Ultrasound and Magnetic Resonance Images of Endodermal Sinus Tumor

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Endodermal sinus tumor, also known as yolk sac tumor, is a rare malignant ovarian tumor that usually occurs in the second decade of life. Here, we report a case of endodermal sinus tumor which occurred in a 14-year-old girl. She presented with lower abdominal pain for about 4 months. Initial ultrasound findings revealed a large ovarian multilocular solid tumor. Doppler study revealed multiple arteriole vessels running in the solid section of the tumor from which the lowest resistance index of 0.30 was recorded. The lactate dehydrogenase level was 261 U/L, and α-fetoprotein marker was 131,630 ng/mL. Axial contrast-enhanced T1-weighted magnetic resonance imaging showed a large cystic mass with solid components in the peripheral portions of the mass. The peripheral solid portions showed strong enhancement, indicating their hypervascularity. She received staging operation for malignant ovarian tumor, and the pathology reports revealed ovarian malignance of endodermal sinus tumor with omental invasion, stage IIIc. This case report suggests that in an adolescent female with elevated α-fetoprotein, low resistance index in the tumor mass and hypervascularity on contrast-enhanced T1-weighted magnetic resonance image, the probability of an endodermal sinus tumor should be highly suspected preoperatively. [*J Chin Med Assoc* 2007;70(11):514–518]

Key Words: adolescent adnexal mass, color Doppler ultrasonography, endodermal sinus tumor, magnetic resonance image

Introduction

Endodermal sinus tumor, also known as yolk sac tumor, is a rare malignant ovarian tumor that usually occurs in the second decade of life. The yolk sac tumor is almost always unilateral and large, measuring between 10 cm and 30 cm. The typical neoplasm manifests as a large complex pelvic mass that extends into the abdomen. The endodermal sinus tumor is often characterized by extremely rapid growth and extensive intra-abdominal spreading with poor prognosis. The cystic areas are composed of epithelial line cysts produced by the tumor or with coexisting mature teratomas.[1] Affected patients can be diagnosed by elevated serum α-fetoprotein (AFP) and lactate dehydrogenase (LDH) levels.[1,2] However, reports of preoperative diagnosis of endodermal sinus tumor in an adolescent by combining images from ultrasound and magnetic resonance imaging (MRI) as well as AFP determination are rare.

Case Report

A 14-year-old girl with body height of 164 cm presented with lower abdominal pain for about 4 months. She was quite healthy before and did well in junior high school. The patient's menarche had started at age 13, and she had had normal menstrual cycles. Her physical and clinical profile was otherwise unremarkable, and her karyotype, 46,XX, obtained at fetal age, was also unremarkable. She was referred to our outpatient department under the impression of large ovarian tumor.

Initial ultrasound findings revealed a large ovarian tumor with multicystic spaces. Transabdominal ultrasound scanning revealed an adolescent uterus measuring 60 × 46 × 43 mm (Figure 1A) with an enlarged right ovary measuring 192 × 109 × 192 mm. The size of the left ovary was normal, with no streak gonad found. The multilocular tumor mass was cystic in nature, with small thin septum measuring 2.5 mm in thickness

