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Endodermal Sinus Tumor in a 14-Year-Old Girl

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A case report on a 14-year-old girl with a large pelvic and abdominal mass is presented. Initial evaluation suggested the mass to be of ovarian origin, and the diagnosis of endodermal sinus tumor was confirmed by the pathological specimen. Alpha fetoprotein (AFP) levels on admission were 12,700 mg/dL, but AFP decreased immediately in the postoperative period and has continued to decline toward normal levels over the past four months of combination chemotherapy. After completion of chemotherapy, AFP levels will continue to be monitored and a second-look procedure may be in order. (Henry Ford Hosp Med J 1986;34:132-3)

Endodermal sinus tumor (EST) is the second most frequent germ cell tumor of the ovaries. Once considered very rare, the tumor is now diagnosed with greater frequency. Although review of Henry Ford Hospital medical records revealed only two other cases of EST, this number may be artifactually low due to the changing classification and nomenclature of this tumor.

EST, because of its specific age incidence, may be considered one of the most common, highly malignant ovarian neoplasms of childhood, adolescence, and young adult life (1). EST is a very aggressive tumor and until recently has carried a grave prognosis, with most patients dying within 12 months of initial diagnosis. However, since the introduction of combined chemotherapy in the mid-1960s, the prognosis of this disease has improved. Preliminary reports on the use of combined chemotherapy in EST patients are encouraging, and in some cases the success of combined chemotherapy will bring into question the value of radical surgery and subsequent loss of childbearing ability in those patients where the tumor is confined to one ovary. The two-year survival rate following combination chemotherapy is over 90% for stage I and II and less than 50% for stage III and IV of the disease. A case report of a 14-year-old patient with EST, stage I-A, is discussed herein.

Case Report

A 14-year-old obese female complaining of abdominal pain was found to have a large abdominal mass upon examination by her pediatrician, who referred her to the Department of Gynecology at Henry Ford Hospital. The patient had a history of asymptomatic bronchial asthma. She was gravida 0 and para 0, and her last menstrual period was August 3, 1985. Three months prior to admission, the patient noted a sharp, intermittent abdominal pain that worsened with movement. The pain was located primarily in the left upper quadrant. One month later, she noted an increase in abdominal girth despite her attempts at weight reduction. The patient also complained of nausea, vomiting, and an overall decrease in appetite. At the time of admission she complained of a persistent left upper quadrant pain, which she described as sharp and constant, possibly associated with emesis, and which was not relieved by use of 500 mg acetaminophen. The patient stated that the pain had become progressively worse during the week prior to admission. She

denied any syncopal episodes, chest pain, shortness of breath, diarrhea, constipation, or recent abdominal trauma. Although a urine pregnancy test was negative, an ultrasound test revealed a 22 cm x 17 cm x 12 cm complex abdominal and pelvic mass. The patient was scheduled for exploratory laparotomy following a complete workup.

Medical history and examination

The patient was recently diagnosed as having an allergy to intravenous pyelogram dye. She denied the use of any medication. Surgical history was noncontributory, and family history was unremarkable except for her mother, who had a history of systemic lupus erythematosus. The patient's gynecologic history involved menarch occurring at age 11 and irregular menstrual periods since then, with menstrual periods lasting anywhere from three to 20 days. The patient's last menstrual period started on August 3, 1985, and had continued for approximately 30 days.

Upon physical examination the patient complained of lower back pain, which had been occurring for several months, but denied the presence of urinary symptoms or pedal edema. Examination revealed a well-nourished and well-developed 14-year-old girl in no apparent distress. Her temperature was 37.3 °C; blood pressure 120/80; pulse 100 beats/min; weight 133.7 kg; and height 70.5 in. Head and neck examination was unremarkable. Thoracic examination revealed the lungs to be clear. Cardiovascular examination was normal. The patient's abdomen was positive for bowel sounds and was distended with an obvious mass that was tender to palpation. The mass was irregular, higher on the left than the right side of the abdomen, and measured approximately 36 cm from the top of the superior pubic symphysis. Pelvic examination revealed the cervix to be small and the cul-de-sac apparently free, but the uterine size could not be assessed because of the abdominal distention. There was no evidence of any lymphadenopathy, and the extremities were normal.

Hospital course

On admission, laboratory data were as follows: hemoglobin 8.7 g/dL; hematocrit was 26%; mean corpuscular volume was 75.5 U³; and electrolyte profiles were normal. BUN was 8 g/dL; creatinine was 1.2 mg/dL; glucose 85 mg/dL; and alpha fetoprotein (AFP) was elevated to

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12,700 mg/dL, with normal being 0 to 20 mg/dL. Intravenous pyelogram showed a right hydroureter and hydronephrosis. Chest x-ray and EKG were normal. Barium enema showed an extrinsic compression proximal to and in the midsigmoid area.

