Society of Radiologists in Ultrasound
2012 Toshiba Residents Program

Andria M. Powers, MD
University of Vermont/ Fletcher Allen Health Care

Clinical History:

A 14 year old girl presented to Emergency Department with several hours of worsending right lower quadrant pain, low-grade fever and mild nausea. She was otherwise healthy and took no medications. CBC, pregnancy test and Basic Metabolic Profile were within normal range. An ultrasound of the pelvis and right lower quadrant and CT scan of the abdomen and pelvis were performed. The studies revealed a dilated, noncompressible, fluid-filled appendix with a small amount of free fluid and adjacent fat stranding. Incidentally found was a large 5-6 cm ovoid mass in the right adnexal region. The patient was evaluated by general surgery for acute appendicitis and taken to the OR for appendectomy. At surgery, acute appendicitis was confirmed, and a large, smooth-walled, well circumscribed mass was identified in the right adnexa. Resection of the adnexal mass was deferred to Gynecologic Oncology.

Tumor markers including CA 125, AFP and CEA were negative. A month after the appendectomy, the patient returned to the OR for right oophorectomy, and the final diagnosis of sclerosing stromal tumor of the ovary was made. The patient did well with resolution of symptoms and normal menstruation.

Imaging

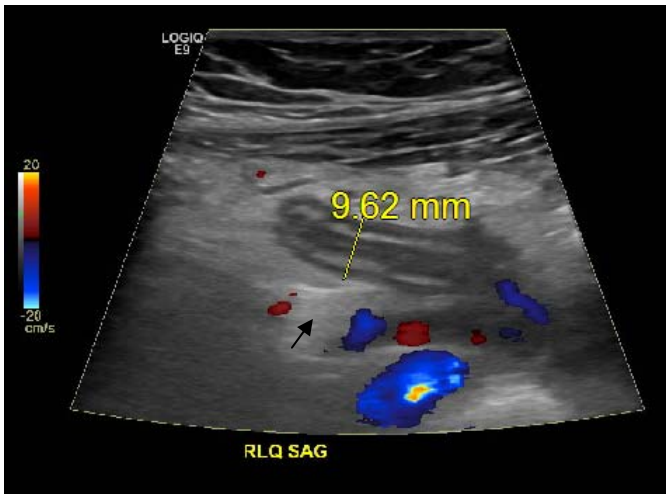

Figure 1. Static image of the right lower quadrant in the area of pain shows a tubular, blind ending structure that originated from the cecum compatible with the appendix. The appendix was enlarged, measuring 9.6mm in diameter, contained hypoechoic fluid as well as a thickened, hypoechoic wall. Real time imaging showed that the structure was noncompressible.

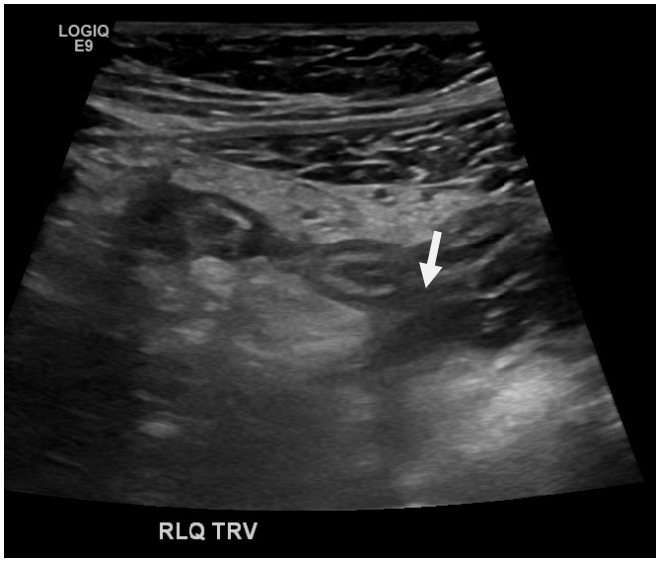

Figure 2. The appendix was vermiform and tortuous; this image shows its origin from the cecum (arrow).

