Endodermal Sinus Tumor of the Vagina - a rare entity treated exclusively with Chemotherapy

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Introduction

Endodermal Sinus Tumor (EST) is the rarest of the vaginal tumors. It commonly occurs in children of under 3 years and usually presents with vaginal bleeding. EST is most frequently encountered in the ovary. It is exceedingly rare in the vulva. Approximately 50 cases of EST of the vagina or uterine cervix have been reported. The presence of Schiller Duval bodies and various histologic patterns characteristic of EST and immuno-histochemical demonstration of alpha-fetoprotein (AFP) clinch the diagnosis. Assay of serum AFP can potentially aid in the diagnosis, monitor effectiveness of therapy and detect recurrences before clinical manifestations. This case report describes the successful treatment of a child with vaginal EST with chemotherapy exclusively.

Case Report

A sixteen-month-old child presented with vaginal bleeding for one month. She was the seventh child and her mother was in mid-thirties at the time of her birth. Pregnancy and childbirth were uncomplicated and there was no history of diethylstilbestrol use or any other particular therapy during pregnancy.

On examination the child’s general health was satisfactory, except for a mild anaemia (haemoglobin 10.5 g/dl). No mass was palpable on abdominal examination. External genitalia were normal and blood stained vaginal discharge was noticed. Per rectal examination revealed a mass anteriorly almost completely filling up the pelvis. However rectal mucosa felt normal.

On ultrasound examination a 3.4x3.8cms heterogeneous mass posterior to the bladder was visualized. Computerized tomographic scan (CT) of the pelvis revealed a homogenous mass with areas of calcification completely filling up the pelvis and displacing the bladder upwards. There was no invasion of pelvic bones, urinary bladder or rectum. Pre-operative blood biochemistry and chest X-ray were normal.

At laparotomy the uterus, ovaries, fallopian tubes and other pelvic and abdominal organs were normal. A digital and speculum examination of the vagina revealed a friable mass filling the upper half of the vagina and arising from the right fornix posteriorly. Size of the tumor was approximately 4 x 4 cms and seemed to be lifting up the uterus. Biopsy was taken from the tumor, the histology of which was reported to be endodermal sinus tumor of the vagina. Characteristic Schiller Duval bodies were also identified. Serum AFP was elevated to 14321 ng/ml (n 10.0 ng/ml). It was decided not to subject the child to any major surgery in view of good results reported with chemotherapy.

Her postoperative recovery was uneventful. She subsequently received six pulses of combination chemotherapy of cisplatinum 30 mg/m² on days one through five, etoposide 100 mg/m² on days two and three and bleomycin 30 mg/m² on days three and four (PEB) at four weekly intervals. She withstood the chemotherapy very well. Serum AFP gradually returned to normal levels after six pulses of initiation of chemotherapy. Complications included alopecia and multiple boils on skin.

CT scan after four pulses of chemotherapy demonstrated a significant reduction in the size of the tumor to 1.8 x 1.7 cms. Serum AFP at that point was 22.6 mg/ml. CT scan was repeated six months after
chemotherapy and the mass was now 1.1 x 1.3 cms. However, the serum AFP was 2.5 ng/ml. As the serum AFP remained within normal limits CT scan was not repeated.

It has been five years since the initial diagnosis. The child was last seen in June 2000. She is growing well and has never had another episode of vaginal bleeding. At repeated general, per rectal and ultrasound examinations, there is no recurrence of the tumor and serum AFP has remained within normal limits.

Discussion

As with all rare disorders the ideal management of EST of the vagina in infancy remains an enigma. The natural history of this malignancy is not clearly understood. Untreated patients die within two to four months of presentation.

Treatment of infantile EST has evolved over this century. Before 1965 local therapeutic modalities, radical surgery and or irradiation were employed with universal failure. Vawter introduced the use of chemotherapy for EST in his report of two patients with recurrent and metastatic disease after surgery. Chemotherapy subsequently appeared as an integral part in most treatments reported since 1970.

Surgery employed for EST ranging from vaginectomy to total pelvic exenteration produces loss of sexual and reproductive functions and possible loss of bladder and rectum as well. Long term radiation causes castration, abnormal growth of pelvic bones, aseptic necrosis of femoral heads, destruction of bone marrow and primary malignancy in the irradiated field. On the other hand complications of chemotherapy are usually transient and do not result in permanent morbidity. There has been a trend to avoid mutilating primary surgery, particularly as multiagent chemotherapy has given equally good if not better results. Therefore, our patient was exclusively treated with combination chemotherapy without resorting to extirpative surgery. Only one case of vaginal EST has been reported to be treated previously using bleomycin, vinblastin and cisplatinum with exclusive chemotherapy. In another case primary chemotherapy (vincristin, actinomycin-D and cyclophosphamide VAC) was followed by partial vaginectomy.

In our patient during chemotherapy the mass reduced in size with simultaneous fall in serum AFP levels. However even with a normal serum AFP, a 1.3x1.3 cms mass was still visible on CT Scan. It was decided not to proceed with excision of the mass as it probably represented scar tissue. Biopsies of such masses after chemotherapy have revealed scar tissue, degenerating and necrotic cells.

Besides our patient, only ten patients with vaginal EST have been reported to survive beyond 5 years.

EST of the vagina appears to be sensitive to the same combination chemotherapy that are used for males with disseminated testicular germinal cancer and females with EST of the ovary. However little is known of long term sequelae of such treatment. There are concerns of possible ovarian failure or oncogenic effects. Cyclophosphamide has been reported to produce ovarian failure in post-pubertal females, but when used in pre-pubertal females normal pituitary-ovarian function and even pregnancy has been reported. More extensive experience with chemotherapeutic treatment of this rare tumor is needed to adequately assess long-term survival and prognosis and the most efficacious chemotherapeutic regimen.

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References