Abstract. Primary leiomyosarcoma of the testis is an extremely rare disease entity of the genito-urinary tract. Although its clinical presentation does not seem to differ from that of other testicular malignancies, the clinical stage with which patients have been treated, as reported in contemporary literature, has always been stage I. Diagnosis is achieved by combining histologic and immunohistochemical findings. Histologic findings refer to the presence of a spindle cell component with nuclear pleomorphism, while immunohistochemical findings refer to its reaction to specific antibodies. Although the number of reported cases is not significant and the clinical and biological behaviour of these tumors are very hard to predict, we demonstrate that radical orchidectomy followed by surveillance appears to be the treatment of choice. Retroperitoneal lymphadenectomy, radiotherapy and chemotherapy do not seem to have any place in the treatment of this type of malignancy.

Primary leiomyosarcoma of the testis is an extremely rare disease entity. Because of its rarity, standard therapy for this condition is difficult to be recommended. Although the number of reported cases is not significant and the clinical and biological behaviour of these tumors is very hard to predict, we demonstrate that radical orchidectomy followed by surveillance appears to be the treatment of choice. Retroperitoneal lymphadenectomy, radiotherapy and chemotherapy do not seem to have any place in the treatment of this disease (1, 2).

Case Report

In 2006, a 73-year-old patient presented to our Department with a one-month history of an enlarging lump in his right testicle causing mild discomfort. The patient had undergone 40 years ago a right hydrocelectomy. He did not present with any other constitutional symptoms such as voiding complaints, weight loss, fatigue or fever. Physical examination did not reveal any superficial lymph node swelling but confirmed a hard, non-tender, right testicular mass. No ulceration of the overlying skin was evident. The digital rectal examination revealed a benign enlarged prostate. Scrotal ultrasonographic examination revealed a solid homogenous echo poor mass of the right testicle measuring 35 mm × 33 mm, consistent with the radiographic appearance of a testicular tumour. The left testicle and both epididymides were unremarkable and there was no evidence of hydrocele or paratesticular pathology. Chest, abdominal and pelvic computed tomography scans revealed no evidence of metastatic disease or lymphadenopathy. Liver function and tumour markers, including a fetoprotein, lactate dehydrogenase, and beta human chorionic gonadotrophin assays, were all within normal ranges. Blood examination revealed a low white blood cell count of 10.9/ nl and C-reactive protein of 0.4 mg/dL.

Right inguinal radical orchietomy with high ligation of the spermatic cord was performed without complication. On gross examination, a transverse opening of the testis showed a well-defined mass, which measured 3.5 by 3.0 by 2.5 cm in the greatest dimension. The tumor was yellowish-white, solid and encapsulated (Figure 1). The tumor existed only in the testis, did not involve the spermatic cord, epididymis or tunica vaginalis and the surgical margins were negative.

Histologically, the tumour was a spindle cell neoplasm with moderate nuclear pleomorphism (Figure 2). Immunohistochemical stains, which included vimentin, desmin, smooth muscle actin, CD34, S100, CD-68 and HMB-45, were performed. Immunohistochemistry revealed positive staining for vimentin, smooth muscle actin and desmin, while S100, CD34, CD-68, HMB-45 were negative. The combined histologic and immunohistochemical findings were diagnostic of primary poorly differentiated leiomyosarcoma of the testis. The patient had an uneventful postoperative course and received no adjuvant therapy. The tumor follow-up, which included computed tomography scan of the abdomen, pelvis, bone, and chest, has remained normal, and the patient has remained free of disease these last 28 months.

Primary Leiomyosarcoma of the Testis. A Case Report

APOSTOLOS P. LABANARIS1, VAHUDIN ZUGOR2, ROBERT SMISZEK1, REINHOLD NÜTZEL1 and REINHARD KÜHN1

1Department of Urology, Martha Maria Medical Center, Nürnberg, Germany; 2Department of Urology, St. Antonius Hospital, Gronau, Germany

Correspondence to: Apostolos P. Labanaris, MD, Ph.D., Department of Urology, Martha Maria Medical Center, Stadtenstraße 58, 90491 Nürnberg, Germany. Tel: +49 9119591351, Fax: +49 9119591352, e-mail: labanaris@web.de

Key Words: Primary leiomyosarcoma of the testis, diagnosis, therapy, prognosis.
Discussion

Leiomyosarcoma is a rare disease entity not often encountered in the genito-urinary system. When presented, 80% arise from the soft tissue of the spermatic cord and 20% originate from the epididymis or darts of the scrotum (3). Although approximately 100 paratesticular leiomyosarcomas have been reported in the literature (4), fewer than 10 cases of primary leiomyosarcoma of the testis have been mentioned (2, 5-10).

Primary leiomyosarcoma of the testis mostly occurs in young men in whom there is an associated history of high-doses of anabolic steroids (5) or chronic inflammation (6). The clinical presentation of this tumor does not seem to differ from other testicular malignancies and the clinical stage has been always stage 1 in all reported cases. The reason why testicular leiomyosarcoma is diagnosed as a local disease is probably due to the fact that it is easily recognized and tends to be slow-growing (10).

Leiomyosarcoma is a soft-tissue tumor that arises from smooth muscle cells of mesenchymal origin. Its origin has been attributed to contractile cells in the tunica propria of the seminiferous tubules, to the muscular layer of blood vessels, and to smooth muscle elements in the tunica albuginea (2). The hormonal stimulation of the proliferation of smooth muscle cells has been suggested to have a role in the carcinogenesis of leiomyosarcoma (5).

The clinical and biological behavior of these tumors is very hard to predict; however, the high mitotic activity is considered an important criteria for malignancy. Histologically, this type of tumor appears as malignant smooth muscle spindle cells with typical nuclei that selectively stain with antibodies to smooth muscle actin and desmin vimentin, but not to S100, CD34, CD-68 and HMB-45.

Although the number of reported cases is not significant, standard therapy for this condition is difficult to recommend. Radical orchidectomy followed by surveillance appears to be the treatment of choice for primary leiomyosarcoma of the testis. Retroperitoneal lymphadenectomy is not recommended since retroperitoneal lymph node involvement is rare, even in the presence of widespread metastatic disease (1). Additionally, leiomyosarcomas seem to be radio-resistant as well as chemoresistant (3).

References


Received August 19, 2009
Revised March 12, 2010
Accepted March 18, 2010