Case Report

Dedifferentiated spermatic cord liposarcoma with macroscopic ossification

Takashi Ando,[1] Ryo Nakayama,[1] Takaki Mizusawa,[1] Takeshi Sakai,[2] Hiroshi Otsuka[3] and Akiyoshi Katagiri[1]

Departments of [1]Urology, [2]Pathology, and [3]Orthopedic Surgery, Niigata Prefectural Central Hospital, Jouetsu, Niigata, Japan

Abbreviations & Acronyms
CDK-4 = cyclin-dependent kinase-4
CT = computed tomography
DDL = dedifferentiated liposarcoma
FISH = fluorescence *in situ* hybridization
MDM2 = murine double minute 2
α-SMA = α-smooth muscle actin

Correspondence: Takashi Ando M.D., Ph.D., Department of Urology, Niigata Prefectural Central Hospital, Shinnancho 205, Jouetsushi, Niigata 943-0192, Japan. Email: hawkcity@med.niigata-u.ac.jp

Received 14 June 2018; accepted 16 July 2018. Online publication 6 September 2018

Introduction: Liposarcoma is a malignant neoplasm of the adipose tissue, and dedifferentiated liposarcoma is a relatively rare subtype. Liposarcomas are typically located in the lower extremities or retroperitoneum, but dedifferentiated liposarcoma of the spermatic cord is rare and no case of it with macroscopic ossification has been reported in the literature.

Case presentation: A male presented to our hospital with a painful, palpable, and hard left suprapubic mass, formed over 3 months. The mass was diagnosed as a spermatic cord tumor and was resected using high orchiectomy. Due to the histological diagnosis of dedifferentiated spermatic cord liposarcoma with ossification and positive margins, a second extended resection and adjuvant radiotherapy were performed.

Conclusion: To differentiate spermatic cord liposarcoma preoperatively is difficult. There is no gold standard treatment for it, although surgical complete resection with clear microscopic margins would be the most effective treatment.

Key words: liposarcoma, orchiectomy, osteogenesis, radiotherapy, spermatic cord.

Keynote message

As dedifferentiated spermatic cord liposarcoma is rare without any standard treatment and to differentiate spermatic cord liposarcoma preoperatively is difficult, further accumulation of relevant knowledge and clinical experience is required.

Introduction

Liposarcoma, a subtype of sarcoma, is a malignant neoplasm of the adipose tissue. In 2013, liposarcoma was histologically classified by the World Health Organization. DDL is a relatively rare subtype of liposarcoma.[1] Liposarcomas are located most commonly in the lower extremities or the retroperitoneum, but spermatic cord liposarcoma is rare. Few cases of spermatic cord DDL have been published in the literature, and there are no reports on spermatic cord DDL with macroscopic ossification.[2,3] The available literature states that the preferred treatment for spermatic cord liposarcoma is surgical resection using orchiectomy with high cord ligation for tumors with negative surgical margins.[3,4] However, the first surgical resection is often insufficient because an accurate preoperative diagnosis of spermatic cord liposarcoma is difficult.[4,5] To prevent local recurrence, the most common type of recurrence, re-resection with wide surgical margins should be performed. Herein, we report the first case of spermatic cord DDL with macroscopic ossification treated with extended re-resection and adjuvant radiotherapy.

Case presentation

A 62-year-old male presented to our hospital with a painful, chronic swelling that had formed over 3 months. Clinical examination and ultrasonography revealed a solid mass with no mobility in the left suprapubic region and normal testes. The patient was afebrile without any signs of urinary tract infection. Laboratory findings, including testicular tumor

