts and capillaries without nuclear atypia or increased cell density (Fig. 2). Based on a pathologic examination of the biopsy specimen, the tumor was unlikely to be malignant. Nevertheless, tumor resection was performed because

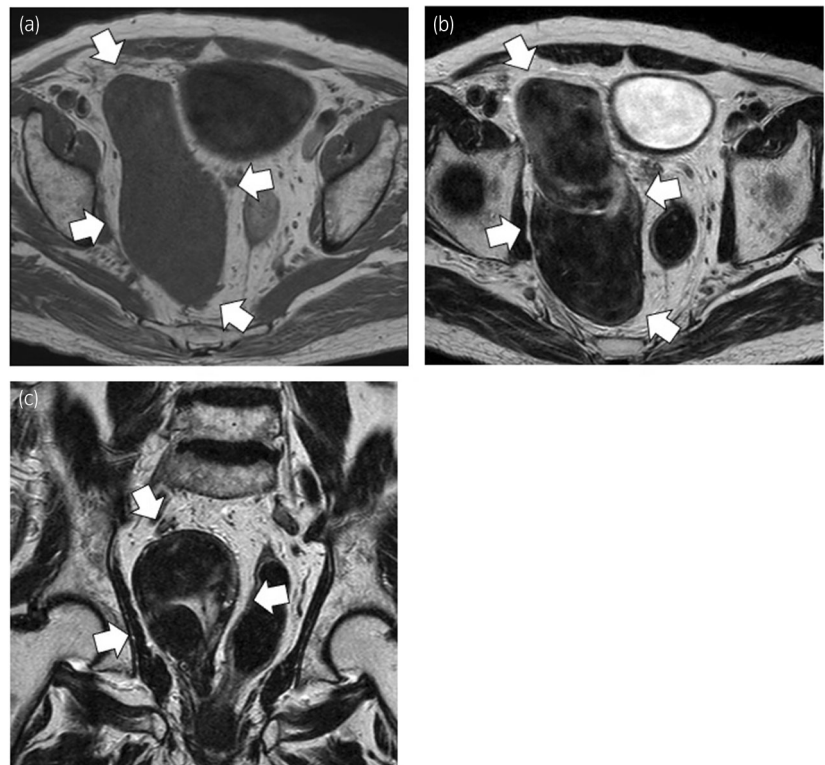


Fig. 1 Magnetic resonance image of the retroperitoneal tumor. (a) T1-weighted sequence, axial. (b) T2-weighted sequence, axial. (c) T2-weighted sequence, coronal. Arrows indicate the tumor.

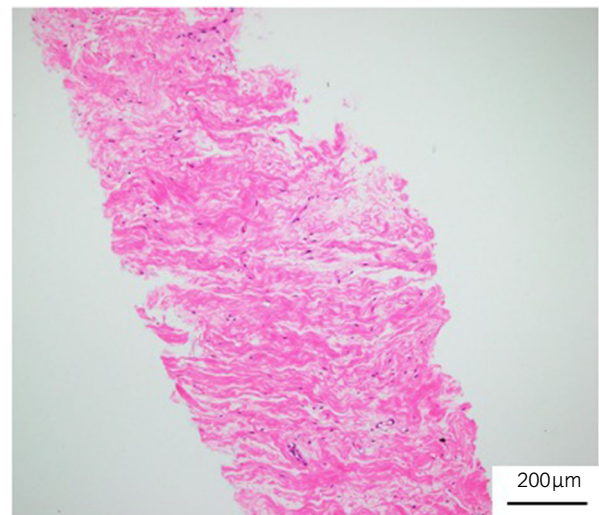


Fig. 2 The pathologic findings of the needle biopsy core. Magnification, $\times 100$. Hematoxylin and eosin staining.

the patient was symptomatic. Laparoscopic surgery was performed because a shorter and easier postoperative recovery period was anticipated in our elderly patient than with open surgery. We used an intraperitoneal approach after the insertion of a right ureteral catheter to prevent a ureteral injury. The locations of the laparoscopic ports are shown in Figure 3a. It was a challenge to resect the retroperitoneal tumor with clear surgical margins; however, the tumor was circumscribed with a solid capsule (Fig. 3b). After complete resection of the tumor along the capsule, the tumor was removed from the lower abdominal wound. The total operative time was 487 min. The

estimated blood loss was 1500 mL, thus requiring a 4-unit red blood cell transfusion. There were no other complications. Figure 3c,d show the gross appearance of the tumor.

