Case report

Cytogenetic study of botryoid rhabdomyosarcoma of the uterine cervix

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Summary. We report a case of sarcoma botryoides of the uterine cervix occurring in a 19-year-old woman. By light microscopy the tumor showed round and spindle cells with hyperchromatic nuclei and, focally, a cambium layer subjacent to the surface epithelium and surrounding endocervical glands. Strap-shaped cells with and without cross-striations and small foci of immature cartilage were also present. Immunohistochemical studies showed positive staining within the tumor cells for myoglobin, desmin, vimentin, muscle-specific actin and CD56. By electron microscopy, tumor cells showed cytoplasmic filaments in an alternating pattern of thick and thin filaments. Chromosomal analysis demonstrated deletion of the short arm of chromosome 1, and trisomies 13 and 18. To our knowledge, this is the first reported case of sarcoma botryoides of the endocervix with chromosomal analysis.

Key words: Botryoid rhabdomyosarcoma – Uterine cervix – Immunohistochemistry – Ultrastructure – Chromosomal analysis

Introduction

Sarcoma botryoides, a subtype of embryonal rhabdomyosarcoma, affects the uterine cervix in young women. It is rare and few cases have been reported in the literature (Montag et al. 1986; Brand et al. 1987; Daya and Scully 1988). Like their counterparts which occur in the vagina of infants, these tumors are characteristically composed of polypoid masses resembling bunches of grapes. Microscopically they show a cambium layer subjacent to the surface epithelium, and primitive rhabdomyoblasts including strap-shaped cells with cross striations.

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To our knowledge, the botryoid type of embryonal rhabdomyosarcoma has not been characterized cytogenetically, while cytogenetic analysis of alveolar rhabdomyosarcoma (AR) has demonstrated a characteristic translocation t(2; 13) in the majority of cases (Turc-Carel et al. 1986; Douglass et al. 1987). Embryonal rhabdomyosarcoma (ER) shows various chromosomal changes but lacks the t(2; 13) which appears to be restricted to the alveolar type (Wang-Wuu et al. 1988).

In this report, we have characterized sarcoma botryoides using immunohistochemistry and electron microscopy. We also describe the cytogenetic findings from cultured tumor cells. Several chromosomal alterations were seen including deletion of the short arm of chromosome 1, and trisomies 13 and 18. There was no evidence of the t(2; 13) translocation observed in alveolar rhabdomyosarcoma.

Case report

The patient, a 19-year-old (gravida I; para O) black female, presented with menorrhagia over several weeks. Her past history was significant for a curettage for an incomplete abortion. Family history revealed a sister diagnosed with ovarian carcinoma at the age of 22 years. Physical examination was remarkable only for a large 2 x 2 cm, friable, necrotic cervical mass. A biopsy demonstrated sarcoma botryoides. A computed tomography scan showed no adenopathy or intra-abdominal disease. A magnetic resonance imaging scan revealed an exophytic, enhancing mass arising from the inferior surface of the cervix. Cystoscopy, proctoscopy and examination under anesthesia confirmed stage IB disease. The clinical impression was that cervical conization would result in inadequate resection, and the patient underwent total abdominal hysterectomy and pelvic lymphadenectomy. She is currently receiving vincristine, actinomycin D and cyclophosphamide chemotherapy.

Pathology

The uterus weighed 72 g. On gross examination, there was an endocervical pedunculated polypoid mass protruding through the cervical os and firmly attached to
Fig. 1. Squamous cervical epithelium overlying hypercellular areas with a focally prominent cambium layer composed of spindle cells. H&E, ×160

Fig. 2. Rhabdomyoblasts showing ample cytoplasm, some with cross striations. H&E, ×200

Fig. 3. Immunoperoxidase stain for myoglobin showing cytoplasmic reactivity in neoplastic cells. ×200

the endocervical wall. The mass measured 3.0 × 1.8 cm and was soft, focally ulcerated and hemorrhagic. The macroscopic examination of the uterus was otherwise unremarkable. By light microscopy, the tumor was covered by benign stratified squamous epithelium and normal endocervical lining epithelium, except where ulcerated. Focally evidence of a cambium layer was seen, composed of spindle cells with an edematous stroma underneath the surface epithelium (Fig. 1), but the cambium layer was best seen around endocervical glands and blood vessels. Tumor cells were seen between entrapped endocervical glands and occasional foci of immature cartilage were present. The tumor cells were round with scanty cytoplasm. In places strap-shaped cells with eosinophilic cytoplasm and cross striations were also identified (Fig. 2). The mitotic count in the more cellular areas ranged from two to five per ten high-power fields. Metastatic tumor was present in one of five iliac lymph nodes excised. Immunohistochemical studies using the standard three-step indirect avidin-biotin-peroxidase method (Hsu et al., 1981) showed that the tumor cells stained positively for myoglobin (Fig. 3), desmin (Dakopatts, Carpinteria, Calif., USA), muscle-specific actin (Enzo Diagnostics, New York, N.Y., USA), and CD56 (Becton-Dickinson, San Jose, Calif., USA). The cells were negative for cytokeratin (Becton-Dickinson), S-100 protein, alpha smooth-muscle actin and neurone-specific enolase (Dakopatts). The vimentin was focally weakly positive. By electron microscopy, cultured tumor cells showed alternating thick and thin filaments and cytoplasmic glycogen granules.

Chromosome analysis

Portions of the tumor and normal myometrium were placed in a sterile culture medium for chromosomal stu-