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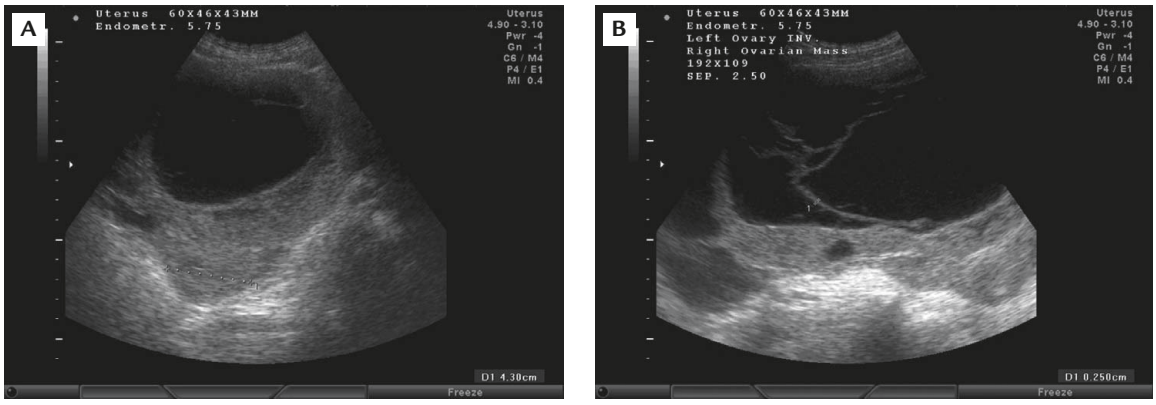

Figure 1. Two-dimensional ultrasound of the endodermal sinus tumor. (A) Transverse scan of the uterus shows a large ovarian tumor mass overlying the uterus (6.0 × 4.6 × 4.3 cm). (B) Transabdominal oblique–longitudinal scan of the ovarian tumor shows multiple thin septa measuring 2.5 mm in thickness in the cystic space.

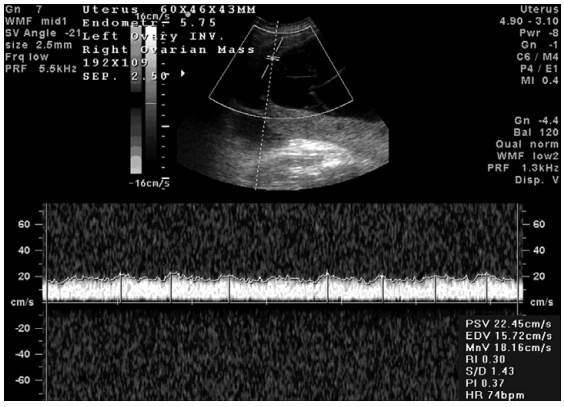

Figure 2. Color Doppler flow mapping of the endodermal sinus tumor. Doppler ultrasound shows a single arterial vessel coursing in the septum of the solid portion, with the lowest resistance index measuring 0.30.

between cysts (Figure 1B). On color Doppler flow mapping, small arteriole blood flow signal with low resistance index (RI) of 0.30 was recorded in the septum (Figure 2). Small arteriole signals were also recorded in the several intramural masses showing low blood flow RI of 0.41.

Subsequent pelvic MRI was performed by 1.5-Tesla MR scanner (Twin Excite®, GE Healthcare, Milwaukee, WI, USA) with an 8-channel phase-arrayed body coil for T1-weighted spin echo. Fast spin echo T2-weighted images (axial and sagittal planes, repetition time [TR]/echo time [TE] = 4,650/90 ms, echo train length [ETL] = 17, matrix size = 512 × 256, field of view = 20 × 20 cm, number of excitations = 4, slice thickness/gap = 5 mm/1 mm) showed a large mixed cystic and solid mass with a diameter of 19 cm occupying the pelvic cavity (Figure 3A). The uterus was displaced posteriorly. The solid portion was located mainly around the periphery of the tumor. On T1-weighted spin echo imaging (axial plane, TR/TE = 400/9 ms, matrix size = 320 × 224, field of view = 20 × 20 cm, number of excitations = 2, slice thickness/gap = 5 mm/1 mm), high signal spots were noted within the solid portion, indicating hemorrhaging (Figure 3B). After intravenous administration of gadolinium, strong enhancement with hypervascularization of the solid portion was demonstrated (Figure 3C).