The pathologic findings of the tumor showed an increase in spindle cells with mild atypia displaying a fascicular pattern with collagen and scattered mast cells (Fig. 4a,b). Based on immunostaining, we diagnosed the tumor as a myofibroblastoma (Fig. 4c–h). The patient's symptoms resolved postoperatively. Moreover, there was no evidence of a local recurrence or distant metastases 6 months postoperatively.

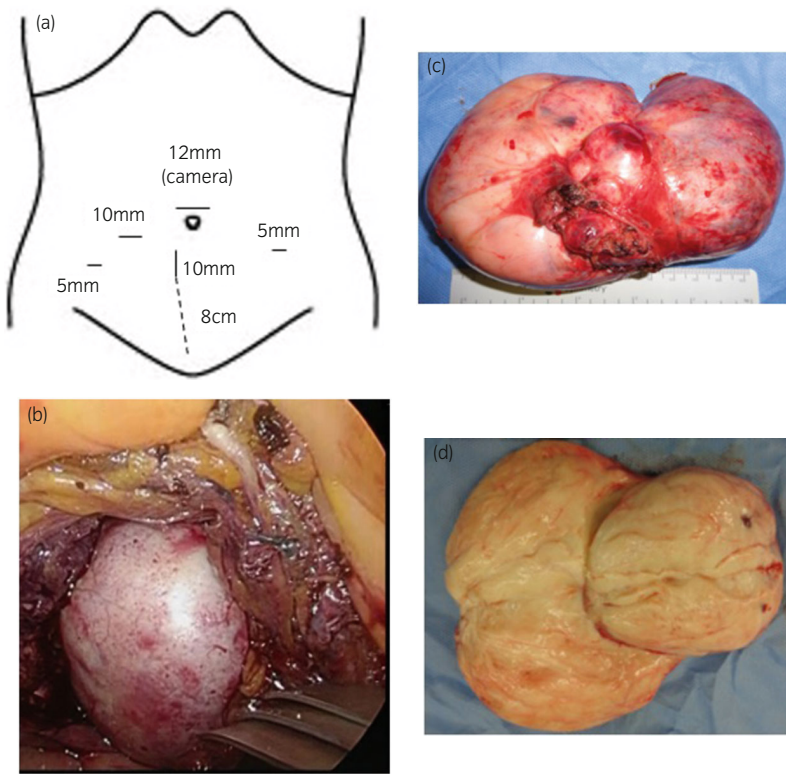


Fig. 3 (a) Location of laparoscopic ports and wound of the removed tumor. The break line represents the wound where the tumor was removed. (b) Endoscopic appearance of the tumor during resection. The tumor was milk-white in color, smooth, and elastic, but hard. (c) Macroscopic image of the tumor. The tumor surface was covered with a capsule. (d) Bisected tumor.

