

Irradiation-associated Paratesticular Leiomyosarcoma

Yi-Shiou Tseng¹, Chia-Tung Shun³, Shiu-Dong Chung^{1,2}, Hong-Jeng Yu², Chao-Yuan Huang²

¹Division of Urology, Department of Surgery, Far Eastern Memorial Hospital, Taipei, Taiwan; ²Department of Urology, and ³Department of Pathology, National Taiwan University Hospital, Taipei, Taiwan

We describe a case of paratesticular leiomyosarcoma occurring in a 37-year-old man, 34 years after receiving wide excision and irradiation for fibrosarcoma of the left thigh. To our knowledge, this is the first report of irradiation-associated testicular leiomyosarcoma of a Chinese man, who presented a progressively enlarged non-tender left scrotal mass. He died from metastatic disease 20 months after radical orchiectomy with adjuvant radiation therapy and systemic chemotherapy. (*JTUA* 20:146-148, 2009)

Key words: sarcoma; testis; radiation; secondary malignancy

INTRODUCTION

Primary paratesticular leiomyosarcoma is an extremely rare tumor especially after adjacent field irradiation with only a few reported cases in the literature and none has been reported in men of Chinese descent. We describe this first case of a primary paratesticular leiomyosarcoma in a younger Chinese male associated with previous exposure history of irradiation due to fibrosarcoma.

CASE REPORT

A 37-year-old man was referred to our department presenting with left scrotal mass noted for more than one year. He had received operation for fibrosarcoma wide excision of the left thigh and followed by localized irradiation at his age of 3-year-old. His medical history was unremarkable especially no steroid usage or diabetes mellitus. Physical examination disclosed a hard, enlarged left scrotal mass with superficial epidermal ulceration and bloody discharge, measuring around 12 × 12 × 11cm. in size (Fig. 1A). No peripheral lymph nodes were palpable. The serum alpha-fetoprotein, beta-hCG or LDH were all in normal range. Chest radiograph did not show any pulmonary lesion. Abdomen-pelvis com-

puterized tomography and bone scan were negative for visceral or bone metastasis. After scrutinized examination, left inguinal radical orchiectomy with adjacent regional lymph node dissection was performed subsequently. Grossly, the tumor mass was an ill-defined yellowish-white (Fig. 1B), solid mass with focal necrosis and hemorrhage, which seemed arising from the paratesticular tissue and the testis per se is compressed atrophy. The spermatic cord was also spared. Microscopic examination showed tumor consists of hyperchromatic spindle neoplastic cells occasional arranged in interlacing fascicles with marked nuclear atypia and frequent mitosis (>20 per 10 high power field) and (Fig. 1C). Immuno-histochemical staining showed the tumor cells were strongly positive for smooth muscle actin (Fig. 1D), vimentin and negative for EMA, S-100, CD68, B-HCG, and alpha-fetoprotein. On the basis of the gross findings, microscopic examination and immunohistochemical studies, the tumor was diagnosed as a paratesticular leiomyosarcoma and associated five lymph nodes obtained did not show metastasis. Adjuvant radiotherapy or chemotherapy was conducted. Unfortunately, metastases to lung, liver and bones developed 15 months later, which did not respond to further chemotherapy. He died 20 months after the radical orchiectomy and adjuvant chemoradiation therapy.

DISCUSSION

Leiomyosarcoma occurs commonly in the gastrointestinal tract such as colon and stomach but is rare in the genital region. The presence of such elements in the scrotal subcutis, dartos muscle, tunica albuginea,

