Extragonadal retroperitoneal endodermal sinus tumor in an eight-month-old female infant

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Summary

We describe a *rare* case of an extragonadal retroperitoneal endodermal sinus (yolk sac) tumor in the minor pelvis. Radiologic investigation, which included abdominal ultrasound and computed tomography (CT), showed a large soft tissue mass occupying the pelvic cavity. Radionuclide bone scans demonstrated bone metastases. The serum alpha fetoprotein was elevated. Pathologic examination of the surgical specimen revealed extragonadal yolk sac tumor. Immunohistochemically, the tumor was positive for a-fetoprotein and cytokeratins. After postoperative combination therapy, follow-up CT showed decreasing tumoral disease, while serum alpha fetoprotein returned to normal.

Key words: Endodermal sinus tumor; Yolk sac tumor; CT; Ultrasound; a-fetoprotein; Retroperitoneal.

Introduction

Endodermal sinus tumor is a malignant tumor that when present in an extragonadal site, behaves in a highly malignant fashion. Endodermal sinus tumors are the most common germ cell tumor in the pediatric age group. Extragonadal germ cell tumors may arise primarily in the retroperitoneum. We report our experience of a case of extragonadal retroperitoneal endodermal sinus tumor in an 8-month-old baby girl.

Case Report

An infant, aged eight months, presented with acute abdominal pain, constipation and anuria. Symptoms were of three months duration prior to referal for radiological examination, with motor disorders and pseudodiarrhea. Physical examination revealed abdominal meteorism.

The radiological examination included conventional abdominal radiograph, sonographic examination and computed tomography (CT). Abdominal ultrasound showed urinary distention and a large echogenic mass, with bladder displacement (Figure 1). CT scan demonstrated a large, inhomogeneous, hypodense soft tissue mass, located between the rectum and sacral bone, occupying the pelvic cavity and which was not calcified (Figure 2). The tumor was not infiltrating the adjacent bones. The preoperative diagnosis included teratoma (of immature or solid mature), embryonal carcinoma, malignant mixed germ cell tumor and endodermal sinus tumor.

Pathologic examination of the retroperitoneal mass, after excisional biopsy, showed a typical endodermal sinus tumor of extragonadal origin. Immunohistochemical evaluation of the tumor revealed positivity for a-fetoprotein and cytokeratins. The serum a-fetoprotein level was elevated (4,000 ng/ml). Radionuclide bone scans revealed multiple bone metastases.

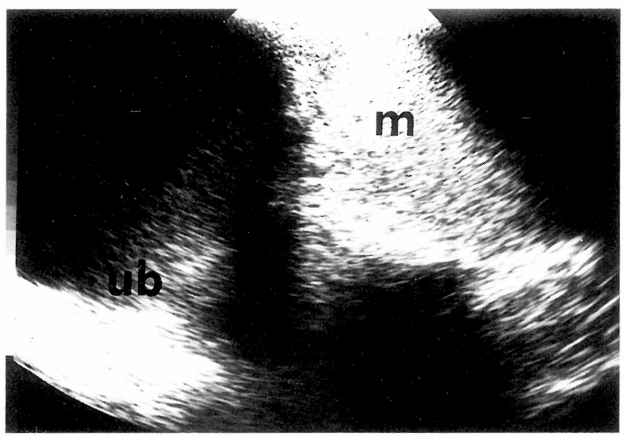

Figure 1. — Ultrasound (longitudinal view). Large echogenic mass elevating the urine bladder.

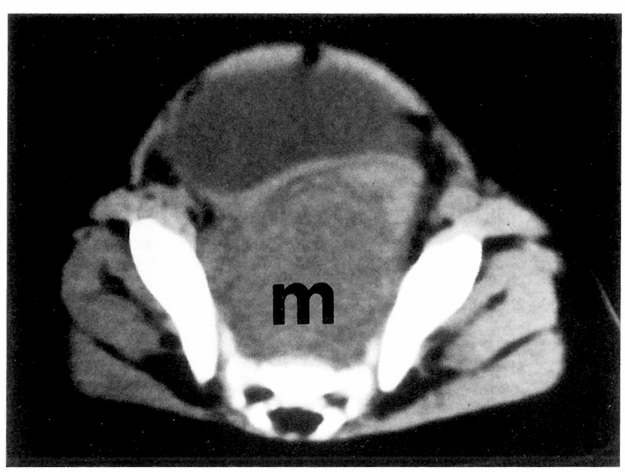

Figure 2. — CT. Large, inhomogeneous, hypodense soft tissue mass occupying the pelvic cavity.