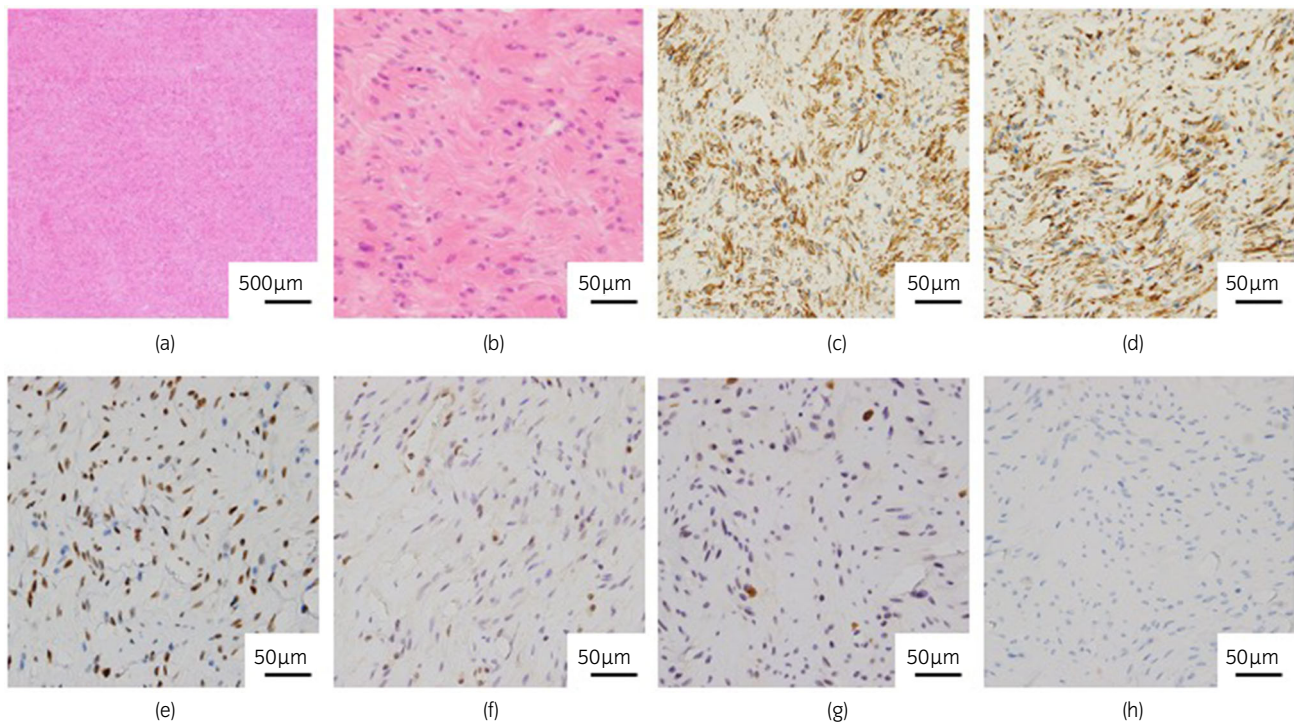


Fig. 4 The pathologic findings of the resected retroperitoneal tumor. (a) Hematoxylin and eosin staining. (b) Hematoxylin and eosin staining. (c) Positive for CD34. (d) Positive for desmin. (e) Positive for estrogen receptor. (f) Loss of nuclear RB1. (g) Negative for anti-STAT6. (h) Negative for ALK. (a) magnification, $\times 40$; (b–h) magnification, $\times 400$.

Discussion

An extramammary myofibroblastoma was first reported in 2001.³ According to a report that summarized the pathologic features of myofibroblastomas, the incidence of abdominal and retroperitoneal lesions was 10% (14 of 143 cases).⁴ To date, five cases of extramammary myofibroblastomas in the pelvic retroperitoneal space have been reported.^{5–8} The present report is of clinical value because there have been few reports that have included details of the clinical course and surgical findings of extramammary myofibroblastomas.

The features reported for mammary myofibroblastomas are as follows: solitary, painless, elastic-to-hard, and covered with a capsule.⁹ In the present case, the extramammary myofibroblastoma, which was located in the retroperitoneum, had similar features. We were able to resect the myofibroblastoma along the capsule of tumor. Most of the previous case reports involving myofibroblastomas occurring in the pelvic retroperitoneum have not included a description of the surgical approach.^{5–7} It has been reported that a prostate myofibroblastoma was resected via a retropubic radical prostatectomy with resection of the anterior pubic arch and reconstruction of the urethra.⁸ In the present case, we attempted laparoscopic surgery of the tumor to achieve a shorter and easier postoperative recovery period. In addition, we did not need to prepare for urinary tract reconstruction. The pelvic region is an area which urologists are very familiar with, and thus we predicted that laparoscopic surgery could be successful.

In previous case reports involving mammary myofibroblastomas, magnetic resonance imaging showed iso-signals in both T1- and T2-weighted images.^{10–13} Moreover, it has been reported that myofibroblastomas have clear demarcation lines at the periphery.^{10–13} In the present case, the demarcation around the tumor was also clear. Indeed, these imaging findings are not specific for myofibroblastomas, thus establishing a diagnosis for myofibroblastomas based on imaging is difficult.

The pathologic findings of a myofibroblastoma are generally reported as a well-circumscribed mass consisting of an increased number of spindle cells with mild atypia.⁴ The spindle cells are arranged in a fascicular pattern, accompanied by collagenous stroma and scattered mast cells. Mitotic figures are not evident. Based on immunostaining, there is diffuse expression of CD34, desmin, and estrogen receptor,^{4,14} in addition to a loss of nuclear RB1 expression in myofibroblastomas. The histologic findings and immunohistochemical staining pattern in our case were also in agreement with previous reports. Furthermore, the anti-STAT6 antibody and ALK-negativity ruled out solitary fibrous and inflammatory myofibroblastic tumors, respectively. Of note, loss of nuclear RB1 has been reported to occur in 90% of myofibroblastomas.⁴ Loss of RB1 is a potential strategy for diagnosing a myofibroblastoma based on a biopsy specimen. An accurate preoperative diagnosis of a myofibroblastoma allows the surgeon to select the optimal procedure for resecting the tumor along the capsule.