Address reprint requests and correspondence to:
Shiu-Dong Chung, MD
Division of Urology, Department of Surgery, Far Eastern Memorial Hospital, Taipei, Taiwan 21, Sec. 2, Nan-Ya South Rd., Taipei, Taiwan 220, ROC
TEL: 886-2-89667000 ext 1313 FAX: 886-2-89665567
E-mail: b3401095@mail2000.com.tw

blood vessels, and testicular parenchyma could provide a site of origin for these rare smooth muscle tumors. Paratesticular leiomyosarcoma is a rare and described as an indolent tumor with the potential for distant metastases. A few cases of primary paratesticular leiomyosarcoma had been reported, however, the age at diagnosis were most after the fifth decade of life.¹ The occurrence in younger cases is suggested to be associated history of anabolic steroid abuse.² Irradiation-related sarcoma were reported as post-irradiation complication and had poor prognosis.³ The criteria for the diagnosis of postirradiation sarcoma was first proposed by Cahen et al in 19484 and consisted of (1) history of radiation therapy, (2) sarcoma arising within the radiation field after the radiation has been given, (3) long latency period, and (4) histologic confirmation of the sarcoma. The case is thought as irradiation-associated

because he had a histologically proven leiomyosarcoma arising 34 years after the irradiation of the left thigh. This is a younger case with an advanced leiomyosarcoma, we speculate previous radiation might contribute the carcinogenesis. After a short-lived initial remission following surgery and chemotherapy, the patient deteriorated because of tumor recurrence and died 20 months after the treatment.

The low incidence rate of the primary leiomyosarcoma of the external genitalia makes it difficult in managing such case. Fagunde et al suggested that adjuvant radiation therapy might reduce the rate of recurrence.⁵ The role of adjuvant chemotherapy was observed as effective in pediatric rhabdomyosarcoma⁵ but not in the leiomyosarcoma.⁷ Retroperitoneal or regional lymph node dissection is not recommended.⁸ The prognosis could hardly be predicted from any data or microscopic

