Pleomorphic rhabdomyosarcoma of the uterine corpus: a case report

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Case Report

A 72-year-old woman presented with metrorrhagia of four months duration and weakness. Menopause occurred at age 49 and the patient did not receive any substitutive hormonal treatment. Physical examination demonstrated uterine enlargement. Ultrasonography and magnetic resonance imaging revealed a mass entirely filling the uterine cavity with extension to the cervix. Histology of the endometrial curettage specimen showed a high grade sarcoma. A total hysterectomy was performed with bilateral pelvic lymphadenectomy. The uterine tumour was a soft whitish-grey, 13 cm in maximal diameter, focally necrotic and polypoid mass filling the uterine cavity with extensive myometrial invasion. Histology revealed ill-defined aggregates of round or pleomorphic cells and short bundles of spindle-shaped cells with focal storiform arrangements. These cells had ovoid vesicular nuclei with distinct nucleoli and abundant eosinophilic, fibrillary or vascuolated cytoplasm (Figure 1). No cross-striations were identified on hematoxylin-eosin-saffron (HES) staining, but numerous tadpole or strap-like cells were highly suggestive of rhabdomyoblastic differentiation. The average mitotic count was 70 per 10 high power fields. There were no other homologous or heterologous elements. A few residual cystic endometrial glands were entrapped superficially. The tumour involved the uterine wall up to the serosal membrane and extended to the cervix and parameters. The ovaries and several pelvic lymph nodes were metastatic. The tumour cells stained positive for desmin (Eurodiagnostic, 1:100), muscle-specific actin (HHF35) (Biogenex, 1:100), alpha-sarcomeric actin (Sigma, 1:800) and were negative for alpha-smooth muscle actin (Sigma, 1:15000) and S-100 protein (Dako, 1:400). The more undifferentiated round cells showed nuclear positivity for MyoD1 (Tebu, 1:20) (Figure 2).

After surgery, the patient received pelvic external radiation therapy (45 Gy) and curietherapy (15 Gy). She was alive without any evidence of recurrence five months later.

Discussion

Pleomorphic rhabdomyosarcoma of the uterine corpus is a very uncommon neoplasm. Most of the cases previously reported have been in fact carcinosarcoma with a rhabdomyoblastic component [1, 2]. These tumours especially affect postmenopausal women and are discovered by abnormal uterine bleeding or hypogastric pain [1, 2]. Pelvic irradiation and tamoxifen therapy are rarely found in the past medical history of the patients [3, 4]. At gross examination, these tumours typically form a polypoid mass in the uterine cavity protuding sometimes through the cervix [1, 2]. Two conditions are required to make the diagnosis of pure uterine rhabdomyosarcoma: exclusion of any epithelial component after extensive sampling of the tumour [1, 4] and demonstration of rhabdomyosarcomatous tissue without other sarcomatous components [1]. Immunohistochemistry is of major interest in diagnosis: skeletal muscle differentiation is proven by staining with alpha-sarcomeric actin, myoglobin, myogenin or MyoD1 antibodies [1, 5]. Desmin and muscle-specific actin are less specific because they can be expressed both in leiomyosarcoma and stromal neoplasms [1, 6]. Differential diagnoses include carcinosarcoma, adenosarcoma with sarcomatous overgrowth, homologous sarcoma (leiomyosarcoma, stromal sarcoma) and embryonal rhabdomyosarcoma [1, 3, 7-9]. For embryonal rhabdomyosarcoma, no distinction can be made with immunohistochemistry. Unlike pleomorphic rhabdomyosarcoma, its background is more edematous, rhabdomyoblasts are less pleomorphic and cross-striations are present [1, 9]. Moreover, it occurs mainly during infancy [1].

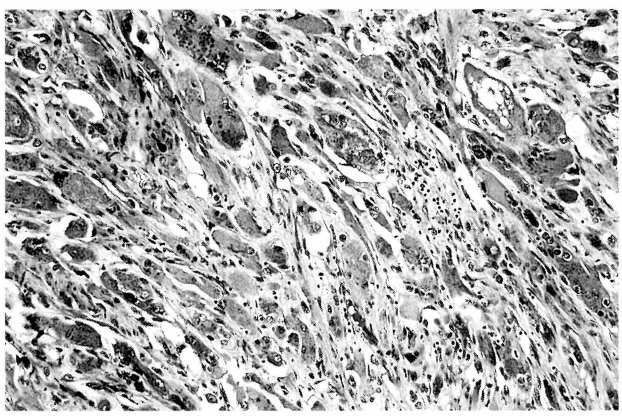

Figure 1. — Disorderly arrangement of round and pleomorphic cells (Hematoxylin-Eosin-Saffron, 100x).

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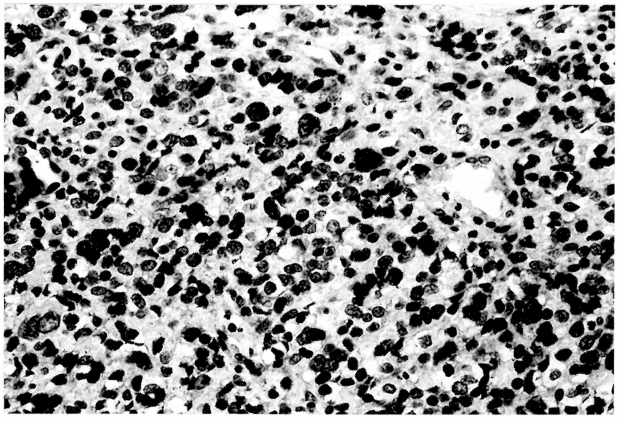

Figure 2. — Expression of MyoD1 in rhabdomyoblasts: nuclear staining (125x).

Histogenesis remains controversial: pleomorphic rhabdomyosarcoma might derive either from primitive mesenchymal cells or represent complete overgrowth of carcinosarcoma [1, 3]. Pleomorphic rhabdomyosarcoma is a highly malignant tumour with early relapse and vaginal, lymph node or pulmonary metastases [1, 2]. Prognosis is extremely poor. Nearly all patients die within two years after initial diagnosis, despite adjuvant radiotherapy and/or chemotherapy [1].

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