Myofibroblastomas have been reported to slowly increase in size over time¹⁵; however, the long-term natural history of

myofibroblastomas is unknown because all reported cases were surgically resected. The prognosis for myofibroblastomas appears to be favorable; specifically, myofibroblastomas are benign tumors that are curable with surgical resection. No recurrences were reported among 13 patients in whom tumors were resected without residual disease after 15 years of follow-up evaluations.¹⁶ Moreover, no recurrences were reported in 8 patients, despite specimens with positive surgical margins.⁴ There have been 2 recurrences among 143 myofibroblastomas⁴; specifically, local recurrences occurred 17 and 20 years after resection.⁴ Thus, myofibroblastomas have a low risk of recurrence, but late recurrences are possible.

Conclusion

We have reported a retroperitoneal myofibroblastoma in an elderly male. The loss of nuclear RB1 expression suggests the diagnosis of a myofibroblastoma preoperatively. Our experience may help determine the optimal surgical strategy for resection of a myofibroblastoma along the capsule.

Author contributions

Shogo Watari: Writing – original draft. Takaharu Ichikawa: Supervision; writing – review and editing. Hiromasa Shiraiishi: Writing – review and editing. Takafumi Sakuma: Writing – review and editing. Risa Kubota: Writing – review and editing. Norihiro Kusumi: Writing – review and editing. Tomoyasu Tsushima: Writing – review and editing. Keina Nagakita: Writing – review and editing. Yoko Shinno: Writing – review and editing.

Conflict of interest

The authors declare no conflict of interest.

Approval of the research protocol by an Institutional Reviewer Board

N/A.

Informed consent

Written informed consent was obtained.

Registry and the Registration No. of the study/trial

N/A.

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Editorial Comment

Editorial Comment to Retroperitoneal myofibroblastoma in an 88-year-old male

Extramammary myofibroblastomas are rare benign tumors. Since the first report by McMenamin *et al.* in 2001,¹ approximately 150 cases of extramammary myofibroblastoma have been published.² Of note, a series by Howitt *et al.* comprised the largest proportion of the published cases of extramammary myofibroblastoma to date.³ The anatomic distribution of these tumors is widespread, with the most common, general site of involvement being the groin/inguinal region.³ Therefore, it has been theorized that extramammary myofibroblastomas occur along the embryonic milk-line from the axilla to the medial groin.² Differential diagnosis for extramammary myofibroblastoma includes both benign and malignant tumors.² Although computed tomography and magnetic resonance imaging are considered valuable in the diagnosis of myofibroblastoma,⁴ extramammary fibroblastoma is difficult to diagnose. Thus, the most appropriate diagnostic test for a clinically or radiologically evolving soft tissue mass is a biopsy.² Treatment strategy should be decided based on pathological diagnosis of biopsy specimen as well as preoperative imaging. In fact, if a malignant tumor is identified from biopsy, a combined resection of adjacent organs should be considered.

Watari *et al.* reported a case of retroperitoneal myofibroblastoma in an 88-year-old male.⁵ Since there have been a few cases of extramammary myofibroblastoma in the pelvic retroperitoneal space, this case was extremely rare in terms of describing detailed surgical treatment. Although extramammary myofibroblastomas are frequently asymptomatic and detected incidentally, large retroperitoneal myofibroblastomas

may be accompanied by symptoms such as dysuria and malaise during defecation.^{2,4,5} In this case, the patient underwent surgical resection due to frequent urination and a “pushing up” sensation from the groin during defecation.⁵ Successful treatment of the large retroperitoneal myofibroblastoma was achieved using laparoscopic radical surgery.⁵ Thus, this report is of clinical value because few reports described the details of clinical course and surgical findings of extramammary myofibroblastomas. Obviously, the selection of open or laparoscopic procedure for tumors should be based on the individual case and the skill of surgeon. Therefore, when faced with a retroperitoneal soft tissue mass, which is detected incidentally or as a slow growing tumor, it is necessary to obtain a biopsy to rule out malignancy. If malignancy cannot be ruled out or the patient is symptomatic, surgical resection is appropriate.

Kyotaro Fukuta M.D. , Kei Daizumoto M.D., Ph.D. and Masayuki Takahashi M.D., Ph.D.
Department of Urology, Tokushima University Graduate School of Biomedical Sciences, Tokushima, Japan
takahashi.masayuki@tokushima-u.ac.jp

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Conflict of interest

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