On the second day after admission the patient underwent an exploratory laparotomy with a right salpingo-oophorectomy, omentectomy, appendectomy, and biopsy of the left ovary. Two units of packed RBCs were given preoperatively and intraoperatively. The tumor was much larger than described by the ultrasound report. It was mostly solid with areas of thin-walled gelatinous cyst on the left superior border. The tumor was loosely adherent to the omentum, the small bowel, mesentery, and sigmoid. Frozen section of the tumor was consistent with endodermal sinus tumor. The patient's postoperative hemoglobin stabilized at 9.2 g/dL. She was then seen by the Oncology staff, who recommended chemotherapy to start two weeks postoperatively. The chemotherapy recommended was cis-platinum, Velban, and bleomycin. Final pathology was reported as endodermal sinus tumor of the right ovary; all other biopsied areas were negative for tumor, and cytology of peritoneal fluid also was negative.

At the time of discharge, our patient's AFP level had decreased to 8100 mg/dL. On October 25, 1985, she was readmitted for the first course of chemotherapy (Platinol-Velban-bleomycin). AFP was 65 mg/dL. By the second readmission on December 31, 1985, the patient's AFP level had decreased to 3.1 mg/dL. Our patient will continue on this regimen, and a computed tomography scan will be included in the follow-up.

Discussion

Endodermal sinus tumor was first classified by Teilum in 1946, who subsequently published a series of articles describing the extraembryonic germ cell origin of the tumor (2-4), thereby refuting the mesonephric origin proposed by Schiller (5). Since its classification, endodermal sinus tumor also has been known as yolk sac tumor, mesoblastoma vetillinum, and Teilum's tumor. The association of elevated serum AFP and its immunohistological identification in tumor cell has strengthened evidence that suggests the close relationship of the tumor to the yolk sac (6,7). The reported age incidence for EST is 16 months to 46 years, with a peak age incidence of 19 years. Our patient presented with classic symptoms of EST: abdominal swelling from rapid tumor growth, abdominal pain secondary to bleeding into the tumor, anemia, and a palpable abdominal mass.

EST may be pure or mixed. In the mixed variety, other germ cell tumors such as the teratoma or choriocarcinoma may be encountered. If a choriocarcinoma is present, the patient may present with endocrine disturbances from the elevated HCG. EST also exhibits a range of histological patterns that differ considerably from each other. Two or more patterns may be present within the same tumor. The presence of Schiller Duval bodies can be considered diagnostic for EST. This tumor is not radiosensitive, and radical surgery has not improved the prognosis. The operative procedure can be tailored to the extent of the disease and the patient's age. In a young patient with a stage I

disease who has not completed childbearing, a unilateral salpingo-oophorectomy combined with thorough staging biopsies and cytological washings may provide adequate surgical management (8). The combination chemotherapies most frequently used include 1) actinomycin D, vincristine, and cyclophosphamide; 2) actinomycin D, 5-fluorouracil, and cyclophosphamide; and 3) cis-platinum, bleomycin, and vinblastine sulfate.

The major unanswered questions concerning this disease include 1) the duration of therapy, 2) the role of second-look laparotomy, 3) the use of AFP for monitoring, and 4) the effect of chemotherapy on future ovarian function. The duration of therapy, which can vary from six to 24 months, will be dictated by the side-effects of these medications. AFP has been used as an excellent marker for persistence or recurrence of the disease. As ESTs recur within one year in 93% of the patients, treatment should be administered for at least 12 months. Normal menstrual cycles have occurred following chemotherapy, and normal pregnancies also have been reported. Infertility may occur less frequently following chemotherapy with cis-platinum, bleomycin, and vincristine than with other therapies that include the alkylating agents (9).

In summary, EST of the ovaries is a highly malignant ovarian cancer which affects children, adolescents, and young adults. Because of poor results with radical surgery and improving prognosis with combination chemotherapy, patients with EST in the early stages can be offered a more conservative surgical approach. Although the exact answers are not yet available regarding second-look laparotomy, patients who present with initial elevation of serum AFP should have monthly follow-up of AFP levels during chemotherapy and after conclusion of such therapy to rule out possible recurrence of the disease.

References

1. Blaustein A. Pathology of the female genital tract. 2nd Ed. New York: Springer-Verlag, 1982:612-22.
2. Teilum G. Endodermal sinus tumor of the ovary and testes, comparative morphogenesis of the so-called mesonephroma ovarii (Schiller) and extraembryonic (yolk sac) structures of the rat placenta. *Cancer* 1959;12:1092.
3. Teilum G. Classification of endodermal sinus tumor and so-called embryonal carcinomas of the ovaries. *Acta Pathol Microbiol Immunol Scand* 1965;64:407-9.
4. Teilum G. The concept of endodermal sinus (yolk sac) tumor. *Scand J Immunol [Suppl]* 1978;8:75-9.
5. Schiller W. Mesonephroma Ovarii. *Am J Cancer* 1939;35:1.
6. Ballas M. Yolk sac carcinoma of the ovary with alpha fetoprotein in serum and ascitic fluid demonstrated by immunoelectrophoresis. *Am J Clin Pathol* 1972;57:511-6.
7. Itoh T, Shirai T, Maka A. Yolk sac tumor and AFP clinico. Pathological studies of four cases. *Cancer* 1974;65:215.
8. Gershenson DM, Del Junco G, Werson J, Rutledge FN. Endodermal tumor of the ovary: The M.D. Anderson experience. *Obstet Gynecol* 1983;61:194-202.
9. Karlen JR, Kastelic JE. Endodermal sinus tumor of the ovary: An improving prognosis. *Gynecol Oncol* 1980;10:206-16.