Yolk sac tumor of the vagina

Parvin Mahzouni, MD, Shervin Pejhan, MD, Mahmoud Ashrafi, MD.

ABSTRACT

Malignant germ-cell tumors (GCT) are rare tumors of childhood accounting for less than 3% of pediatric malignancies. Endodermal sinus tumors (EST) form the most common histologic subtype of malignant GCT. The vagina is an extremely rare site for GCTs. An 8-month-old female was admitted with a short history of vaginal bleeding, and a mass protruding from the vagina. A mass was palpable anteriorly on rectal examination. Computed tomography showed a tumor mass posterior to the bladder. A biopsy revealed a vaginal EST. The serum alpha-fetoprotein was elevated. Vaginohysterectomy was carried out. She was subsequently referred to the oncologist for further management.

Saudi Med J 2007; Vol. 28 (7): 1125-1126

From the Departments of Pathology (Mahzouni, Pejhan), Pediatric Surgery (Ashrafi), Alzahra Hospital, Isfahan University of Medical Sciences, Isfahan, Iran.

Received 29th May 2006. Accepted 19th September 2006.

Address correspondence and reprint request to: Dr. Parvin Mahzouni, Associate Professor of Pathology, Department of Pathology, Alzahra Hospital, Isfahan University of Medical Sciences, Isfahan, Iran. Tel. +98 (217) 7860197. Fax. +98 (212) 2585085. E-mail: P_Shervin@yahoo.com

Malignant germ-cell tumors (GCT) are rare tumors in childhood accounting for less than 3% of pediatric malignancies.[1] In children less than 3 years old, extragonadal and testicular GCTs are the most common sites of germ cell tumors.[2] Endodermal sinus tumor (EST) is the most common histologic subtype of GCT.[3,4] We report an EST of the vagina, an extremely rare pediatric malignancy primarily affecting infants,[5,6] in order to emphasize that clinicians should be suspicious of such an underlying pathology in cases of vaginal bleeding in infants.

Case Report. An 8-month-old female was admitted with a history of vaginal bleeding for one month, and protrusion of a mass from the vagina of recent onset. The child was apparently healthy until she developed vaginal bleeding. On examination the patient was pale. A mass was palpable anteriorly on rectal examination. Pelvic CT, (with and without) contrast, revealed a solid, cystic tumoral mass (4 x 3.5 x 3 cm), located anterior to the rectum, and at the posterio-superior aspect of the bladder (**Figure 1**). There was no calcification or retroperitoneal lymphadenopathy. Contrast-enhanced computed tomography (CECT) did not show extension of the tumor beyond the vagina. A biopsy of the tumor mass showed tumor tissue arranged in a loose reticulum, and microcystic areas lined by flat and cuboidal cells. Areas of hemorrhage and necrosis were observed. Festooning pseudopapillary processes with central vessels (Schiller-Duval bodies), and hyaline droplets were identified (**Figure 2**). Immunohistochemical stain demonstrated the presence of alpha-fetoprotein (AFP) in the tumor cells (**Figure 3**), and a diagnosis of vaginal EST was made. The serum AFP was elevated at 926 ng/ml. The surgical team carried out a vaginohysterectomy. The tumor was arising from the superior part of the vagina. The uterus and cervix were free of tumor, and there was no infiltration into the bladder and rectum. The gross appearance of the tumor was that of a pale tan-yellow slimy tumor with foci of necrosis. Microscopically, the biopsy findings of yolk sac tumor were confirmed. The patient had a smooth postoperative recovery and was referred to the oncologist for further management and chemotherapy. She will be followed up with AFP level estimation and CT scan when necessary.

Discussion. An EST is the most common GCT in children. In females, it is usually encountered in the ovary.[7] An EST of the vagina is a rare, highly malignant GCT that exclusively involves children less than 3 years of age.[8,9] The clinical presentation includes a history of bloody vaginal discharge, often accompanied by a polypoid mass protruding from the vagina. A vaginal examination under anesthesia is the best way to diagnose a yolk sac tumor of the vagina.[10] Sarcoma botryoids - a variant of embryonal rhabdomyosarcoma, is the most common differential diagnosis. It is a common vaginal tumor of infancy and presents with vaginal bleeding and a mass. However, it has a typical grape like appearance and there are edematous and cellular areas formed of immature skeletal muscle cells, which help in distinguishing it from a yolk sac tumor. Formerly, a yolk sac tumor of the vagina was confused

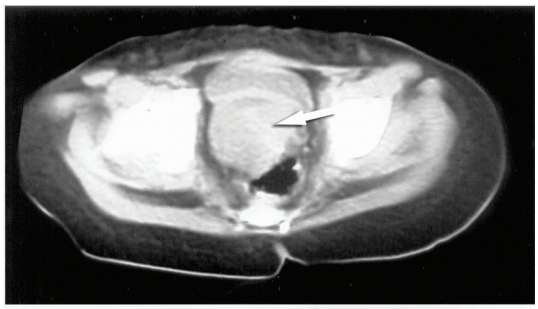

Figure 1 - Contrast–enhanced CT of pelvis showing tumor in the vagina.

