

Role of radiotherapy in the treatment of fibrosarcoma of the spermatic cord: a case report and review of the literature

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ABSTRACT

Background. Spermatic cord sarcomas are rare. The therapeutic approach is based only on case reports and small series. The standard treatment is radical orchiectomy with wide local resection, while the role of adjuvant therapies is not clear. We present a case of fibrosarcoma of the spermatic cord treated with surgery and adjuvant radiotherapy. A review of the literature about the role of adjuvant treatments is also discussed.

Case report. A 59-year-old man presented a right testicular mass of about 4 × 3 cm in size. Biopsy showed a high-grade polymorphous sarcoma, consistent with a diagnosis of poorly differentiated fibromyosarcoma. He underwent a right radical inguinal orchiectomy and adjuvant radiotherapy (total dose: 5940 cGy). During treatment the patient developed a G3 skin toxicity (RTOG score) in the inguinal fold. After a follow-up of 57 months, he is alive and without evidence of local or distant recurrence. No late toxicity was noted.

Conclusion. The optimal adjuvant management of spermatic cord sarcoma is still uncertain. Looking at the literature, it seems that adjuvant radiotherapy can improve loco-regional control and disease-free survival without additional late toxicity.

Introduction

Spermatic cord sarcomas are rare tumors and the therapeutic approach is based only on case reports or small series¹. Due to the rarity of this disease, it is difficult to know its natural history and to reach conclusions about the treatment results. Even if radical orchiectomy with wide local resection of the surrounding soft tissues remains the standard², tumor-negative margins are not easy to obtain because of local anatomical constraints³. Although most sarcomas of the spermatic cord are well differentiated, the propensity for local recurrence is high: local and/or ipsilateral nodal recurrences are frequent (50-75%) after surgery³⁻⁵. A strategy to reduce this high local recurrence rate could be the combination of surgery and radiation therapy. However, the role of loco-regional and/or systemic therapies such as retroperitoneal node dissection, adjuvant radiation therapy or chemotherapy remains controversial^{6,7}. We present a case of fibrosarcoma of the spermatic cord treated with surgery and adjuvant radiotherapy. A review of the literature about the role of adjuvant treatments is also discussed.

Case report

In January 2005, a 59-year-old man was admitted with a right testicular mass. Ultrasonography showed a roundish, heterogeneous and hypoechoic mass (about 4 × 3

Key words: spermatic cord, fibrosarcoma, radiotherapy, adjuvant therapy.

Conflict of interests: The authors have no conflict of interests related to this work.

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Received January 11, 2011;
accepted February 11, 2011.

cm in size), with hypervascularization on color-Doppler imaging. A biopsy was performed and histopathological examination showed a high-grade polymorphous sarcoma, consistent with a diagnosis of poorly differentiated fibromyosarcoma. Total-body CT scan did not reveal any nodal or distant metastasis. A right radical inguinal orchiectomy was performed with removal of the testis, epididymis and all spermatic cord as far as the internal inguinal ring. Postoperative total-body PET scan was negative for residual disease and distant metastasis.

Because of the poor differentiation of the disease, adjuvant radiotherapy was delivered in order to reduce the risk of local relapse. The clinical target volume included the tumor bed and ipsilateral pelvic and inguinal nodes (Figure 1). Three-dimensional conformal radiotherapy (3D-CRT) with 2 opposed 10-MV fields (AP-PA) was delivered by Linear Accelerator. The total dose prescribed at the isocenter was 5940 cGy (in daily fractions of 180 cGy).

The patient developed G3 skin toxicity (RTOG score⁸) in the inguinal fold after 3960 Gy. For this reason, treatment was interrupted for 6 days. After its recovery the patient finished the radiation therapy without further interruptions. He has been followed with clinical and laboratory evaluations every 3 months and 6-monthly CT and/or PET scans. After 57 months' follow-up, the patient is alive and there is no evidence of local or distant recurrence. At the last follow-up visit we did not find any signs or symptoms of late toxicity according to the RTOG-EORTC score⁸.

Discussion

Soft tissue sarcomas include a variety of neoplasms arising from tissues of mesodermal origin. Genitourinary sarcomas are uncommon in adults, making up less than 2.7% of all sarcomas, and paratesticular sarcomas represent less than 1% of them⁹. The majority of paratesticular neoplasms originate from mesenchymal tis-

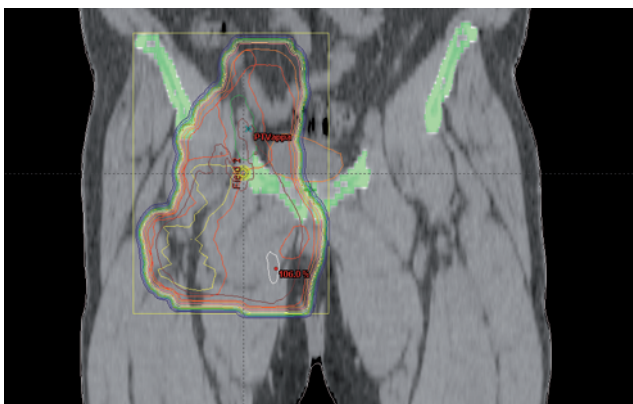


Figure 1 - Treatment plan including tumor bed and ipsilateral pelvic and inguinal nodes.

sue¹⁰. These cancers predominantly affect patients during the sixth and seventh decades of life¹¹ and up to 30% are malignant. Approximately 10% of spermatic cord sarcomas are leiomyosarcomas¹².