The patient underwent a staging operation plus appendectomy under the impression of malignant germ cell tumor on the 5th day after admission. The castration effects for this young girl were seriously considered. Therefore, the uterus and the left adnexae were preserved. An ovarian tumor measuring 20 × 18 × 16 cm adhering to the upper portion of the omentum was removed. The gross specimen revealed areas of gelatinous necrosis in the cyst and hemorrhages in the solid portion (Figure 4A). Sections of the tumor showed a picture of yolk sac tumor, which was composed of a loose meshwork of communicating spaces lined by primitive tumor cells with clear or vacuolated cytoplasm. The lining of the papillary infolding and the cavity was irregular, with a glandular pattern. The Schiller-Duval body consisted of a cystic space, lined with a layer of flattened or irregular endothelium, into which projected a glomerulus-like tuft with a central vascular core (Figure 4B). AFP was measured in the tumor by means of an immunoperoxide technique. The pathologic diagnosis was an endodermal sinus tumor, stage IIIc, with omentum involvement > 2 cm in size. All lymph nodes and ascites fluid sampled were free of tumor cells. AFP decreased from a preoperative level of 131,630 ng/mL (normal range, 95% confidence interval, 0–20 ng/mL) to 77,401 ng/mL 10 days after the operation, and decreased further to

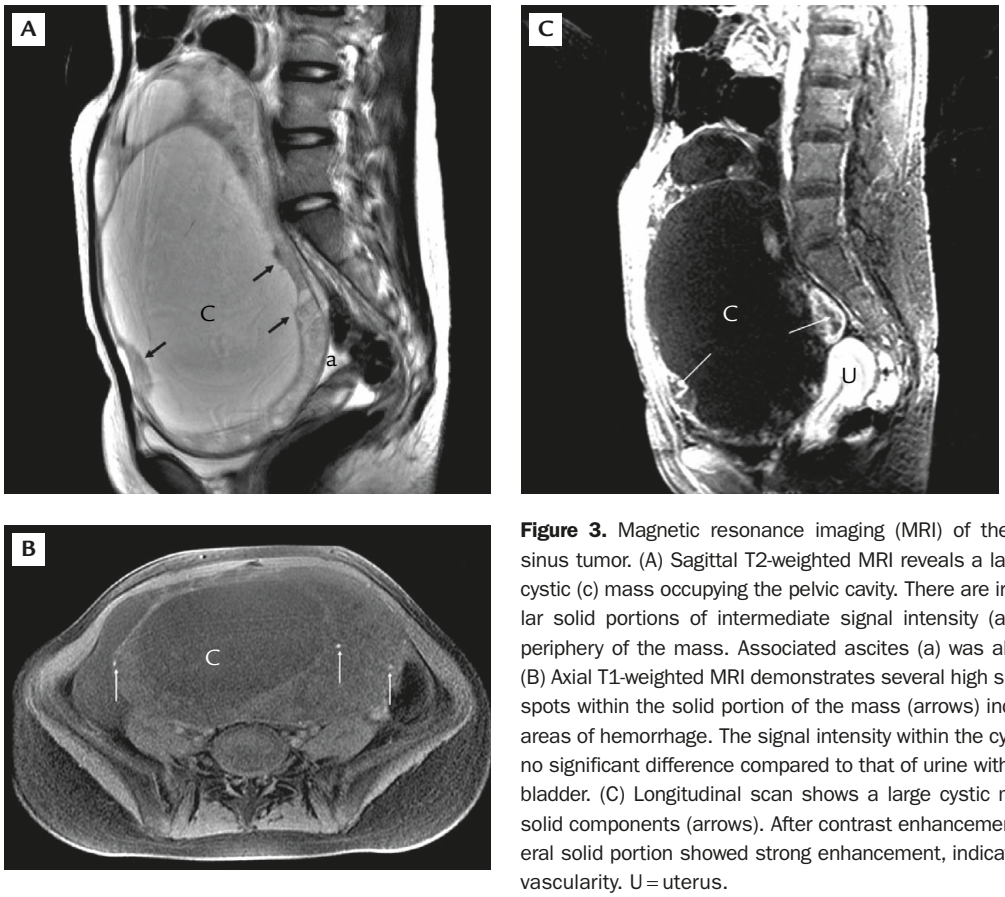

Figure 3. Magnetic resonance imaging (MRI) of the endodermal sinus tumor. (A) Sagittal T2-weighted MRI reveals a large unilocular cystic (c) mass occupying the pelvic cavity. There are irregular, nodular solid portions of intermediate signal intensity (arrows) at the periphery of the mass. Associated