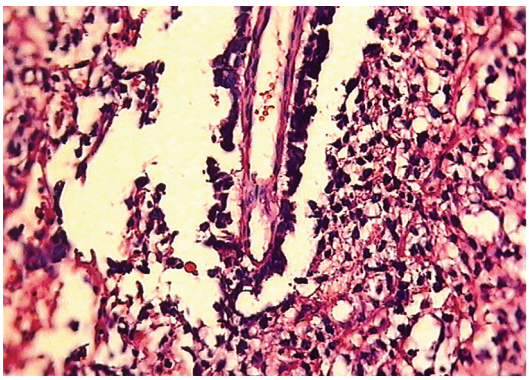

Figure 2 - Schiller–Duval bodies in the biopsied tissue. (Hematoxylin and eosin; original magnification x 400).

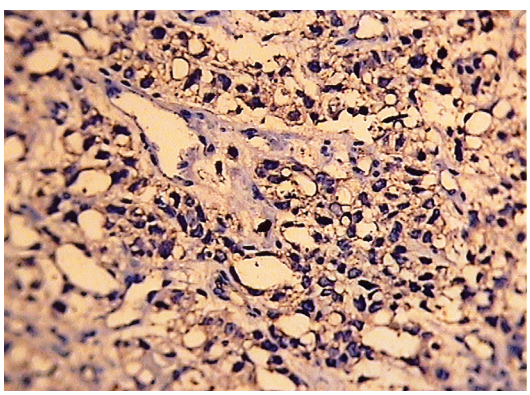

Figure 3 - The tumor cells showed positive staining for alpha-fetoprotein in the cell cytoplasm (original magnification x 400).

with a clear cell carcinoma of the vagina. However, the former has always occurred in children less than 3 years of age, and the later has not been reported at that young an age. The histopathological examination confirms the differentiation between the 2. The AFP level in blood is usually elevated at presentation and should be used as guide to monitor therapy as well as detect a recurrence.[11] The histologic diagnosis of EST is based on the finding of Schiller-Duval bodies, as demonstrated in our case. The usefulness of monitoring serum AFP in the patients with EST is well established.[12] In our case, the serum AFP was checked after histological diagnosis, as the first clinical and radiological diagnosis, based on the site of the tumor, was sarcoma botryoides. In patients suspected of having vaginal EST, elevated serum AFP is a diagnostic tumor marker that can be used to monitor the course of the disease.[13]

To conclude, we emphasize on the need for complete evaluation of infants presenting with painless vaginal bleeding. As the source of blood found in diapers of young girls without overt cause is uncertain, one should maintain a high index of suspicion for any potential underlying pathological condition. Apart from radiological investigations as diagnostic tools, one might consider measuring AFP in such patients.

References

1. Rescoria FJ. Germ cell tumors. ***Semin Pediatr Surg*** 1997; 6: 29-37.
2. Einhorn LH, Donohue J. Cis-diamminedichloroplatinum, vinblastine, and bleomycin combination chemotherapy in disseminated testicular cancer. ***Ann Intern Med*** 1977; 87: 293-298.
3. Oldham KT, Colombani PM, Foglia RP, Skinner MA, editors. Principles and practice of pediatric surgery. Philadelphia (PA): Lippincott Williams & Wilkins; 2005. p. 1625.
4. Davidoff AM, Hebra A, Bunin N, Shochat SJ, Schnaufer L. Endodermal sinus tumor in children. ***J Pediatr Surg*** 1996; 31: 1075-1078.
5. Horng YC, Tsai WY, Lin DT, Huang SF, Lin KH, Li YW, et al. Infantile endodermal sinus tumor presenting with vaginal bleeding: report of a case. ***J Formos Med Assoc*** 1994; 93: 164-166.
6. Arora M, Shrivastav RK, Jaiprakash MP. A rare germ-cell tumor site: vaginal endodermal sinus tumor. ***Pediatr Surg Int*** 2002; 18: 521-523.
7. Kurman RJ, Norris HJ. Endodermal sinus tumor of the ovary, a clinical and pathological analysis of 71 cases. ***Cancer*** 1976; 38: 2404-2419.
8. Hwang EH, Han SJ, Lee MK, Lyn CJ, Kim BS. Clinical experience with conservative surgery for vaginal endodermal sinus tumor. ***J Pediatr Surg*** 1996; 31: 219–222.
9. Ishi K, Suzuki F, Saito A, Koyatsu J, Kubota T. Cytodiagnosis of vaginal endodermal sinus tumor. A case report. ***Acta Cytol*** 1998; 42: 399-402.
10. Copeland IJ, Sneige N, Ordonez NG, Nancock KC, Gershenson DM, Saul PB, et al. Endodermal sinus tumor of the vagina and cervix. ***Cancer*** 1985; 55: 2558-2565.
11. Deshmukh C, Bakshi A, Bhagwat R, Kurkure P. Yolk Sac tumor of vagina. ***Indian J Pediatr*** 2005; 72: 367-367.
12. Kumar V, Kini P, Vepakomma D, Basant M. Vaginal endodermal sinus tumor. ***Indian J Pediatr*** 2005; 72: 797-798.
13. Rosai J, editor. Rosai and Ackermans Surgical Pathology. Philadelphia (PA): Mosby; 2004. p. 1683-1685.