A case of rhabdomyosarcoma of the vagina in an elderly woman

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Summary

Most rhabdomyosarcomas of the vagina (RMSV) occur in infants and children up to six years old. RMSV in elderly patients is extremely rare. We report a case of a 70-year-old woman with RMSV. She had received surgery for uterine endometrial cancer one year before and a vaginal polypoid tumor was noted during routine follow-up vaginal examination. She was referred to our department for radiation therapy following partial tumorectomy of the lesion. She was given three sessions of intra-vaginal radiation therapy, once a week with 6 Gy at 7.5 mm below the vaginal surface and external irradiation of 50 Gy to the pelvis. However, paraaortal lymph node metastasis developed during initial radiation therapy. Furthermore, multiple bone metastases appeared at the completion of the radiation therapy. Six months after initial treatment the patient died from progression of the disease. Autopsy demonstrated small residual tumor at the primary site as well as multiple systemic metastases.

Key words: Rhabdomyosarcoma, Vagina, Elderly woman, Radiation therapy.

Introduction

Rhabdomyosarcoma of the genitourinary tract in children is a relatively common disease [1]. Most rhabdomyosarcomas of the vagina (RMSV) occur in infants and young children. RMSV in elderly patients is extremely rare [2] and only a few cases have been reported in patients over 50 years old [2, 3]. The treatment of RMSV in children has been almost standardized. Chemotherapy plays an important role because RMSVs are particularly susceptible to chemotherapy. The Intergroup Rhabdomyosarcoma Study (IRS) report (study III) reported that 17 of 20 RMSV group III patients, who were mainly treated with chemotherapy, had no evidence of disease [4]. However, the treatment for RMSV in older patients is not standardized because of its rarity. We report a primary RMSV with rapid progression in a 70-year-old woman who was treated with radiation therapy alone and discuss the difficulty of treatment.

Case

A 70-year-old woman diagnosed with endometrial cancer of the uterus who had undergone total hysterectomy and bilateral salpingo-oophorectomy one year before was diagnosed with a vaginal polypoid tumor on routine vaginal examination. The vaginal examination revealed a bulky polypoid mass bleeding within the vagina. Pelvic computerized tomography demonstrated left iliac lymphadenopathy as well as a vaginal mass (Figure 1). Partial tumorectomy was subsequently performed for histological diagnosis. The tissue specimen showed highly anaplastic small round cells (Figure 2). Immunohistochemical staining of the specimen was strongly positive for desmin (Figure 3), and negative for cytokeratin and LCA. Accordingly, the histological diagnosis was a mixed-type rhabdomyosarcoma. Thus the patient was classified as group III according to the IRS grouping system [5]. Surgery and chemotherapy were not indicated because of her poor general health and hematological condition. Hence, she was referred to our department for radiation therapy. She was given three sessions of intravaginal radiation therapy, once a week with 6 Gy at 7.5 mm below the vaginal surface and external irradiation of 50 Gy over 25 fractions to the pelvis (with 30 Gy over 15 fractions for the whole pelvis and with 20 Gy over 10 fractions for the center shielded pelvic field). Unfortunately, paraaortal lymph node metastases developed during the initial radiation therapy. She was given external irradiation of 50 Gy over 25 fractions to the paraaortal lymph nodes. In addition, multiple bone metastases appeared following radiation therapy (Figure 4). Her general condition

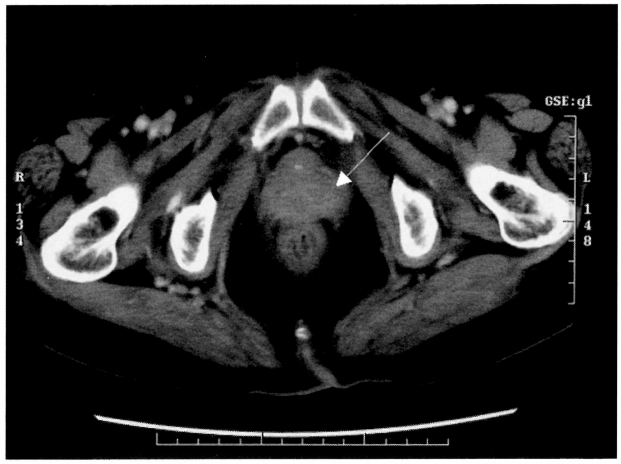

Figure 1. — Computed tomography of the pelvis showing an enhanced tumor at the vagina.

