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Red alert – Infant vaginal bleeding

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Abstract Infant vaginal bleeding is an alarming symptom in an infant. Although several causes can be listed, the possibility of malignancy still needs to be ruled out in view of the guarded prognosis of these uncommon infantile tumors. This case report aims to raise the awareness towards the workup and management of infantile malignancies in a baby girl.

Key words Yolk sac tumor; endodermal sinus tumor; germ cell tumor; vagina; bleeding; infant.

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INTRODUCTION

Malignant germ cell tumors (MGCTs) account for less than 3% of pediatric malignancies, and yolk sac tumors (YSTs), also called endodermal sinus tumors (ESTs), are the most common histological subtype [1]. In children younger than three years of age at presentation, the primary location of MGCTs is extragonadal (41%) and testicular (27%). The vagina (8%) is a very rare location for MGCTs [1, 2]. There are only fifty odd cases of vaginal ESTs reported in literature. Although rare, vaginal ESTs are both locally aggressive and capable of

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metastasis [3]. Early diagnosis and treatment is very important because of its aggressive nature and positive response to chemotherapy [3]. Here we report a case of a vaginal yolk sac tumor presented to us.

CASE REPORT

Parents brought their eight-month-old female child with a complaint of vaginal bleeding for ten days. Parents denied history of trauma. The baby girl was active and alert on examination. The abdominal examination and the inspection of the perineum were normal. No features of precocious puberty were noted.

Baseline blood tests, including hemogram and urea/creatinine, were within normal limits. The alfa feto protein (AFP) was high, measuring >1000 ng/ml. The hormonal profile of follicle-stimulating hormones, luteinizing hormones, estrogen, and progesterone was normal. Sonography revealed a solid, homogenously hypoechoic, and vascular mass arising from the right ovary measuring 3.4 x 2.8 cm.

After a provisional diagnosis of a functional ovarian tumor, the child was scheduled for surgery. An exploratory laparotomy by Pfannensteil incision revealed cystic bilateral ovaries with a bulky uterus and a soft friable mass of 4 x 3 cm arising from the vaginal fornix, which was sampled. A histopathological examination showed tumor cells arranged in a nested and reticular pattern with Schiller-Duval bodies. Histochemistry showed Periodic acid–Schiff (PAS) positive and diastase-sensitive glycogen, PAS positive and diastase-resistant hyaline globules, cytokeratin, alfa-1-antitrypsin, and placental alkaline phosphatase. Based on the above features, the child was diagnosed with having a vaginal yolk sac tumor.

The metastatic work-up of the child computed tomography (CT) of the chest and abdomen) was negative. The tumor board decided to place her on a chemotherapy schedule of six cycles of combination chemotherapy (JEB) using carboplatin, etoposide, and bleomycin. The child has completed one cycle of chemotherapy.

DISCUSSION

Endodermal sinus tumors—or yolk sac tumors—of the infantile vagina, which was first described by Norris et al., have distinct clinico-pathological features [4]. There is a paucity of case reports concerning this rare neoplasm in literature, with only about fifty odd cases reported to date.

Yolk sac tumors are the most common germ cell tumor in children. They have been

found in pure form or as part of a mixed germ cell tumor. The most common clinical presentation is frank vaginal bleeding or bloodstained vaginal discharge. In more advanced cases, a mass can be seen protruding through the vaginal orifice. Occasionally, a child may present urinary retention due to compression of the bladder by the tumor. The origin of vaginal EST is thought to be embryonic remnants of midline mesonephric structures. This disease in patients is usually advanced and is rarely present after the age of three years. The lung and the liver are the most frequent sites of metastasis [1-6].

Imaging findings of YSTs of the infantile vagina determined soft tissue masses with irregular margins and heterogeneous enhancement. They may extend through the uterus and fallopian tubes, and the origin of larger lesions may not be recognizable. Magnetic resonance imaging (MRI) is superior to CT because it provides more information about anatomical details [5].

Assays of serum AFP values can potentially aid in preoperative diagnosis, monitor the effectiveness of treatment, and detect recurrences before clinical manifestation and staging. The immunohistochemical demonstration of AFP clinches the diagnosis [4].

Histopathological subtypes of ESTs are known as reticular (microcystic), macrocystic, endodermal sinus type, papillary, solid, glandular myxomatous, sarcomatoid, polyvesicular, hepatoid, and parietal. PAS stain can be used to identify the hyaline material. The diagnosis is based on histological patterns, typical Schiller-Duval bodies (which were hence called “mesonephroma ovarii”), PAS stain positive diastase resistant hyaline globules, and increased serum AFP [3].

The differential diagnoses of YSTs include functional ovarian tumors; embryonal rhabdomyosarcomas (sarcoma botryoides) on gross examination (soft, polypoid growths with a grapelike configuration); and clear cell carcinomas on microscopic examination (with characteristic architectural patterns). Additionally, clear cell carcinomas may be associated with adenosis, whereas yolk sac tumors occur commonly in children fewer than three years old. A vaginal examination under anesthesia is the best way to diagnose a yolk sac tumor of the vagina. [3].

As with all rare disorders, the ideal management of ESTs of the vagina in infants continues to be an enigma. Additionally, the natural history of this malignancy is not clearly understood.

Untreated patients do not survive beyond two to four months after initial presentation [3,4], underlining the dismal prognosis of this tumor.

Different treatment approaches used in the past included surgery, surgery with radiotherapy, and surgery with chemotherapy. Surgery for the treatment of ESTs varies from partial vaginectomy to total pelvic exenteration, which results in the loss of sexual and reproductive functions and possibly the loss of bladder and rectum functions as well. Long-term radiation may be an important cause of morbidity, as it can cause abnormal growth of pelvic bones, aseptic necrosis of femoral heads, destruction of bone marrow, and primary malignancy in the irradiated field [3,4]. A partial vaginectomy in combination with chemotherapy, with the aim of preserving childbearing potential, is possibly the most accepted treatment [4]. Chemotherapy with bleomycin, etoposide, and cisplatin regime has improved the outcome of these patients, as confirmed by several studies [4,7,8]. In 80% of patients complete remission can be achieved, and in at least 60% of cases long-term remission may be possible [4].

Vaginal bleeding in an infant is almost always pathological, requiring further evaluation. Vaginal yolk sac tumors are uncommon neoplasms, requiring an aggressive, multidisciplinary team approach.

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