Huge Primary Intermediate-grade Myofibroblastic Sarcoma in the Retroperitoneum

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ABSTRACT

Myofibroblastic sarcoma is exceedingly rare, with low-grade features in most cases, and rarely involves the retroperitoneum. The 2020 World Health Organization (WHO) classification of soft tissue tumours still lists only low-grade myofibroblastic sarcoma and shows no consensus on the definitions of high- and intermediate-grade myofibroblastic sarcomas, in contrast to the 2013 WHO classification. Surgical resection of the tumour and adjacent structures is the standard of care for most patients, and intermediate- and high-grade myofibroblastic sarcomas have very poor survival. We describe a patient with intermediate--grade myofibroblastic sarcoma in the retroperitoneum, who underwent *en bloc* resection and ureteroplasty without adjuvant therapy and was free of pain and any other discomfort during 19 months of follow-up.

Key Words: *Myofibroblastic sarcoma, Intermediate grade, Retroperitoneum, Surgery*.

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INTRODUCTION

Myofibroblastic sarcoma is a rarely observed malignant soft tissue tumour with low-grade features in most cases and occurs mainly in the head and neck, followed by the limbs and the trunk,[1] and rarely involves the retroperitoneum.[2] The 2020 World Health Organization (WHO) classification of soft tissue tumours still lists only low-grade myofibroblastic sarcoma and shows no consensus on the definitions of high- and intermediate-grade myofibroblastic sarcomas, in contrast to the 2013 WHO classification.[3] Surgical resection of the tumour and adjacent structures is the standard of care for most patients, and the therapeutic effects of adjuvant chemotherapy or radiation therapy are still unclear.[1] Intermediate- and high-grade myofibroblastic sarcomas have very poor survival, as reported in previous literature.

In this report, we describe a patient with intermediate-grade inflammatory myofibroblastic sarcoma who underwent *en bloc* resection and ureteroplasty without adjuvant therapy and completed 19 months of follow-up.

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CASE REPORT

A 57-year male was admitted with constant and dull pain in the lower left portion of his abdomen. The pain appeared one day prior to admission, with no obvious inducing factor. Physical examination revealed a huge palpable mass in the lower left abdominal quadrant with tenderness, an unclear boundary and slight mobility. The results of laboratory examinations, including tumour markers, routine blood tests, biochemical tests, and urine and stool tests, were within normal limits. No abnormalities were found on the electrocardiogram or chest computed tomography (CT). Abdominal ultrasonography, contrast CT and computed tomographic arteriography (CTA) of the abdomen revealed a large soft tissue mass in the abdominal and pelvic cavity measuring 18×13 cm, compression or infiltration of the left ureter, compression of the left external iliac blood vessels and a small amount of ascites.

Upon consultation with a multidisciplinary tumour board involving radiologists, urologists, vascular surgeons, anaesthesiologists and pathologists, it was decided that exploration and surgical resection of the tumour should be performed. On June 6, 2019, the patient underwent retroperitoneal tumour resection and ureteroplasty. A large mass with surface bulging was present in the peritoneal cavity, and 300 ml of pale yellow ascites was found in the hepatorenal recess, bilateral paracolic gutters and pelvic cavity. In the course of the operation, the left ureter was ligated and repaired because it was surrounded by the tumour and difficult to separate from the tumour. The left external iliac arteries surrounded by infiltrated tumour tissue were successfully separated.