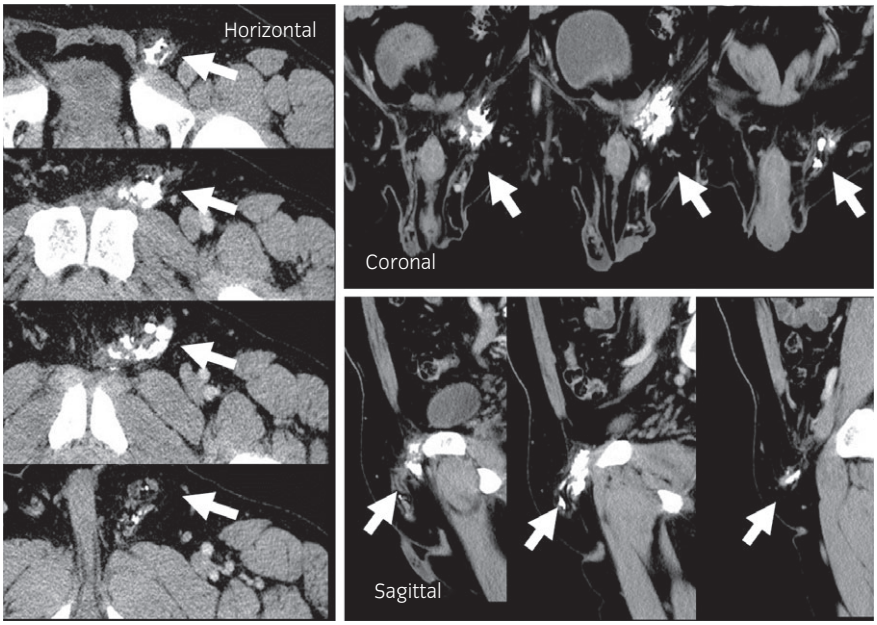

Fig. 1 Enhanced CT scan showing a solid soft tissue mass with calcification originating from the spermatic cord (white arrows).

markers (serum levels of α-fetoprotein, human chorionic gonadotropin β, and lactate dehydrogenase), were normal. CT exhibited a solid soft tissue mass (approximately 10 cm) with calcification originating from the left spermatic cord (Fig. 1). Retroperitoneal and pelvic lymph nodes were normal without any metastasis. A spermatic cord tumor was clinically diagnosed, and surgical resection was performed using orchiectomy with high spermatic cord ligation. The hard tumor surrounding the spermatic cord was resected along with the left testis and spermatic cord (Fig. 2), and the testis was normal. Pathological examination revealed the presence of a spindle cell neoplasm, atypical adipocytes, atypical stromal cells, and ossification (Fig. 3). Immunohistochemical tumor markers including CDK4 (+) and MDM2 (+) in the spindle cells as well as atypical adipocytes and stromal cells suggested DDL. In addition, vimentin (+) and α-SMA (+) in the spindle cells indicated differentiation. As the pathological surgical margin was positive, our orthopedic surgeons performed a second extended surgery to remove skin, fat, and the left portion of the pubic bone surrounding the tumor. The second surgical specimen did not reveal any remaining tumor cells. The patient underwent adjuvant local radiotherapy (60 Gy) and strict follow-up with chest/abdomen CT every 3 months for the first year. We also planned long-term follow-up with chest/abdomen CT every 6 months within 10 years from the surgery.

Discussion

Neoplasms originating from smooth muscle, fibrous tissue, or fat composing the spermatic cord are rare. Benign tumors such as lipomas are the most common neoplasms

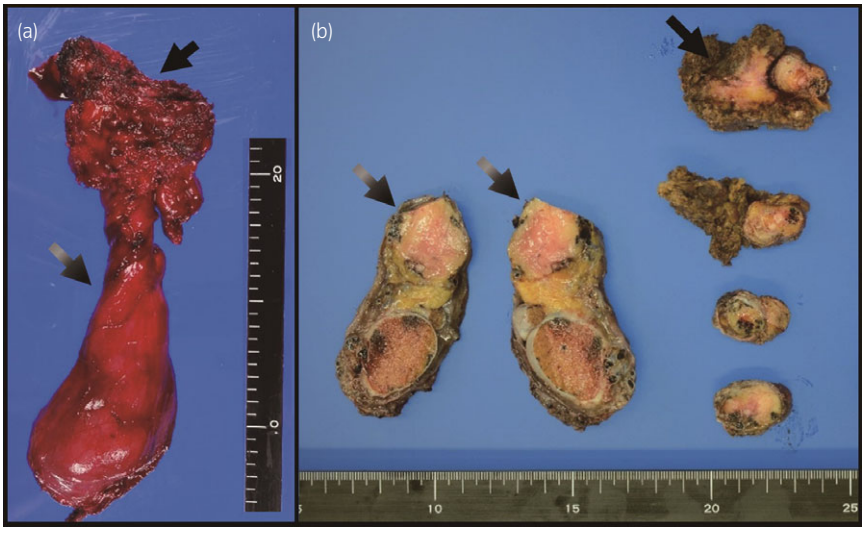

Fig. 2 Tumor resection along with the left testis and ipsilateral spermatic cord (a) and specimen after formalin fixation (b). Ossification is indicated by the black arrows.