ascites (a) was also observed. (B) Axial T1-weighted MRI demonstrates several high signal intensity spots within the solid portion of the mass (arrows) indicating small areas of hemorrhage. The signal intensity within the cyst (c) showed no significant difference compared to that of urine within the urinary bladder. (C) Longitudinal scan shows a large cystic mass (c) with solid components (arrows). After contrast enhancement, the peripheral solid portion showed strong enhancement, indicating its hypervascularity. U = uterus.

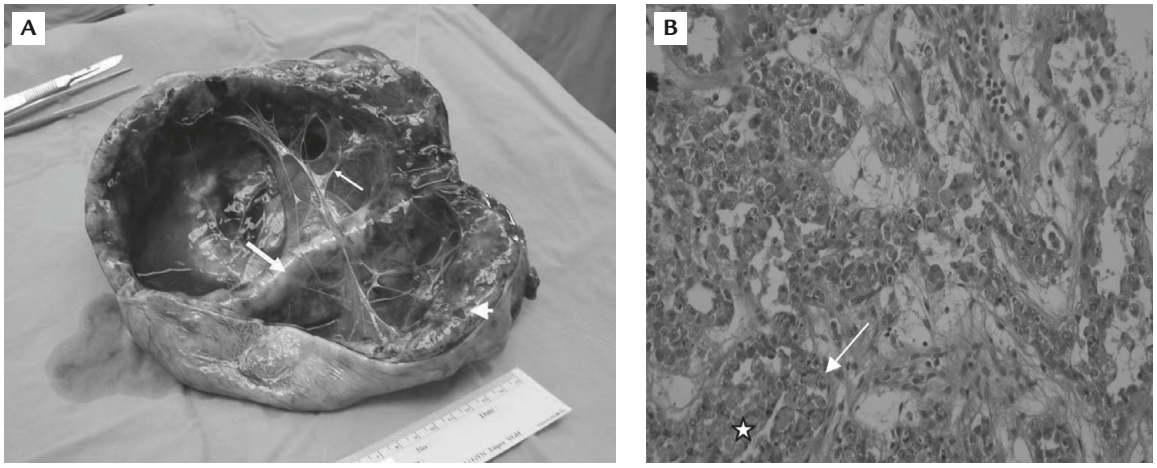

Figure 4. Gross and microscopic findings of the endodermal sinus tumor. (A) The beef-like appearance of the tumor presented with fine multilocular septa (small arrow) and a coarse septum (large arrow). The solid wall is filled with small hemorrhagic areas (arrowhead). (B) The characteristic microscopic feature of endodermal sinus. The lining of the papillary infolding and the cavity is irregular, with a glandular pattern (arrow). The Schiller-Duval body consists of a cystic space, lined with a layer of flattened or irregular endothelium, into which projects a glomerulus-like tuft with a central vascular core (☆).

50.13 ng/mL after the second cycle of bleomycin, etoposide and cisplatin (BEP). Similarly, CA-125 decreased from a preoperative level of 319 U/mL (normal range, 95% confidence interval, 0–35 units/mL) to 169 U/mL after the operation, and decreased further to 11.89 U/mL after the third cycle of BEP was completed. Chemotherapy was limited to 3 cycles to preserve ovarian function. The patient was followed-up

for 1 year and 3 months after discharge and the AFP and CA-125 levels last measured were 1.20 ng/mL and 15.17 U/mL, respectively.

