

Spermatic Cord Liposarcoma Associated with Prostate Cancer

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ABSTRACT

We report on a case of liposarcoma of the spermatic cord treated with radical orchidectomy and wide excision of the tumour without adjuvant treatment. After 16 years of follow up, neither local nor distant recurrences were detected. Spermatic cord liposarcoma is a rare disease. The basic treatment for all patients with spermatic cord liposarcoma is radical orchidectomy with wide local resection and high ligation of the spermatic cord. The role of postoperative irradiation depends on pathologic findings. Local recurrences are rare but because of late events, a long follow-up is mandatory.

Keywords: Spermatic cord, Liposarcoma, Prostate cancer

ÖZET

Spermatik Kord Liposarkomuna Eşlik Eden Prostat Kanseri

Sunulan olgu, radikal orşiektomi ve geniş lokal eksizyon ile tedavi edilmiş ve başka adjuvan tedavi almamış spermatik kord yerleşimli liposarkomdur. 16 yıl boyunca ne lokal ne de uzak metastaz saptanmadı. Spermatik kord liposarkomu nadir görülen bir hastalıktır. Spermatik kord liposarkomalı tanılı bütün hastalarda ana tedavi radikal orşiektomi ve geniş lokal eksizyon ve spermatik kordun yüksek bağlanmasıdır (high ligation). Postoperatif radyoterapinin rolü patolojik bulgulara bağlıdır. Lokal nüks nadirdir olup geç olaylar için uzun süre takip gerekmektedir.

Anahtar Kelimeler: Spermatik kord, Liposarkom, Prostat kanseri

INTRODUCTION

Spermatic cord liposarcoma is a rare malignant tumour. Most available information on these tumors derives from small series or case reports.¹⁻¹¹ Sarcomas are uncommon tumors with an overall incidence of 4-6/100,000. Sarcomas of the genitourinary tract account for about 5% of these cases, and 2% of all urological tumors. The spermatic cord is the most commonly involved urological site and accounts for approximately 30% of all genitourinary sarcomas.^{6,9,12,13} Herein, we report a case of a liposarcoma of the spermatic cord with a 16-year follow-up, and review the literature.

CASE REPORT

A 57-year-old man initially presented to his physician in January 1980 with one-month history of a painless, slowly growing right inguinal mass. Physical examination revealed a solid mass 2-3 cm superior to the right testis extending into the inguinal canal. Both testes were normal in size and consistency. There were no signs of inguinal hernia. The tumour was not adherent to the testis or epididymis, and was not translucent. Ultrasonography was carried out, and confirmed the presence of an extra-testicular mass.

Based on the above-mentioned findings, surgical exploration was done with an incision extending into the right inguinal canal in the spermatic cord. The right testis and epididymis were found to be macroscopically normal. A solid mass was found 3 cm proximal to the testis. A liposarcoma was suspected on frozen sections. Radical orchidectomy with en bloc excision of the tumour and a high ligation of the spermatic cord at the internal inguinal ring was performed.

On microscopic examination, the mass was interpreted as a poorly-differentiated liposarcoma of the sclerotic variant, measuring 7 x 5.5 x 6 cm. Surgical margins were free of tumour. As the operation was radical, no adjuvant therapy was proposed.

Sixteen years later, in March 1996, the patient was referred to our department for radical radiotherapy of a well differentiated prostatic carcinoma. This tumour was discovered based on an elevated PSA (9.3 µg/l) and a transurethral resection of the prostate, which was performed thereafter. Digital rectal examination revealed a T2a tumour. There were no palpable signs of local recurrence concerning right inguinal area. Chest X-ray revealed no signs of metastases. Abdominopelvic CT scan and bone scintigraphy were free of any pathologic finding.

Facing the long survival of this patient with spermatic cord liposarcoma, diagnosed initially as poorly differentiated, the pathological slides were reviewed and the diagnosis changed to that of well-differentiated liposarcoma, sclerosing variant.

