Managing spindle cell sarcoma with surgery and high-intensity focused ultrasound: A case report

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Abstract
BACKGROUND
Undifferentiated pleomorphic sarcomas, also known as spindle cell sarcomas, are a relatively uncommon subtype of soft tissue sarcomas in clinical practice.

CASE SUMMARY
We present a case report of a 69-year-old female patient who was diagnosed with undifferentiated spindle cell soft tissue sarcoma on her left thigh. Surgical excision was initially performed, but the patient experienced a local recurrence following multiple surgeries and radioactive particle implantations. High-intensity focused ultrasound (HIFU) was subsequently administered, resulting in complete ablation of the sarcoma without any significant complications other than bone damage at the treated site. However, approximately four months later, the patient experienced a broken lesion at the original location. After further diagnostic workup, the patient underwent additional surgery and is currently stable with a good quality of life.

CONCLUSION
HIFU has shown positive outcomes in achieving local control of limb spindle cell sarcoma, making it an effective non-invasive treatment option.
INTRODUCTION

Limb spindle cell sarcomas are undifferentiated pleomorphic sarcomas, and they account for 12.18% of soft tissue sarcomas (STS) of the extremities\(^2\). STS in adults are a heterogeneous group of tumors of mesenchymal origin that share similar biological patterns of local tumor growth and metastasis. They can be carcinogenic or tumorigenic in their morphology\(^2\). This rare malignancy accounts for only up to 1% of all cancers\(^3\). Due to its rarity and complexity, there are few effective studies on its clinicopathological features and diagnosis. Here, we present a case involving a lower-extremity spindle cell sarcoma, detailing its treatment (including HIFU) and the evaluation of the treatment efficacy, in order to raise awareness about treatment options for this rarely detected tumor.

CASE PRESENTATION

Chief complaints

A 69-year-old female patient presented with a 3-cm-diameter firm mass that had gradually increased over the prior 6 years on the left thigh, with local pain.

History of present illness

Lumpectomy was performed at The Second Affiliated Hospital of Chongqing Medical University. Postoperative pathology results confirmed the mass to be a spindle cell soft tissue sarcoma. Postoperative immunohistochemistry results indicated CK(-), EMA(-), Vim(+), S100(-), SMA(±), Act(-), CD34(+), BCL-2(-), CD9(±), Ki-67(+), 50% AB(+), MBP(-), NF(-), and CD68(+), confirming the diagnosis of spindle cell soft tissue sarcoma (Figure 1A,B). The patient was treated with an expanded resection.

History of past illness

However, after 2 years, a firmer mass with some tenderness was found at the surgical site. Therefore, the patient underwent another expanded resection, followed by
radioactive particle implantation. Postoperative immunohistochemistry results indicated CK(-), EMA(±), DES(-), S100(-), SMA(-), CD34(+), SDX-10(-), CDK4(-), MDM2(-), CD68(-), CD99(±), BCL-2(+), Vim(+), and Ki-67(+) >50%.

**Personal and family history**

Nevertheless, after 16 mo, magnetic resonance imaging (MRI) revealed that the patient had relapsed. Subsequently, the patient underwent three lumpectomies and radioactive particle implantation.

**Physical examination**

Despite this, after 5 mo, the follow-up pathology results revealed another relapse. A new treatment plan was designed: five sessions of HIFU (which occurred on March 5, June 11, August 20, October 13, and November 24, 2021), using an IC (integrated circuit)-type HIFU tumor treatment system (Chongqing Haifu Medical Technology Co., Ltd., China), which mainly consists of an ultrasonic generator, a focused ultrasonic transducer, a motion system, a control system, and a B-ultra real-time guidance system. The vertical scanning mode with a slice thickness of 2 mm was used. The ultrasonic transmitter worked at frequencies of 0.85 and 1.5 MHz. The ultrasonic power was 150–238W. The duration of each treatment was 275–1325s. The focal length was 135 mm and the lesion had a diameter >5 cm.

**Laboratory examinations**

The ablation effect was assessed by MRI. After the first HIFU session, MRI indicated grayscale changes for the whole mass at the lesion site, mild skin edema, and orange peel-like changes, without induration. MRI indicated coagulative necrosis in the treated region, with homogeneous enhancement at the edge of the tumor (Figure 2A). Residual tumor cells were not found in repeated biopsies at 2 and 4 wk after 5 HIFU (Figure 2B, C).
Imaging examinations

During the course of the disease (April 26th, 2017 to April 2nd, 2022), the patient underwent seven chest computed tomography (CT) scans, all of which were free of lung metastases, four whole-body bone scans (whole-body scans before and after HIFU are shown in Figure 3A, B), all of which were free of bone metastases but showed localized bone damage, and ten MRI scans (MRI scans before and after HIFU are shown in Figure 4A, B). HIFU completely ablated the tumor without complications except for localized bone damage. No further chemotherapy, radiotherapy, or biological therapy was required for tumor control.