This histological type derives from the smooth muscle of different areas such as the testicular duct wall, blood vessels, and cremaster muscle, and is thought to arise as a result of malignant degeneration of previous leiomyomatous neoplasms¹³. In fact, spermatic cord sarcomas may arise from any mesodermal-derived cord structure¹⁴.

Evaluation of a scrotal mass requires a physical examination, measurement of serum tumor markers including alpha-fetoprotein and human chorionic gonadotropin¹⁵, and appropriate imaging studies. Ultrasonography should be the primary imaging modality for scrotal or inguinal masses¹⁶. If a neoplasm is found, CT should be used to demonstrate the extratesticular origin of the mass and to determine its scrotal extension for local staging. Moreover, CT is necessary for pelvic staging and searching for distant metastasis¹⁷.

Radical orchiectomy including tumor resection with negative microscopic margins is the standard treatment. The role of adjuvant treatments has not been defined because of the rarity of this tumor but, in our opinion, these tumors should preferentially be treated by a multidisciplinary team. There is a body of evidence supporting this idea. First of all, the tumors arise in a site where wide circumferential resection margins may be difficult to obtain³. This is analogous to sarcomas of other sites such as the head and neck region, where there are similar surgical margin constraints and where local relapse rates as high as 50% with surgery alone have dropped to 10-20% with the addition of adjuvant radiation¹⁸. In a report of Merimsky *et al.* the median time to first relapse in 13 patients was 5.5 months¹⁹. Local infiltration of the scrotum, inguinal canal or pelvic tissues along the pathways of the vasa deferens may be another cause of local failure²⁰. Moreover, even if radical orchiectomy is an essential component in the management of spermatic cord sarcoma, the reported survival rates after surgery alone (50% to 80%) indicate a need for additional treatment³. Blitzer *et al.* reported a 75% local recurrence rate in cases treated with radical orchiectomy alone and emphasized the need for adjuvant therapies⁴.

Moreover, it is generally accepted that, unlike soft tissue sarcomas of other locations where regional lymph node spread is uncommon (with the exception of certain histologies such as synovial cell sarcoma, rhabdomyosarcoma, and epithelioid sarcoma), spermatic cord sarcoma frequently disseminates to regional lymphatics. The most commonly involved nodes are the external iliac, hypogastric, common iliac and para-aortic nodes. Banowsky *et al.* reported that the probability of metastasis to the regional lymph nodes is higher than 29%²¹. Merimsky reported lymph node metastases in 2 of 6 patients who underwent lymph node dissection²².

Locoregional treatments include prophylactic retroperitoneal lymph node dissection (RPLND) and adjuvant radiation therapy. No significant benefit from the addition of RPLND to radical orchiectomy has been demonstrated, however¹⁸.

Even if there are no sufficiently large series to evaluate the efficacy of adjuvant radiation therapy for spermatic cord sarcoma and prospective trials are not possible because of its rarity, the limited series that have been studied seem to confirm the efficacy of adjuvant radiotherapy in drastically reducing local relapse rates. In the series of Fagundes *et al.*²⁰, 5 of 9 patients treated with radical orchiectomy alone subsequently developed locoregional failure, in 2 cases limited to lymph nodes. By contrast, there were no locoregional recurrences among the 9 patients who received adjuvant radiation to regional lymph nodes following radical orchiectomy. This correlated with a significant difference in locoregional control and disease-free survival in favor of adjuvantly treated patients²⁰. In the report by Catton³, 2 of the 14 patients (14%) with primary spermatic cord sarcoma treated with orchiectomy alone developed local relapse, while all of the 5 patients who received adjuvant irradiation maintained local control. Finally, even if the role of adjuvant treatment still remains uncertain²², radiation therapy should be recommended in addition to surgery at least in cases with evidence of more aggressive tumor behavior (i.e., high-grade tumor, lymphatic invasion, inadequate resection margin, or recurrence). Regardless of the initial therapy, the high risk of local and distant recurrence always necessitates long-term follow-up²³.

Conclusions

The optimal management of spermatic cord sarcoma is still uncertain. The role of radiotherapy remains unclear. However, looking at the literature, it seems that adjuvant radiotherapy to the inguinal canal and ipsilateral pelvic nodes can improve locoregional control and disease-free survival without late toxicity.