DISCUSSION

The majority of primary malignant tumours of the spermatic cord in adults are usually sarcomatous in nature.^{14,15} Liposarcomas, leiomyosarcomas and rhabdomyosarcomas are the most common soft tissue malignancies in adults and infants, respectively. These malignant tumours are thought to be of mesenchymal origin, and are divided into two groups, namely, highly aggressive childhood rhabdomyosarcoma and undifferentiated sarcomas, which rapidly disseminate via lymphatics and blood vessels. The sarcomas of the second group are less aggressive, and usually occur in adults. Liposarcoma accounts for about 7% of the paratesticular sarcomas.^{9,16}

The origin of spermatic cord liposarcoma is controversial. The structures of the spermatic cord originate from the mesonephric duct, and the histological types of these neoplasms do reflect their embryological origin. The tumour genesis is unclear: some authors hypothesize that the tumour arises from the retroperitoneal fat. Others postulate that those lesions may arise as a result of a malignant transformation of a pre-existing lipoma² but, to date, it is commonly accepted that reported cases of malignant transformation of lipoma are merely diagnostic errors; the tumour being an under-recognized liposarcoma from the beginning.¹⁷

Clinical differential diagnosis with other scrotal masses, both benign and malignant is often difficult. Different radiological examinations like ultrasound, CT scan and MRI can help to establish a diagnosis before surgery.

Liposarcomas can be classified roughly into three categories according to the dominant cell type¹⁷; i.e., well differentiated, myxoid-round cell, and pleomorphic. However, the most recent WHO classification recognizes five main categories, i.e., well differentiated (including lipoma-like, sclerosing, inflammatory dedifferentiated), myxoid, round cell, and pleomorphic. For each category, clinical setting, morphological findings, and most importantly cytogenetic abnormalities are specific. Liposarcoma are usually soft; consistency, color and aspect of the cut surface vary depending on the histological structure. The aspect is pale yellow and mucoid, translucent in

the myxoid forms: it is yellowish or pale orange, soft, oil, friable, in the well-differentiated forms, or in any case rich in adipoblasts-adipocytes; whiter and firmer in the forms having a larger spindle-cell and collagen quota; softer, encephaloid, in scarcely differentiated forms.¹⁷ Well-differentiated and dedifferentiated liposarcomas are characterized by the presence of giant marker and ring chromosomes derived from the q13-15 region of chromosome 12.^{18,19} Well-differentiated liposarcoma must be distinguished from inflammatory lesions (liponecrosis and reactions to silicone implants) and from varieties of lipoma, myxoid liposarcoma has such abundant and typical vascularisation that it may easily be distinguished from intramuscular myxoma. Round-cell liposarcoma must be differentiated from vacuolized round-cell sarcomas, such as embryonal rhabdomyosarcoma, but in the latter recognition of the rhabdomyoblasts will orient diagnosis.

Radical orchidectomy with wide local resection and high ligation of the spermatic cord at the internal inguinal ring is currently the procedure of choice for well-differentiated liposarcomas.^{4,8,10,20-22} Complete excision is difficult to obtain for liposarcoma of the spermatic cord as well as for the retroperitoneal location. Retroperitoneal lymphadenectomy is not indicated (reported only in one case) since well-differentiated liposarcomas tend to spread primarily by local extension. The most suitable type of treatment, for myxoid and well-differentiated liposarcoma -nearly all stage I-, is wide removal. For high grade liposarcoma -stage II- radical removal may be preferred, as that which offers the highest guarantee. Radiotherapy is effective, particularly in the myxoid liposarcoma, and is thus widely used in association with surgery.²⁰⁻²² Hematogeneous and lymphatic spread are usually late events seen in high-grade tumors, especially in well differentiated liposarcomas undergoing dedifferentiation. In liposarcomas of the spermatic cord, regional lymph node metastases have not been described. Hematogeneous metastases are cited in two case reports with lung, bone, or brain localizations without histological confirmation.^{11,23,24}

Postoperative radiotherapy was proposed to reduce the incidence of recurrence after radical surgery.^{5,9,22,24} Local failure is not rare for such tumours, especially after inadequate surgery or in a high-grade malignant tumour (i.e., dedifferentiated pleomorphic and round cell types).^{7,11} Those tumours can be considered as radiosensitive (dose range in the literature from 40-60 Gy).

Chemotherapy, especially doxorubicin and ifosfamide, has been reported occasionally in case of recurrent disease.^{5,11,22,25}

In conclusion, well-differentiated liposarcoma of the spermatic cord is a rare malignancy of good prognosis. Wide excision of the tumour with orchidectomy is the treatment of choice. Local recurrences are described after inadequate surgery and/or in high-grade lesions. In such patients, adjuvant radiotherapy and/or chemotherapy may be of benefit. Since recurrence may occur for many years after the initial diagnosis, long-term follow-up is mandatory.

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