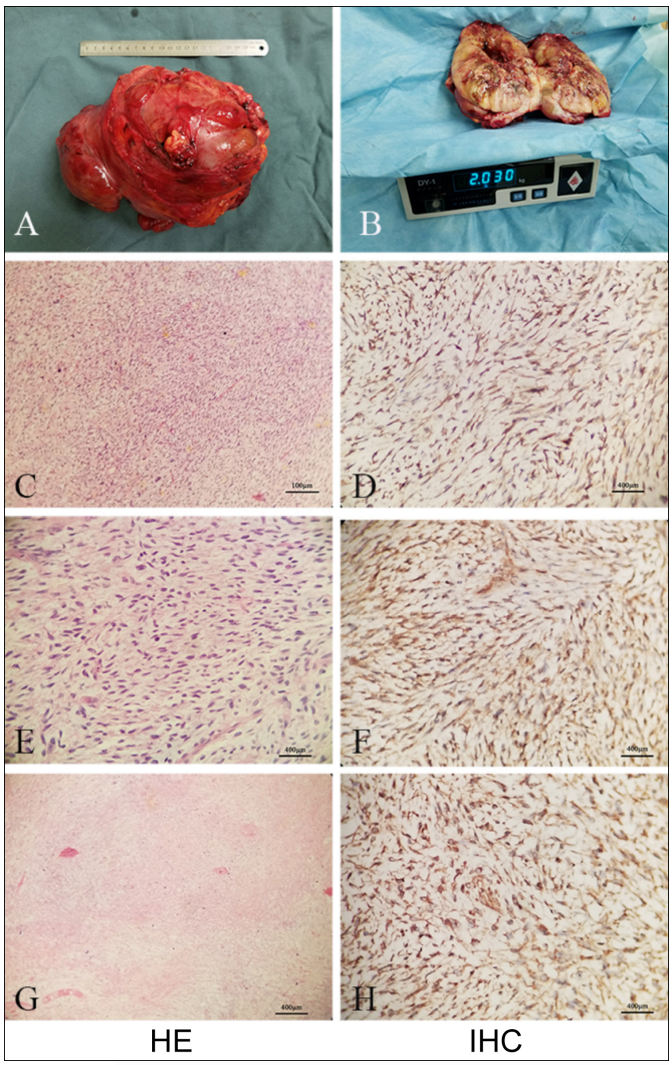

Figure 1: (A) **On macroscopic examination, the tumour's dimensions were 18×13 ×12 cm, and the surface was nodular. (B) The excised section showed greyish-yellow tissue with tan and dark red areas, and the texture was tender, sticky and firm; the tumour, weighed 2.03 kg, was translucent with a necrotic area. (C) A large number of spindle cells arranged in bundles (haematoxylin-eosin, ×100). (E) Some mitotic figures (haematoxylin-eosin, ×400). G: Some areas with necrosis (haematoxylin-eosin, ×400). Immunohistochemical images showing tumour positivity for vimentin (D), SMA (F), and desmin (H) (×400).**

On macroscopic examination, the tumour's dimensions were 18×13 ×12 cm (Figure 1A), and it weighed 2.03 kg. Sectioning revealed a yellow, lobulated tumour with a large, central necrotic area of 2×3 cm (Figure 1B). Histopathological examination revealed a large number of spindle cells that were arranged in bundles (Figure 1C) and contained necrosis in some areas (Figure 1G), small- to medium-sized nucleoli with readily identifiable mitotic figures, and <9 mitotic cells per 10 high-power fields (HPFs) (Figure 1E). Immunohistochemistry analysis showed that the tumour cells were positive for smooth muscle actin (SMA) (Figure 1F), desmin (Figure 1D), and vimentin (Figure 1H) and negative for HMB45, MDM2, S-100, ALK, h-caldesmon, Sox10, dog-1, STAT6, NF, CK, EMA, CD34, CD31, MyoD1, PR, Bcl-2, and CD117 and had a Ki-67 index of approximately 25%. On the basis of the histopathological and immunohistochemical features, the final diagnosis of intermediate-grade myofibroblastic sarcoma was established according to the French federation of cancer centres sarcoma group (FNCLCC) system.[4]

The patient had an uneventful recovery without adjuvant chemotherapy and was discharged on the 12th postoperative day. During the 19-month follow-up, the patient was free of pain and any other discomfort. Physical examination and abdominal CT showed no signs of recurrence or metastasis.

DISCUSSION

Myofibroblastic sarcoma is a rare malignant tumour with hybrid characteristics of smooth muscle cells and fibroblasts. Since Mentzel first reported it in 1998,[5] myofibroblastic sarcoma has been found to arise mainly in the head and neck regions. Involvement of other organs, including the trunk, bone, breast, liver, pancreas and adrenal gland, has been reported less frequently, and few cases have been reported in the retroperitoneum.[2] Because most reports comprise case reports or case series, the incidence rate is difficult to ascertain.

At present, there is no consensus on the definitions of high- and intermediate-grade myofibroblastic sarcomas, and the tumour grading classification generally depends on the three-tiered FNCLCC system.[4] The present case showed small necrotic areas (< 50%) and less than 9 mitoses per 10 HPFs and was considered to be intermediate grade according to the FNCLCC system. Compared with low-grade myofibroblastic sarcoma, intermediate- and high-grade myofibroblastic sarcomas have obvious pleomorphism, mitotic activity, local necrosis and significant invasion, with higher incidences of local recurrence and metastasis.

The tumour in this case was successfully removed by general surgeons and urological surgeons. This finding suggests that macroscopic complete resection improves the survival rate of patients with myofibroblastic sarcoma, and in retroperitoneal sarcoma, treatment should be carried out by a dedicated multidisciplinary team, from the diagnosis stage to the beginning of treatment, especially the first surgical treatment.[6] Given the limitations of the database, the therapeutic effects of adjuvant chemotherapy or radiation therapy are still unclear.[1] Due to the high possibility of distant metastasis, patients with grade 3 retroperitoneal sarcomas are good candidates for adjuvant chemotherapy. Surgery combined with radiotherapy and chemotherapy may be an effective method for the treatment of high-grade myofibroblastic sarcomas. However, the patient described herein refused further postoperative adjuvant chemotherapy or radiation therapy.

Geetha *et al*. reported a case of a high-grade retroperitoneal myofibroblastic sarcoma in the retroperitoneum in a 65-year male who underwent surgical excision of the tumour. Recurrence of the lesion occurred eight months after surgery, and the patient gradually deteriorated to death two months after the recurrence.[2] Given the risks of recurrence and metastasis, close

clinical follow-up is recommended. The present patient was doing well 19 months after the operation, with follow-up visits every 3 months for 2 years, every 6 months for the next 3 years, and every year after 5 postoperative years.

In summary, retroperitoneal myofibroblastic sarcoma of intermediate grade is extremely rare and may have a significantly lower degree of malignancy and lower incidence of recurrence and metastasis than that of high grade. Surgical resection with adjuvant radiation and chemotherapy may be the ideal choice of treatment, and close clinical follow-up is recommended.

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PATIENT'S CONSENT:
Patient has provided informed consent for the publication of the case.

COMPETING INTEREST:
All authors have contributed significantly to the manuscript and declare that the work is original and has not been submitted or published elsewhere. None of the authors has any financial disclosure or conflict of interest.

AUTHORS' CONTRIBUTION:
ZW: Supervised manuscript writing, reference selection, and final submission.
XM and YC: Performed the surgical operation.
QH: Histopathologic slides and details.
HH: Manuscript drafting and literature search.
All the authors have read and approved the final version of the manuscript.

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