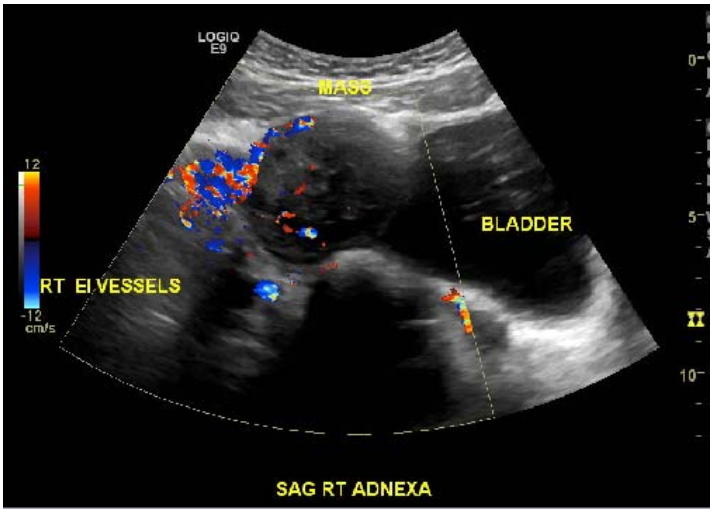

Figure 3. Ultrasound evaluation of the right pelvis revealed a large heterogeneous, but predominantly hypoechoic mass in the region of the right adnexa. Small nearly anechoic foci in

the central portion of the mass may be cystic. A right ovary could not be identified. The left ovary (not pictured) appeared normal.

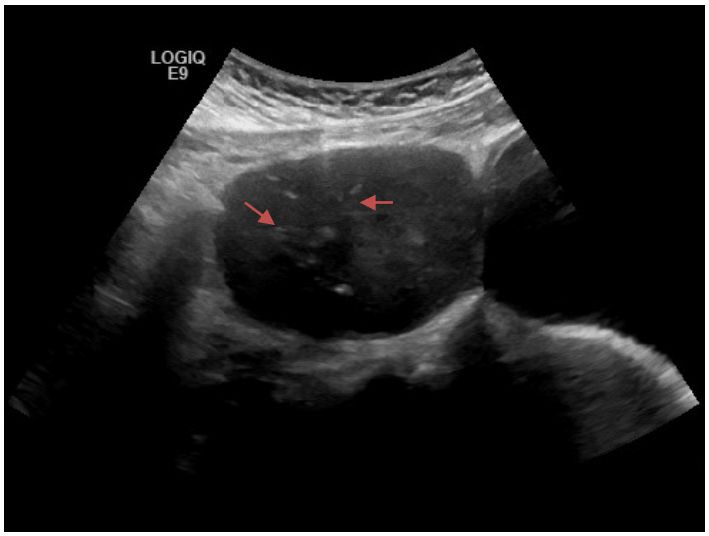

Figure 4. Well circumscribed ovoid mass in the right adnexal region. The mass is predominantly hypoechoic with a few linear regions of increased echogenicity (red arrows) and several small anechoic cystic regions.

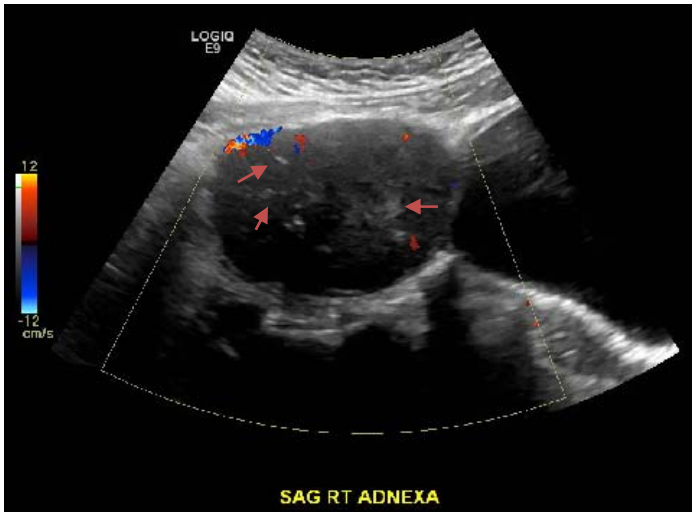

Figure 5. Ultrasound with Color Doppler flow imaging shows a few foci of color Doppler flow peripherally in the mass. This image also shows hyperechoic foci in a somewhat spoke wheel pattern of arrangement (red arrows). Several small central anechoic foci are again identified.

CT Abdomen/Pelvis

Figure 6. Contrast enhanced axial CT of the Abdomen and Pelvis shows a heterogeneously enhancing mass in the right pelvis that is contiguous with the right gonadal vessels. Central portions of the mass are hypoattenuating and poorly enhancing. The enlarged appendix to the right of the mass is partially visualized in this image (arrow) with adjacent fat stranding.

