Sclerosing Stromal Tumor of the Ovary in a three-year-old Girl

Chia-Ming Chang1 Shin-Lin Shih1,2

Department of Radiology1, Mackay Memorial Hospital, Taipei, Taiwan
Department of Radiology2, Taipei Medical University, Taipei, Taiwan

ABSTRACT

Sclerosing stromal tumor of the ovary is a rare, benign ovarian tumor, seen primarily in young women. We report a 3-year-old girl who presented with a palpable lower abdominal mass. Computed Tomography (CT) study revealed a huge pelvic mass with an enhancement pattern typical of ovarian sclerosing stromal tumor. The diagnosis was confirmed by pathology examination of the surgically resected tumor.

Sclerosing stromal tumor (SST) of the ovary is a rare, benign sex-cord stromal tumor [1]. It mainly affects young women in the second and third decades of life, often manifesting with menstrual irregularity. There are only a few published reports of cases in premenarchal girls. We report a 3-year-old girl with typical radiographic and pathologic features of ovarian SST.

CASE REPORT

A 3-year-old girl presented with a 10-cm palpable mass in the lower abdomen for two months. She had no abdominal pain, fever, vomiting, change in bowel habits, anorexia, or decreased activity level. Serum levels of human β-chorionic gonadotropin and α-fetoprotein were within normal limits, as were levels of estradiol, follicular stimulating hormone, and luteinizing hormone. Ultrasound examination showed a huge heterogeneous mass in the pelvic cavity. Color Doppler sonography demonstrated hypervascularity, especially in the lower portion of the tumor (Fig. 1). CT showed a huge lobulated mass measuring 10.5 × 8.3 × 5.8 cm in the pelvic cavity. There was strong peripheral enhancement after contrast medium administration, with centripetal progression on delayed images (Fig. 2). CT impression of this lesion was a huge vascular tumor, such as hemangioma. Laparotomy was performed, revealing a huge solid mass arising from the right ovary. The gross specimen of the tumor was measured 9 × 7.5 × 5.4 cm. It was a smooth, solid mass with whorls of fibrotic tissue. Microscopic examination revealed spindle cells of varying cellularity, forming a pseudolobular pattern with high vascularity, presenting as “hemangiopericytoma-like” pattern (Fig. 3). The final pathology diagnosis was SST of the ovary. Eighteen months after surgery, the girl remained well and had no evidence of recurrence on sonography.

DISCUSSION

The mean age of presentation of SST of the ovary is 28 years, 5 to 18 years younger than the average age of patients with other types of stromal tumors and much younger than those with ovarian epithelial tumors [2, 3]. Fewer than 10 cases of ovarian SST in premenarchal girls have been reported. To the best of our knowledge, our 3-year-old patient with ovarian SST is the youngest to be reported in the English literature. Though unexpected in a patient
Sclerosing stromal tumor of the ovary

this young, the imaging characteristics help to clarify the diagnosis.

Primary ovarian neoplasms are classified as epithelial, germ cell, and sex cord-stromal tumors. In adults, epithelial ovarian tumors are the most common, followed by germ cell tumors, and then stromal tumors. The pathology spectrum in pediatric patients is similar to that in adults, but the frequency and clinical behavior are different. Germ cell tumors account for more than two thirds of pediatric ovarian tumors, epithelial tumors 17%, and stromal tumors 13% [4, 5].

Sex cord-stromal tumors of ovary include SST, granulosa cell tumor, fibroma-thecoma, Sertoli-Leydig cell tumor, steroid cell tumor, and others. SST accounts for only 6% sex-cord stromal tumors [4] and was first identified as a distinctive subtype within the fibroma-thecoma group.

Figure 1

Figure 1. Abdominal sonogram shows a huge pelvic mass with abundant vascularity.

Figure 2

Figure 2. Contrast-enhanced abdominal CT revealed a 10.5 × 8.3 × 5.8 cm lobulated mass in the pelvic cavity with peripheral strong enhancement in post-contrast study a, and centripetal progression in delay images b.
Sclerosing stromal tumor of the ovary by Chalvarjian and Scully in 1973 [1]. The most common presenting symptom of ovarian SST is menstrual irregularity, although pelvic pain and palpable mass are also reported. Most ovarian SSTs are hormonally nonfunctional, although a few cases have involved androgenic or estrogenic manifestations [6]. Surgical removal of the tumor is curative; it is benign and does not recur locally or metastasize [7]. The histological features of ovarian SSTs include typical pseudolobular appearance, cellular heterogeneity, and “hemangiopericytoma-like” pattern related to prominent vasculature [8].

Imaging studies play an important role in the diagnosis of ovarian tumors. Epithelial tumors are typically primarily cystic. Malignant epithelial tumors often demonstrate a thick, irregular wall; thick septa; papillary projections; and varying portions of solid component [4].

Benign germ cell tumors (mature teratoma) may vary from purely cystic to a mixed mass with all components of the three germ cell layers. Fat attenuation within a cyst, with or without mural calcification, is a CT feature diagnostic for mature teratoma. Malignant germ cell tumors usually appear as a large, complex mass with cystic areas of necrosis. Calcifications may be seen in malignant teratoma or dysgerminoma. Tumor markers may help to distinguish among these entities [4].

The appearance of sex-cord stromal tumors of the ovary ranges from small solid masses to large multicystic masses. Granulosa cell tumors are usually large multicystic masses with solid components. Fibrothecoma, SST, and Sertoli-Leydig cell tumors are usually solid. On CT image, the appearance of sex cord stromal tumor is sometimes difficult to differentiate from that of other malignant ovarian tumors. Further imaging study may be required. For example, fibromas have very low signal intensity on T2-weighted MR images [4].

Ovarian SST may appear as a mixed solid and cystic mass, making it difficult to differentiate from malignant tumors on ultrasonography, and unnecessary extensive surgical excision, such as bilateral oophorectomy, may be performed [9]. On dynamic contrast enhanced CT and MRI studies, ovarian SST shows strong early peripheral enhancement with centripetal progression, findings similar to those in hepatic hemangioma [10]. The enhancement pattern is crucial in differentiating SST from other stromal tumors, such as fibroma or thecoma, that have mild and slow enhancement, as well as from epithelial tumors, where early enhancement and fast washout are usually observed [10].

In conclusion, we demonstrate the characteristic imaging features of ovarian SST, particularly the enhancement pattern on CT. Even though it was surprising to find the tumor in such a young patient, the typical imaging pattern was helpful in diagnosis and management. While ovarian SST would not normally be in the differential diagnosis of a pelvic mass in a very young child, knowledge of and careful attention to specific imaging patterns may

**Figure 3.** Histopathology (100x). Microscopically, the tumor is composed of spindle cells with various cellularity forming a pseudolobular pattern and high vascularity, presenting as “hemangiopericytoma-like” pattern.
Sclerosing stromal tumor of the ovary

be quite helpful, especially in planning surgery that will be curative but not overly extensive.

REFERENCES