FINAL DIAGNOSIS

Unfortunately, the patient returned 4 mo later with a 2×4 cm tissue breakdown at the site of the original lesion.

TREATMENT

The patient underwent a comprehensive evaluation followed by a successful surgery on March 3, 2022 (Figure 5A-D).

OUTCOME AND FOLLOW-UP

The patient is now in a stable condition and her limb has been preserved. Further collection and analysis of follow-up data is underway.

DISCUSSION

Diagnosing spindle cell cancers can be challenging due to their rarity and complexity. Differentiating this condition from clear spindle cell tumor, spindle cell/sclerosing rhabdomyosarcoma, and pleomorphic rhabdomyosarcoma is crucial. Factors such as age of onset, gender, clinical manifestations, and imaging findings should be carefully considered to make an accurate diagnosis. Immunohistochemical tests, including β-catenin nuclear staining, Ki-67 staining, and anti-CD34 antibody tests,
are critical in achieving a definitive diagnosis\textsuperscript{(4-6)}. De Vita et al. suggested that immunohistochemistry for MyoD1, Myogenin, and base depletion can be used in the diagnosis\textsuperscript{(7)}. By combining clinical imaging and postoperative pathology, our medical team was able to reach a definitive diagnosis of spindle cell sarcomas for this patient.

The management of limb spindle cell sarcomas requires a comprehensive approach that considers the tumor’s behavior\textsuperscript{(8)}, individual patient factors, and the potential risks and benefits of various treatment options. Surgery is typically the primary treatment option\textsuperscript{(9)}. Aggressive management is necessary for regional and distant metastases. However, despite the diligent efforts made, it is worth mentioning that limb spindle cell sarcomas still exhibit a notable recurrence rate\textsuperscript{(10-12)}, which, in certain instances, may necessitate the consideration of amputation as a treatment option. Additionally, there is a risk of radiation toxicity associated with radiotherapy\textsuperscript{(13)}. Consequently, physicians have been actively seeking alternative treatments that are both safe and effective.

Recently, HIFU has gained popularity as a treatment options for solid tumors\textsuperscript{(14)}. It utilizes a piezoelectric transducer with a fixed aperture and focal length to produce ultrasound waves (1-7 MHz). These waves have thermal and mechanical impacts on tissue. Thermal effects involve heating the targeted tissue, resulting in coagulative necrosis and cell death at higher energy doses (\textgreater 55 °C). CT or MRI guidance helps convert the ultrasound waves into heat energy, which fuses at the focal point, causing coagulation and necrosis\textsuperscript{(15)}. HIFU accurately ablates lesions without affecting surrounding normal tissues\textsuperscript{(16)}. It has been widely used for treating uterine fibroids\textsuperscript{(17)}, pancreatic cancer\textsuperscript{(18)}, prostate cancer\textsuperscript{(19)}, thyroid nodules\textsuperscript{(20)}, hepatocellular carcinoma\textsuperscript{(21)}, and even bone metastases\textsuperscript{(22,23)}. HIFU is also utilized in the treatment of soft tissue sarcomas. The application of MRI-guided focused ultrasound (MR-HIFU) in myxofibrosarcoma (MFS) has shown promise. Studies by Vanni S et al\textsuperscript{(24)}, and Zhao YX et al\textsuperscript{(25)} have reported positive results in using MR-HIFU for recurrent MFS cases, both as a palliative treatment to alleviate pain and as a curative treatment for effective local disease control. While HIFU demonstrated effectiveness in our patient, it is important to note that there was bone damage observed at the treatment site. To assess tumor
progression and bone damage during HIFU treatment, a combination of MRI and whole body bone imaging can be utilized\textsuperscript{(26,27)}.

Based on research findings, HIFU has been proven to be a safe, effective, reproducible, and minimally invasive treatment option. It offers the advantage of avoiding the potential side effects associated with radiotherapy and chemotherapy\textsuperscript{(28)}. It achieves a delicate balance by effectively preventing bleeding, minimizing tissue damage, and enhancing the overall quality of life for patients\textsuperscript{(25)}. In this case, the utilization of HIFU treatment proved to be a safe, effective, reliable, and noninvasive approach for addressing a soft tissue lesion in the left lower extremity of the patient. Nevertheless, it is important to note that the follow-up period in this study was relatively brief, and therefore, additional extensive clinical studies are required to validate and substantiate these findings.

**CONCLUSION**

In conclusion, limb spindle cell sarcomas are rare, but this case report highlights the effectiveness of non-invasive HIFU for local control of such tumors. Further studies on HIFU for treating spindle cell sarcomas are necessary to improve the management of this condition.
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