CASE REPORT

Endodermal Sinus Tumor of the Vagina in a Child

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Introduction

Malignant germ cell tumors are rare tumors of the childhood accounting for less than 3 % of pediatric malignancies. Endodermal sinus tumor (EST) is the most common histologic subtype of malignant GCT. The vagina is an extremely rare site for GCTs [1]. They are usually diagnosed before 3 years of age. Historically, the approach to therapy has included radical surgical resection with adjuvant irradiation and chemotherapy [2]. We report a case of EST of the vagina which was managed conservatively with chemotherapy alone.

Case Report

A 6-month-old female child was admitted on April 8, 2010, to our hospital with a history of intermittent episodes of bleeding per vaginum for 2 months. The child was apparently healthy until she developed vaginal bleeding. An ultrasound of the pelvis showed a mass of 23 × 20 mm in

the vagina with increased vascularity and a possibility of vaginal polyp or mass. The uterus and cervix appeared normal. On examination, her vitals were stable. An examination under anesthesia was done, which revealed a friable growth from the anterior vaginal wall. The growth bled on touch and tissue was obtained and sent for histopathologic examination to the Norman Institute of Pathology, Christian Medical College, Vellore. She was diagnosed to have EST on histopathologic examination. She was then admitted for further evaluation to the Christian Medical College, Vellore. Her serum alpha-fetoprotein was elevated to 2,410 IU/ml. She was then started on chemotherapy with etoposide, carboplatin, and bleomycin in an attempt to preserve pelvic function. She was given four cycles of chemotherapy, which were completed on July 23, 2010. An MRI of the pelvis done on the August 5, 2010, showed no evidence of residual vaginal, uterine, or other pelvic soft tissue mass lesion. She will be pursued for follow-up with AFP level estimation and CT scan or MRI.

Histology

The biopsy of the tumor mass showed fused papillaroid structures, cords and clusters of polygonal cells with moderate amounts of eosinophilic to clear cytoplasm, and moderately pleomorphic hyperchromatic to vesicular nuclei, many with nucleoli exhibiting mitosis. There were interspersed vascular channels including within the cores of the papillae. There were foci of necrosis and occasional

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foci of glands of columnar epithelium with elongated hyperchromatic nuclei (Fig. 1). Immunohistochemical stains revealed the tumor cells diffusely positive for pancytokeratin (Fig. 2) and focally positive for alpha-fetoprotein. A diagnosis of vaginal yolk sac tumor was made.

Discussion

EST of the vagina is a rare and highly malignant germ cell tumor that primarily occurs in infants. In females, EST is usually encountered in the ovary, either in the pure form or as a component of a mixed germ cell tumor. The clinical presentation includes vaginal bleeding with a polypoidal friable mass. With this clinical presentation, many cases of EST are misdiagnosed as sarcoma botryoides [3]. However, it has a characteristic grape-like appearance and there are edematous and cellular areas formed of immature skeletal muscle cells which help in distinguishing it from an EST. The histopathologic examination confirms the differentiation between the two.

Alpha-fetoprotein level in blood is usually elevated at presentation and should be used as a guide to monitor therapy as well as to detect a recurrence [4]. In our case, examination under anesthesia with vaginal biopsy and elevated alpha-fetoprotein levels confirmed the diagnosis of EST of the vagina.

Management of this vaginal tumor should take into consideration the chemosensitivity of the tumor and preservation of the organ and its function. EST is extremely chemosensitive [5]. The current survival rate for genital malignant GCT is excellent. The 4-year event-free survival rate was 76.2 ± 13.1 % and the 4-year overall survival rate was 91.7 ± 8.4 % in one study [6].

Fig. 1 High power magnification showing papilloroid structures with hyperchromatic nuclei and interspersed vascular channels

Fig. 2 Immunohistochemical stain showing tumor cells positive for pancytokeratin

We emphasize primary chemotherapy with close follow-up of the patient as the treatment of choice to allow preservation of sexual function.

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