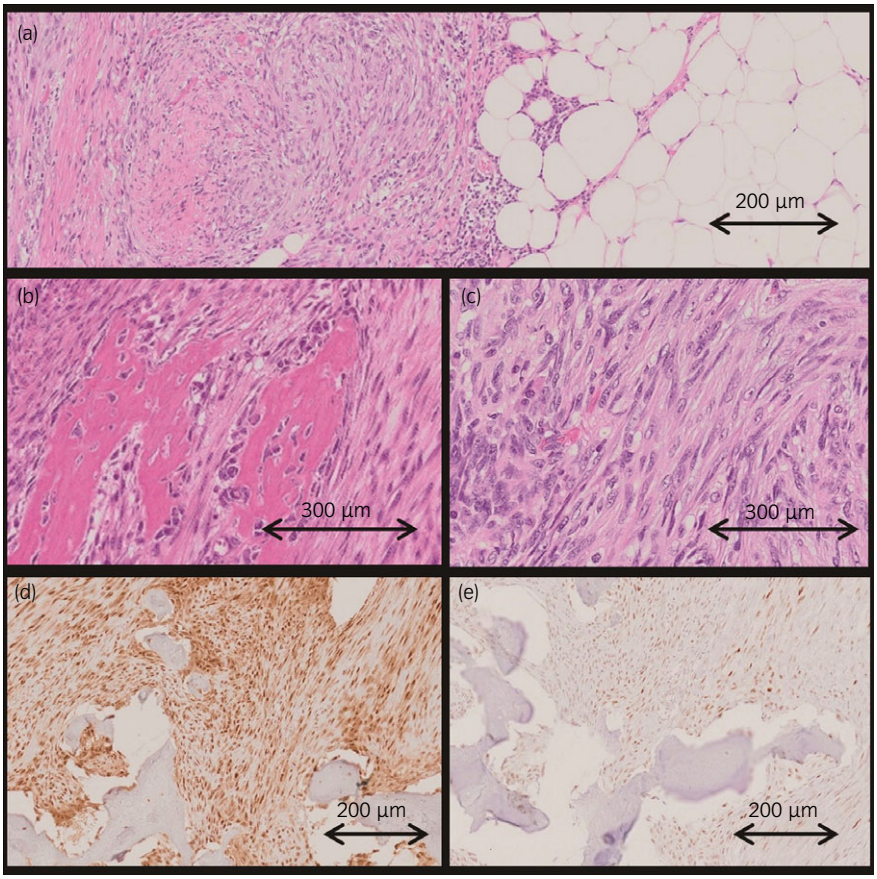

Fig. 3 Spindle cell neoplasm with atypical adipocytes, stromal cells, and mature-looking adipocytes. Hematoxylin and eosin stain (HE) (a). Presence of ossification. HE (b). Presence of spindle cell neoplasm. HE (c). Immunostaining by CDK-4 in the spindle cells (d). Immunostaining by MDM2 in the spindle cells (e).

of the spermatic cord. Spermatic cord liposarcomas have been discussed in the literature since 1845 when Lesauvage *et al.* reported the first case.[5] Typically located in the lower extremities or retroperitoneum, liposarcomas most frequently occur in adults and are classified into five subgroups: (i) well differentiated, (ii) dedifferentiated, (iii) myxoid, (iv) round cell, and (v) pleomorphic.[1] Moreover, spermatic cord liposarcoma accounts for 3–7% of all spermatic cord tumors,[5] and spermatic cord DDLs have been primarily reported in individual case reports.[3] According to the literature, spermatic cord DDLs originate in the distal and middle portions of the cord. DDLs with ossification located in the retroperitoneum are rarely reported,[2,6] and there are no reports of spermatic cord DDL with macroscopic ossification, similar to that in the present case. Due to its rarity, the clinical role of ossification within DDL remains unknown.

