Int J Clin Exp Pathol 2015;8(2):2183-2185
www.ijcep.com /ISSN:1936-2625/IJCEP0004769

Case Report

Yolk sac tumor of vagina: a case report

Ahmed Alhumidi, Safa Al Shaikh, Abdullah Alhammadi

Department of Pathology, King Khalid University Hospital, King Saud University, Riyadh, Saudi Arabia

Received December 12, 2014; Accepted February 4, 2015; Epub February 1, 2015; Published February 15, 2015

Abstract: Malignant germ-cell tumors (MGCT) are rare tumors of childhood accounting for less than 3% of pediatric malignancies. Yolk sac (endodermal sinus) tumor is one of the malignant germ cell tumor that usually involves the gonads (ovaries and testes). Its occurrence in the vagina is extremely rare. We report a 6-months old girl presented with a vaginal mass diagnosed as a yolk sac tumor. This diagnosis is confirmed by histopathologic examination, immunehistochemical studies as well as elevated serum alpha fetoprotein (AFP).

Keywords: Yolk sac tumor, endodermal sinus tumor, vagina, alpha fetoprotein

Introduction

Yolk sac tumor (YST) was first described by Teilum in 1959 [1]. Such tumors occur mostly in the ovaries and testes of young patients and they are usually related to midline structures. Only 10%-15% are extragonadal. In children less than 3 years of age, the most common sites for germ cell tumors are extragonadal and testicular [2]. Yolk sac tumors of vagina are extremely rare with only few cases have been reported in the literature. The patients usually present with vaginal bleeding.

Case report

A six months old girl, product of full term spontaneous normal vaginal delivery, presented with a three days history of vaginal bleeding. Radiological studies (**Figure 1**) (CT scan, U/S, and MRI) showed a heterogeneous mass measuring 3 × 2.7 cm occupying the vagina. Serum alpha fetoprotein (AFP) was highly elevated (5321 KIU/L), B-human chorionic gonadotropin (B-HCG) was normal (less than 0.1 KIU/L).

The patient was taken to the surgery, where an excisional biopsy was obtained. Microscopic examination showed neoplastic cells arranged in a reticular growth pattern with glomerular-like structures composed of a central blood vessel enveloped by atypical large pleomorphic cells with prominent nucleoli (Schiller-Duval bodies) floating in a loose myxoid stroma (**Figure 2**).

Immunohistochemical studies revealed that the tumor cells were focally positive for alpha fetoprotein (AFP), more diffusely positive for placenta alkaline phosphatase (PLAP) & cytokeratin (AE1/AE3) (**Figure 3**), and negative for skeletal muscle markers (desmin, myo D1 and myogenin), smooth muscle actin, and CD30. Histopathologic examination and immunohistochemical studies support the diagnosis of yolk sac tumor.

The patient received seven cycles of PEB chemotherapy (Cisplatin, Etoposide, and Bleomycin). After that, the patient was taken to surgery, where a partial vaginectomy was done. Histological examination showed extensive necrosis of the tumor with only few viable neoplastic cells indicating a good response to chemotherapy. AFP returned to normal level few weeks after surgery.

Discussion

Malignant germ cell tumors (GCT) account for only 3% of cancers in the pediatric population, and the most common histological subtype is yolk sac tumor (YST). In children, the tumors are frequently located in the sacrococcygeal region, testis, and ovary, and in the areas adjacent to these locations, the thorax, the vagina, and