References

1. Coleman J, Brennan MF, Alektiar K, Russo P: Adult spermatic cord sarcoma: management and results. *Ann Surg Oncol*, 10: 669-675, 2003.
2. Sclama AO, Berger BW, Cherry JM, Young JD Jr: Malignant fibrous histiocytoma of the spermatic cord: the role of retroperitoneal lymphadenectomy in management. *J Urol*, 130: 577-579, 1983.
3. Catton CN, Cummings BJ, Fornasier V, O'Sullivan B, Quirt I, Warr D: Adult paratesticular sarcomas: a review of 21 cases. *J Urol*, 146: 342-345, 1991.
4. Blitzer PH, Dosoretz DE, Proppe KH, Shipley WU: Treatment of malignant tumors of the spermatic cord: a study of 10 cases and a review of the literature. *J Urol*, 126: 611-614, 1981.
5. Ballo MT, Zagars GK, Pisters PW, Feig BW, Patel SR, von Eschenbach AC: Spermatic cord sarcoma: outcome, patterns of failure and management. *J Urol*, 166: 1306-1310, 2011.
6. Schwartz SL, Swierzewski SJ, Sondak VK, Grossmann HB: Liposarcoma of the spermatic cord: report of 6 cases and review of the literature. *J Urol*, 153: 154-157, 1995.
7. Kanso C, Roussel H, Zerbib M, Flam T, Debré B, Vieillefond A: Adult spermatic cord sarcoma: diagnosis and management. *Prog Urol*, 21: 53-58, 2011.
8. Cox JD, Stetz J, Pajak TF: Toxicity criteria of the Radiation Therapy Oncology Group (RTOG) and the European Organization for Research and Treatment of Cancer. *Int J Radiat Oncol Biol Phys*, 31: 1341-1346, 1995.
9. Russo P, Brady MS, Conlon K, Hajdu SI, Fair WR, Herr HW, Brennan MF: Adult urological sarcoma. *J Urol*, 147: 1032-1037, 1992.
10. Bajaj P, Agarwal K, Niwedita SR, Pathania OP: Leiomyosarcoma arising from tunica vaginalis testis: a case report. *Indian J Pathol Microbiol*, 44: 145-146, 2001.
11. Rao CR, Srinivasulu M, Naresh KN, Doval DC, Hazarika D: Adult paratesticular sarcomas: a report of eight cases. *J Surg Oncol*, 56: 89-93, 1994.
12. Enoch S, Wharton SM, Murray DS: Management of leiomyosarcomas of the spermatic cord: the role of reconstructive surgery. *World J Surg Oncol*, 3: 23, 2005.
13. Avila Padilla J, Capell González M, Muniesa Calderó M, Yelletisch Araújo A, Badía Torroella F, Abós Fanlo P: Leiomyosarcomas of the spermatic cord. *Actas Urol Esp*, 17: 464-467, 1993.
14. Srigley JR: The paratesticular region: histoanatomic and general considerations. *Semin Diagn Pathol*, 17: 258-269, 2000.
15. Ou SM, Lee SS, Peng YJ, Sheu LF, Yao NS, Chang SY: Production of beta-HCG by spermatic cord leiomyosarcoma: a paraneoplastic syndrome? *J Androl*, 27: 643-644, 2006.
16. Yang DM, Kim HC, Lim JW, Jin W, Ryu CW, Kim GY, Cho H: Sonographic findings of groin masses. *J Ultrasound Med*, 26: 605-614, 2007.
17. Cardenosa G, Papanicolaou N, Fung CY, Tung GA, Yoder IC, Althausen AF, Shipley WU: Spermatic cord sarcomas: sonographic and CT features. *Urol Radiol*, 12: 163-167, 1990.
18. Tran LM, Mark R, Meier R, Calcaterra TC, Parker RG: Sarcomas of the head and neck. *Cancer*, 70: 169-177, 1992.
19. Merimsky O, Terrier P, Bonvalot S, Le Pechoux C, Delord JP, Le Cesne A: Spermatic cord sarcoma in adults. *Acta Oncol*, 38: 635-638, 1999.
20. Fagundes MA, Zietman LA, Althausen AF: The management of spermatic cord sarcoma. *Cancer*, 77: 1873-1876, 1996.
21. Banowsky LH, Schultz GN: Sarcoma of the spermatic cord and tunics: review of the literature, case report and discussion of the role of retroperitoneal lymph node dissection. *J Urol*, 103: 628-631, 1970.
22. Kanso C, Roussel H, Zerbib M, Flam T, Debré B, Vieillefond A: Adult spermatic cord sarcoma: diagnosis and management. *Prog Urol*, 21: 53-58, 2011.
23. May M, Seehafer M, Helke C, Gunia S, Hoschke B: Liposarcoma of the spermatic cord-report of one new case and review of the literature *Aktuelle Urol*, 35: 130-133, 2004.