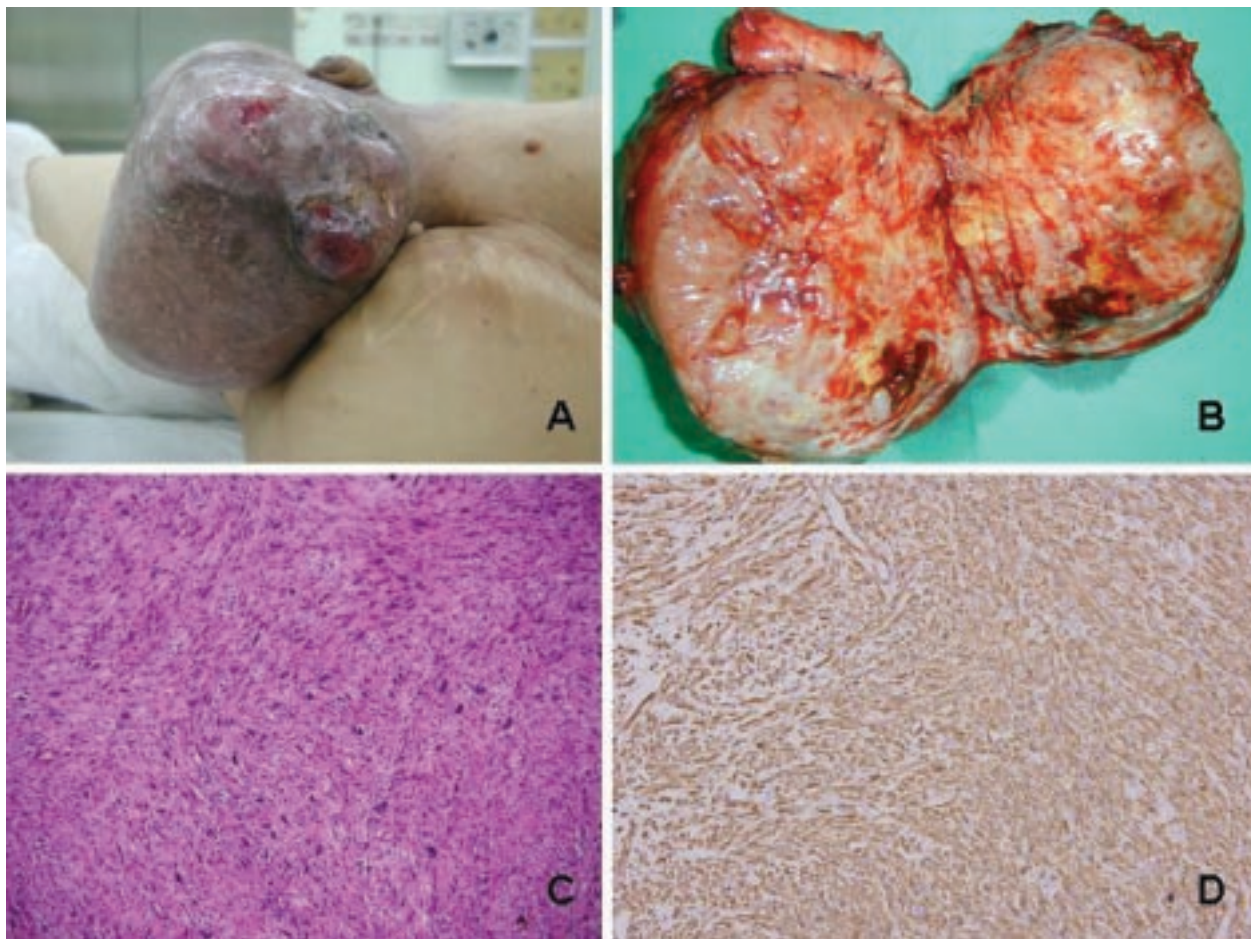


Fig. 1. (A) Enlarged left scrotum with bloody discharge. (B) Gross feature of the tumor is yellowish-white. (C) Hyperchromatic spindle neoplastic cells occasional arranged in interlacing fascicles (haematoxylin and eosin stain; original magnification 100 \times). (D) Tumor cells positive for smooth muscle actin staining (ABC method; original magnification 200 \times)

picture. The malignant behavior might be correlated with the increased mitotic rate microscopically and Farrell et al thought the variation of survival rate may be explained by different degrees of the malignancy.⁷ Although the number of reported cases is very few, radical orchiectomy followed by surveillance appears to be the treatment of choice for paratesticular leiomyosarcoma. There was rare consensus on the adjuvant therapy because no documented follow-up data of a case series with large case number.

In summary, irradiation-associated paratesticular leiomyosarcoma is a rare complication of radiotherapy with a detrimental outcome.

REFERENCES

1. Varzaneh, F. E., Verghese, M., and Shmookler, B. M.: Paratesticular leiomyosarcoma in an elderly man. *Urology* 2002;60:1112.
2. Froehner, M., Fischer, R., Leike, S. et al: Intratesticular leiomyosarcoma in a young man after high dose doping with Oral-Turinabol: a case report. *Cancer* 1999;86: 1571-5.
3. Fuchs, B., Valenzuela, R. G., Petersen, I. A. et al: Ewing's sarcoma and the development of secondary malignancies. *Clin Orthop Relat Res* 2003;415:82-9.
4. Cahen WG, Woodard HQ, Higginbotham NL, et al: Sarcoma arising in irradiated bone: a report of eleven cases. *Cancer* 1:3-29,1948.
5. Fagundes, M. A., Zietman, A. L., Althausen, A. F. et al: The management of spermatic cord sarcoma. *Cancer* 1996;77:1873-6.
6. de Vries, J. D.: Paratesticular rhabdomyosarcoma. *World J Urol* 1995;13:219-25.
7. Catton, C. N., Cummings, B. J., Fornasier, V. et al: Adult paratesticular sarcomas: a review of 21 cases. *J Urol* 1991; 46:342-5.
8. Farrell, M. A. and Donnelly, B. J.: Malignant smooth muscle tumors of the epididymis. *J Urol* 1980;124:151-3.