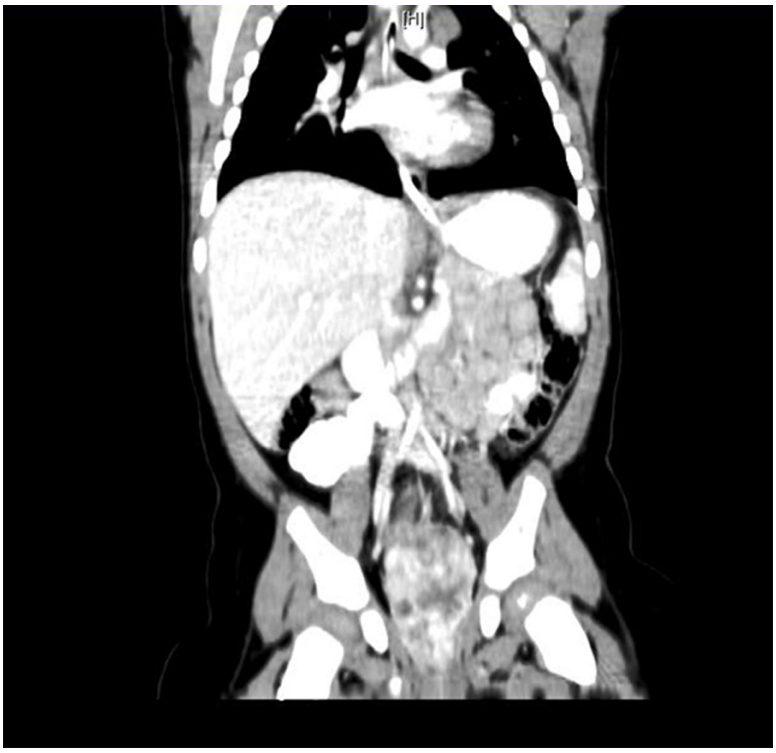

Figure 1. CT scan of the abdomen and pelvis shows heterogeneous mass occupying the vagina.

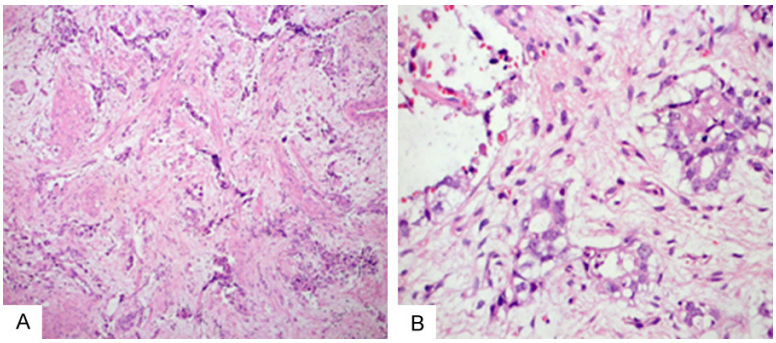

Figure 2. A. Low power shows neoplastic cells arranged in a reticular pattern with a myxoid background (H/E stain × 100). B. High power shows a central blood vessel enveloped by atypical large pleomorphic cells with prominent nucleoli (Schiller-Duval bodies) (H/E stain × 400).

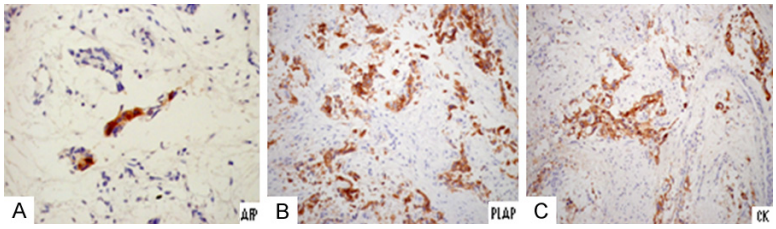

Figure 3. Immunohistochemistry stain shows focal positivity of AFP (A) and more diffuse positivity of PLAP (B) and cytokeratin (C) (Immunohistochemistry × 200).

other sites. [3, 4]. The extragonadal location of YST could be the result of germ cells "lost" during their migration from the yolk sac along the dorsal wall of the embryo to the gonadal folds [5].

YST of the vagina is a rare, highly malignant GCT that exclusively involves children less than 3 years of age. The clinical presentation includes a history of bloody vaginal discharge, often accompanied by a polypoid mass protruding from the vagina [6].

These tumors typically show a loose reticular mesh network and papillae-like structures consisting of a central vascular core lined by a single layer of cells (Schiller-Duval bodies). Some authors classify the histological patterns of yolk sac tumors as follows: festoon, reticular, solid, and polyvesicular. Most tumors exhibit more than one of these patterns [3]. These tumors are immunohistochemically positive for AFP.

Partial vaginectomy with combination chemotherapy is the most recommended line of treatment because it improves the prognosis, has acceptable complication rates, and preserves childbearing potential. The surgery eradicates local tumor cells and makes subsequent chemotherapy more effective. Incomplete excision results in local recurrence even with combination chemotherapy [6]. The serum AFP level is a useful marker for diagnosis and monitoring the recurrence of vaginal YST in infants. In our case the serum AFP levels fell to normal after surgery, indicating complete tumor excision.

To conclude, any infants presenting with vaginal bleeding or a mass protruding through the vagina we should consider measuring serum AFP in such patients as diagnostic tools in addition to radiological studies and microscopic examination. Serum AFP level is a useful marker for diagnosis & monitoring the recurrence of vaginal yolk sac tumor. Primary conservative surgery (at least partial vaginectomy) and adjuvant chemotherapy are the recommended choice of treatment because it improves the prognosis, and preserve both reproductive and sexual function.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Abdullah Alhammadi, Department of Pathology, King Khalid University Hospital, King Saud University, Riyadh, Saudi Arabia. E-mail: alhammadi.1987@gmail.com

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