Revised manuscript accepted for publication March 3, 2004

Fig. 2

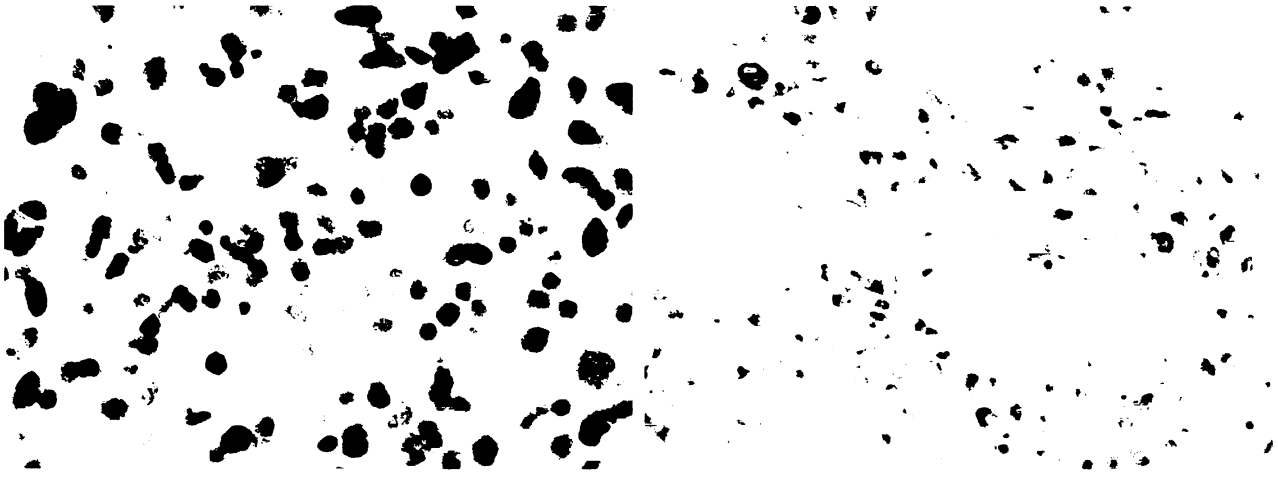

Figure 2. — Hematoxylin and eosin staining of the vaginal tumor (x 400) revealing highly anaplastic small round cells.
Figure 3. — Desmin immunohistochemical staining of the vaginal tumor (x 400). Most of the tumor cells are positive for desmin (densely stained lesions).

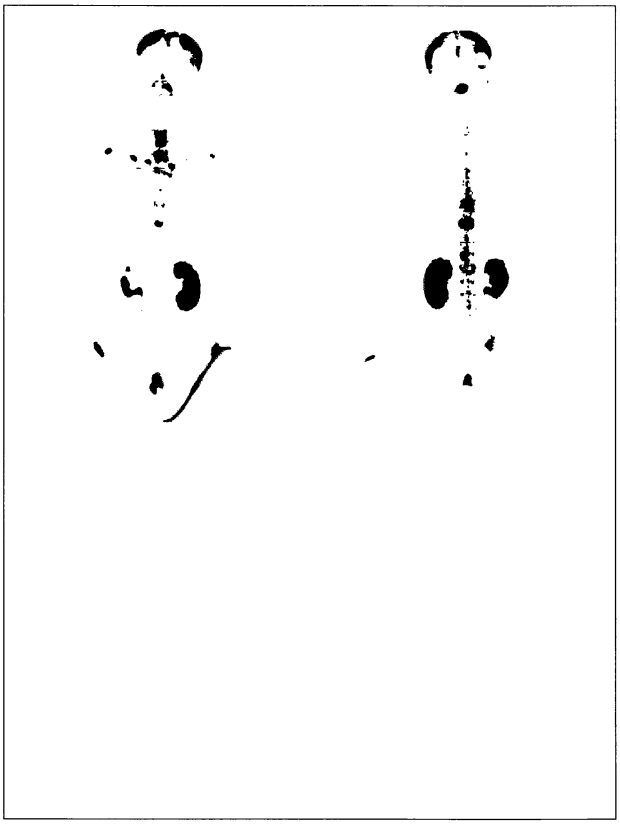

Figure 4. — ^{99m}Tc bone scintigraphy showing multiple bone metastases to the skull, spine, femoral bone and ribs.

rapidly worsened and she died of disease progression six months after initial treatment. The autopsy revealed a small residual tumor in the vagina and multiple metastases to the lung, liver, paraaortal lymph nodes and bone. These were histologically confirmed as rhabdomyosarcoma. However, part of the paraaortal lymph nodes involved metastatic tumors of uterine endometrial cancer.

Discussion

In general, accurate histologic diagnosis of rhabdomyosarcoma is difficult because the existence of rhabdomyoblasts strongly suggests rhabdomyosarcoma, however, the probability of the appearance of rhabdomyoblasts is not so high. Therefore, immunohistochemical staining with cytokeratin, S-100, vimentine, leukocyte common antigen and desmin is generally used to distinguish between rhabdomyosarcoma and other malignant tumors. Desmin is a particularly reliable marker that distinguishes between RMS and melanoma because it is more sensitive for poorly differentiated rhabdomyosarcoma than the other tumor markers [6]. In this case, both the primary tumor and metastatic lesions were positive for desmin, and negative for cytokeratin and LCA. Given these results, the patient was diagnosed with rhabdomyosarcoma.

The treatment of RMSV in children has been essentially standardized in recent years. Chemotherapy plays a central role in pediatric RMSV whereas radiation therapy is utilized under certain circumstances. The IRS study (III) reported that 17 of 20 RMSV group III patients who were primarily treated with chemotherapy had no evidence of disease up to 66 months [4].

However, because of its rarity, the treatment for RMSV in adult patients is not standardized. Hilgers *et al.* reported that of three adult RMSV patients who had not received chemotherapy, all died within a year after initial treatment. Two of the three who were treated with radiation therapy alone died within six months [3]. In the present case, chemotherapy was contraindicated given the patient's poor hematological and general condition, as well as rapid disease progression. As a result, she was treated with radiation therapy alone and died of recurrence only six months after radiation therapy. Shy *et al.* reported in their literature review that all nine vaginal rhabdomyosarcomas in patients over 20 years old, which

were treated with combined therapy including chemotherapy, lived for more than 20 months [2]. These findings, including the present case, suggest that chemotherapy is necessary for adult RMSV patients, as well as for infants, to improve the prognosis of the disease. In order for future clinical investigations to improve the prognosis of the disease in adults, the indications for chemotherapy, the optimal quantity of anti-cancer drugs and the number of treatment cycles has to be determined. Taking into account the rare incidence of adult RMSV, a multi-center study would be required to establish the optimal chemotherapy for adult RMSV.

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