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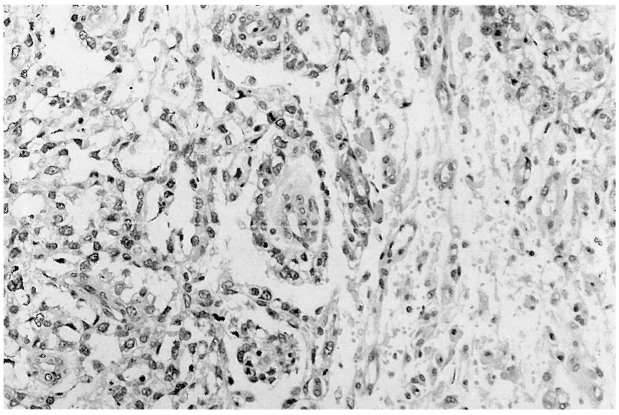

Figure 3. — Endodermal sinus tumor with characteristic Shiller-Duval bodies and PAS-positive extracellular hyaline doplets.

Discussion

Malignant gonadal and germ cell tumors are rare in children, accounting for approximately 3% of childhood cancers [1, 2]. Extragonadal tumors are believed to result from aberrant migration of germ cells from the yolk sac into the germinal ridge of the developing fetus. These are, typically, more malignant than gonadal tumors [2]. Endodermal sinus tumor, also known as yolk sac tumor, is the most common malignant germ cell tumor in infants and children [3]. Common locations of development include the ovaries, the vagina [1] and the testes [4] and, less common sites are the omentum [5], the retroperitoneum [6, 7] and the pelvis [8]. The sacrococcygeal area is the major site of involvement in the newborn and infant; in older children and adolescents, the ovary is the most common location for endodermal sinus tumors. When germ cell tumors are located in the pelvis, signs and symptoms reflect obstruction of the rectum or urinary tract by the growing mass and may be limited to constipation and/or anuria with bladder distention [2].

On imaging, CT and US provide useful information for the presence, the texture, and the location of the tumor. Furthermore, CT is a useful method to identify local tumor extension into the adjacent bone. In ultrasound examination the mass had an echogenic appearance. On CT scans the yolk sac tumor had an inhomogeneous, hypodense soft tissue appearance with heterogeneous enhancement after IV administration. The origin of the tumor was indistinguishable by computed tomography. CT and radionuclide bone scans are also used to evaluate the presence of metastatic disease.

The precice diagnosis is established histologically after complete surgical excision or a biopsy. In our case the histologic appearance was typical. There were microcystic areas formed by a loose meshwork lined by flat cells in a myxoid background. Shiller-Duval bodies and PAS-positive extracellular hyaline doplets were present (Figure 3). Immunohistochemically, the tumor was positive for a-fetoprotein [9, 10], and cytokeratin [11]. The treatment of endodermal sinus tumors is combined chemotherapy and, whenever possible, surgical excision. Human serum a-fetoprotein level is one of the most reliable tumor markers of endodermal sinus tumors originating in the ovary and extragonadal sites that can be used to monitor the effectiveness of treatment and to detect recurrences [2, 12].

After two cycles of chemotherapeutic modality (cisplatin, bleomycin, and etoposide) [2, 13], CT scan showed marked decrease in tumoral disease and the serum a-fetoprotein level returned to normal.

In conclusion, we suggest that ultrasound examination and computerized tomography are quite sensitive methods in the diagnosis of pelvic masses in infancy as well as for monitoring the response to therapy.

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