Paratesticular Leiomyosarcoma — A Case Report

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Paratesticular leiomyosarcoma is very rare. We report a 72-year-old man with this scrotal tumor. The mass had been painless for eight years, but became painful and larger recently. A non-tender, firm, elastic mass was palpable on the right hemiscrotal wall, measuring 6 centimeters in diameter. The mass was not adhering to the testis or to the vas deferens. We performed a wide excision of the mass. The pathological diagnosis was compatible with a grade-2 leiomyosarcoma. The patient refused postoperative radiotherapy, of which the role remains controversial. He was doing well at nine months of follow-up. Local recurrence and/or distant metastasis have been reported for paratesticular leiomyosarcoma, though this rare tumor is often mistaken as a benign tumor. Long-term follow-up is therefore necessary for this case. (JTUA 19:122-4, 2008)

Key words: leiomyosarcoma, dartos muscle, scrotum, skin.

INTRODUCTION

Paratesticular leiomyosarcoma is very rare. The tumor usually presents with a painless, slow-growing scrotal mass in middle-aged or older men. The diagnosis is always based on histological examination. Complete and radical local excision with a tumor-free margin is necessary to achieve the best outcome. Local recurrence and/or distant metastasis with dismal prognosis have been reported,¹ though the tumors are often mistaken as benign tumors. Long-term follow-up is necessary for these patients. We report on a case of paratesticular leiomyosarcoma.

CASE REPORT

The 72-year-old man had been aware of a painless scrotal tumor for about eight years. The tumor was becoming larger and painful over the past three months before he visited our clinic. The man had a history of hypertension but no other medical diseases. Physical examination revealed a non-tender, solid mass on the right hemiscrotal wall, measuring 6 centimeters in diameter. There was no inguinal lymphadenopathy. We performed a wide local excision of the mass under general anesthesia. Grossly, the tumor was pinkish, oval-shaped with irregular surface, and elastic. It was mobile under the scrotal skin and not adhering to the testis or to the vas deferens. The pathological diagnosis was compatible with a paratesticular leiomyosarcoma. The tumor was composed of highly cellular fascicles of spindle-shaped cells with cigar-shaped nuclei and eosinophilic cytoplasm. Some bizarre giant tumor cells with prominent pleomorphic nuclei were present. And the mitotic count was high (about 20 mitoses/10 HPF) with focal necrosis (Fig. 1), suggestive of a grade-2 tumor accord-

Fig. 1. Section through the tumor showing characteristics of leiomyosarcoma, such as interlacing bundles of spindle-shaped cells with eosinophilic cytoplasm and cigar-shaped nuclei (black arrow), bizarre giant tumor cells with prominent pleomorphic nuclei and eosinophilic cytoplasm (white arrow), and several mitoses per high power field. × 40.
paratesticular leiomyosarcomas. Immuno-histochemical studies showed the tumor stained positive for actin and desmin, but negative for CD-34 and S-100. Postoperative abdominal computed tomography showed no obvious intra-abdominal lesions. We proposed postoperative radiotherapy, but the patient refused. The patient was doing well at nine months of follow-up, with no signs of local recurrence or distant metastasis.

DISCUSSION

Leiomyosarcoma is one type of smooth muscle neoplasms. Smooth muscle of skin area is present only in the walls of blood vessels, along hair follicles (arrectores pilorum), and the dartos muscle in the scrotum. There are two types of leiomyosarcomas, cutaneous leiomyosarcomas arising from the dartos muscle or arrectores pilorum, and subcutaneous leiomyosarcomas arising from the muscle lining of arterioles and veins in the subcutaneous tissue. Leiomyosarcomas of the scrotal wall usually arise from the dartos muscle, but it is difficult to differentiate the tumor from other leiomyosarcomas of the skin by histology alone. This case could be either a case of cutaneous leiomyosarcoma arising from the dartos muscle or a subcutaneous superficial leiomyosarcoma. Up to 2004, only less than 40 cases of leiomyosarcomas arising from the dartos muscle had been reported.

There are three typical histological features of leiomyosarcomas, including perpendicularly arranged fascicles of spindle cells with eosinophilic cytoplasm, hyperchromatic blunt-ended nuclei, and scattered paranuclear vacuoles. On immunohistochemical staining, most leiomyosarcoma express smooth muscle actin, muscle-specific actin, and desmin; while CD-34, myogenin, Ki-67, S-100 protein, and cytokeratin had also been reported in some cases. This case was positive only for actin and desmin.

The etiology of leiomyosarcomas remains unclear, though some authors suggested local irradiation at childhood could be a potential cause. Clinically most paratesticular leiomyosarcomas present in men of middle or older age as painless, slow-growing scrotal tumors. The diagnosis is always based on histological examination. Other rare tumors, including benign leiomyoma, fibrous mesothelioma, various benign fibrous tumors and pseudotumors, and fibromatosis, should be considered in the differential diagnosis of paratesticular leiomyosarcomas.

Paratesticular leiomyosarcomas are often mistaken as benign tumors preoperatively. The prognosis is usually good if without local recurrence, though a local recurrence rate of 40% has been reported. Three prognostic factors have been proposed: the presence of a tumor-free margin, the subtype, and the grades of the tumors. A positive margin at initial excision greatly increased the risk of local recurrence. Therefore, a complete and radical local excision with a tumor-free margin is necessary. Postoperative radiation therapy with or without chemotherapy have been reported in some cases, but the benefits remain inconclusive due to small number of cases. Regional lymphadenectomy is indicated for those with clinically suspected or proven lymphatic spread. Distal metastasis to the lungs, liver, bone, or lymph nodes has also been reported. Though still controversial, it is generally accepted that distant metastasis is more common for leiomyosarcomas of subcutaneous type than for cutaneous type. According to the NCI system, leiomyosarcomas were classified into three grades according to the degree of mitoses, pleomorphism of nuclei, and focal necrosis. Paratesticular leiomyosarcomas are mostly of low-grade tumors, but high-grade tumors may behave more aggressively. In the large series of 24 cases of paratesticular leiomyosarcomas, none of the cases with grade-1 or grade-2 tumors was dead of disease during the follow-up period. Therefore, long-term follow-up is necessary for patients with this rare tumor.

REFERENCES

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