Discussion

Germ cell tumors more frequently occur in girls with dysgenetic gonads with Y-chromosome component. This unique gonadal dysgenesis is associated with ambiguous genitalia, enlarged clitoris, scrotal folds, urogenital sinus and Turner stigmata.[3] The karyotype is associated with mosaicism of 45,X/46,XY. The internal genital organs are associated with hypoplastic uterus and streak gonad.[3] Our case with normal karyotype is quite different from those with dysgenetic gonads.

Endodermal sinus tumors are highly vascularized and characterized by ultrasound scanning as having multiple small arterioles. Vessels with low blood flow resistance can be detected by color Doppler flow velocity measurement (Figure 2), while vessel-enriched solid portions of the malignancy can be confirmed by MRI (Figure 3C). Microscopic examination of the endodermal sinus revealed multiple patterns. The most common appearance is a reticular, honeycombed structure of communicating spaces lined with primitive cells. Schiller-Duval bodies, which resemble the endodermal sinus of the rodent placenta, are also present. The Schiller-Duval body in our patient consisted of a cystic space, lined with a layer of flattened endothelium, into which projected a glomerulus-like tuft with a central vascular core (Figure 4B). It is speculated that the central vascular core of the Schiller-Duval body from which the small arteriole blood flowed with lower RI value was recorded in the solid portion of the endodermal sinus tumor. The vessel-enriched mass could also be confirmed by contrast T1-weighted MRI. The combined findings of enriched contrast medium in T1 imaging with lower blood flow RI in an ovarian intramural mass should serve as an alert in the diagnosis of endodermal sinus tumor in adolescent female patients.

Reported imaging findings of endodermal sinus tumors range from entirely solid to predominantly cystic.[1] But there have also been reports of heterogeneous appearance consisting of a mixture of solid and cyst.[1] The cystic portion might be due to cystic degeneration or necrosis, reflecting its rapid nature of growth. Our ultrasonographic findings were in accordance with the heterogeneous appearance, with the tumor being predominantly solid at the periphery but also having areas of cyst. Because extensive areas of hemorrhage and necrosis are common gross findings in yolk sac tumor,[4] identification of hemorrhage on MRI can help to identify this tumor in young women with solid ovarian tumors. In our case, axial T1-weighted MRI demonstrated several high signal intensity spots within the solid portion of the mass indicating small areas of hemorrhage (Figure 3B). The ultrasonographic signal intensity within the cyst had no significant difference as compared with that of the urine within the urinary bladder, indicating no evidence of intracystic hemorrhage. According to the imaging findings, the hemorrhage within the solid part of the tumor could be subtle and could be detected only by MRI, which is sensitive to deoxyhemoglobin.[5]

Another important disease entity to be distinguished from yolk sac tumor is sclerosing stromal tumor. Sclerosing stromal tumors are known to show striking enhancement which is much more prominent than that of uterine myometrium, and are also encountered in young women. The rim of hypointensity on T2-weighted images and the pattern of pseudolobulation, which consists of low intensity nodules set against high-intensity stroma on T2-weighted images, may help to diagnose sclerosing stromal tumor, distinguishing it from yolk sac tumor.[6,7] In addition, necrosis and hemorrhage have not been described in sclerosing stromal tumors in either radiologic or pathologic literature.[4,6,7] Elevated serum AFP level is key to diagnosing endodermal sinus tumor.

In conclusion, endodermal sinus tumor has common imaging findings reflecting its pathologic characteristics. When the solid portion of an ovarian tumor mass presents with (1) multiple small arterioles with lower RI detectable by color Doppler ultrasound, (2) hemorrhagic spots that can be demonstrated by T1-weighted MRI, (3) hypervascularity that can be shown on contrast-enhanced T1-weighted scans, and (4) the presence of ascites, the physician should be on the alert for endodermal sinus tumor. Although the majority of pelvic masses in young women are functional ovarian cysts, when a pelvic mass is observed with such imaging findings of malignancy, evaluation of the serum AFP level will further discriminate the tumor type.

Acknowledgments

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