Spermatic cord liposarcoma must be differentiated from paratesticular disease; however, it may be misdiagnosed as another common disease, such as inguinal hernia or hydrocele, owing to its nonspecific symptoms, including painless swelling and palpable mass.[5] Although CT or magnetic resonance imaging may be used to differentiate spermatic cord tumor from other common diseases, malignancy is difficult to differentiate preoperatively. Immunohistochemistry for MDM2 and CDK4 has high sensitivity for diagnosing DDL,[3] whereas immunohistochemistry for vimentin or α-SMA indicates differentiation toward non-epithelial cells or smooth muscles respectively. Recently, the usefulness of FISH for diagnosing and categorizing of liposarcoma was reported, although we could not perform it because of equipment insufficiency.[7,8] FISH could identify chromosomal translocations and amplification of MDM2 or CDK4 in tumor cells. Using FISH technique, the existence of osteogenic differentiation in a part of DDL was investigated in detail and it seemed that ossification within DDL was neoplastic rather than metaplastic according to recent reports.[9,10]

Due to its rarity, there is no gold standard treatment for spermatic cord liposarcomas, including DDL, although previous reports recommend surgical resection using radical high orchiectomy from as close to the deep ring as possible as the most effective treatment.[3,4] These reports acknowledge the importance of complete resection with clear microscopic margins and re-resection with wide resection in cases with positive margins in order to achieve a negative margin and to prevent local recurrence. Routine lymph node dissection is not recommended unless there is an evidence of a tumor. The use of adjuvant radiotherapy for spermatic cord liposarcoma is controversial due to the lack of data on its effectiveness. The recommended dose for adjuvant radiotherapy of liposarcoma is 60 Gy, and the radiation field should cover the internal inguinal ring.[5] However, there is no definitive role of chemotherapy in managing localized spermatic cord liposarcoma.

Reportedly, local recurrence is the most common form of spermatic cord liposarcoma recurrence, accounting for approximately 20% of recurrence cases.[3] DDL has the potential to cause distant metastasis and recurrence, and although late recurrence has been reported, no specific outcome data are available to date, thus long-term follow-up of at least 10 years is recommended.[3–5]

In conclusion, we reported the first case of spermatic cord DDL with macroscopic ossification. Due to its rarity, there is no standard treatment for spermatic cord liposarcomas, including DDL, and further research and clinical experience in treating these tumors is necessary.

Conflict of interest

The authors declare no conflict of interest.

References

1 Jo VY, Fletcher CD. WHO classification of soft tissue tumours: an update based on the 2013 (4th) edition. *Pathology* 2014; **46**: 95–104.
2 Chondros K, Heretis I, Papadakis M *et al.* Dedifferentiated paratesticular liposarcoma with osseous metaplasia. *Case Rep. Urol.* 2015; **2015**: 965876.
3 Kryvenko ON, Rosenberg AE, Jorda M, Epstein JI. Dedifferentiated liposarcoma of the spermatic cord: a series of 42 cases. *Am. J. Surg. Pathol.* 2015; **39**: 1219–25.
4 Chalouhy C, Ruck JM, Moukarzel M, Jourdi R, Dagher N, Philosophe B. Current management of liposarcoma of the spermatic cord: a case report and review of the literature. *Mol. Clin. Oncol.* 2017; **6**: 438–40.
5 Chintamani, Tandon M, Khandelwal R *et al.* Liposarcoma of the spermatic cord: a diagnostic dilemma. *JRSM Short. Rep.* 2010; **1**: 49.
6 Arakaki R, Yoshikawa T, Yamada H, Kamoto T, Mikami Y. A case of retroperitoneal well-differentiated liposarcoma with metaplastic bone formation. *Hinyokika Kiyo.* 2010; **56**: 697–700.
7 Asif A, Mushtaq S, Hassan U *et al.* Fluorescence in situ hybridization (FISH) for differential diagnosis of soft tissue sarcomas. *Asian Pac. J. Cancer Prev.* 2018; **19**: 655–60.
8 Shahi F, Alishahi R, Pashaiefar H *et al.* Differentiating and categorizing of liposarcoma and synovial sarcoma neoplasms by fluorescence in situ hybridization. *Iran J. Pathol.* 2017; **12**: 209–17.
9 Yamashita K, Kohashi K, Yamada Y *et al.* Primary extraskeletal osteosarcoma: a clinicopathological study of 18 cases focusing on MDM2 amplification status. *Hum. Pathol.* 2017; **63**: 63–9.
10 Macagno N, Fuentes S, de Pinieux G *et al.* Paravertebral well-differentiated liposarcoma with low-grade osteosarcomatous component: case report with 11-year follow-up, radiological, pathological, and genetic data, and literature review. *Case Rep. Pathol.* 2017; **2017**: 2346316.