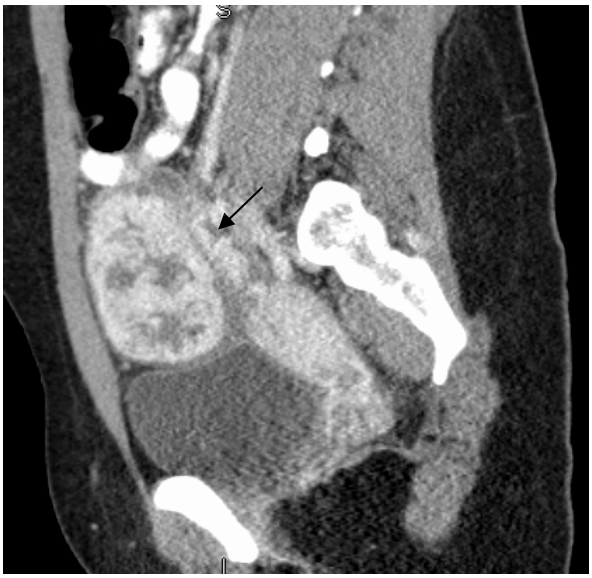

Figure 7. Sagittal reconstruction of contrast enhanced CT shows the enhancing, heterogeneous mass superior to the urinary bladder in the right hemipelvis. The enhancing portions are

nodular and predominantly peripheral with bands projecting to the center. The enhancing right gonadal vessels are contiguous with the mass (arrow). The uterus is normal appearing for age.

Figure 8. Coronal reconstruction of contrast enhanced CT shows the heterogeneous, enhancing right adnexal mass causing mass effect on the supralateral portion of the urinary bladder. The nodular regions of enhancement extend from the periphery to the center in a semi-radiating pattern.

Summary of imaging findings:

Ultrasound:

1. Enlarged, fluid filled non-compressible appendix with a thickened hypoechoic wall.

2. Large, heterogeneous but predominantly hypoechoic mass in the right adnexa with peripheral vascularity.

CT:

1. Inflamed, dilated and fluid filled appendix.

2. Heterogeneously enhancing ovoid mass in the right adnexa that is contiguous with the right gonadal vessels. The enhancement is predominantly peripheral with radiating pseudolobulations extending to the center. There are irregular hypoattenuating regions within the central portion of the mass that may be cystic.

Differential Diagnoses:

1. Acute appendicitis

2. Heterogeneous right ovarian mass in a young woman:

Sclerosing Stromal Tumor of the Ovary

Ovarian Fibroma

Granulosa Cell Tumor

Uncommon: Ovarian metastasis (Krukenberg Tumor)

Final Diagnosis:

Acute appendicitis with incidental right ovarian sclerosing stromal tumor.

Discussion:

This patient presented with acute appendicitis with the incidental discovery of a right ovarian mass. Appendicitis is the most common cause of emergent gastrointestinal surgery in children, and can be associated with complications such as perforation or abscess formation (1). Diagnosis of acute appendicitis often relies heavily on imaging support of clinical suspicion, which can be demonstrated by CT or ultrasound. Ultrasound can be used as a first line evaluation for appendicitis followed by CT if needed or if the appendix is not visualized, especially in pediatric populations (2). Multiple findings are used to identify acute appendicitis on ultrasound including a maximal outer diameter(MOD) of greater than 6 mm without compressibility, periappendiceal fluid, presence of fecalith, hyperechoic periappendiceal fat stranding, hyperemia on Doppler flow imaging, mesenteric lymphadenopathy (3-5). Current literature reports high sensitivity and specificity (98.7% and 95.4% respectively) when using MOD >/= 7mm or appendiceal wall thickness of >1.7mm (6).

Sclerosing stromal tumor of the ovary is a subtype of the ovarian sex cord stromal tumors representing a benign, rare tumor that is distinct from granulosa cell tumors, thecomas and fibromas (7). The prevalence is reported as 6-8% of ovarian stromal tumors, and they most commonly occur in women below age 30 (8-9). Often asymptomatic, a reported symptom is menstrual irregularity and symptoms improve after surgical resection (10). Typically this tumor is unilateral.

Although the imaging appearance is not pathognomonic, characteristic features of sclerosing stromal tumor of the ovary have been described. On ultrasound, it tends to appear as a heterogeneous, partially solid mass with central echogenic regions. Cystic portions are centrally located, and may be multiple, round or cleft shaped (7). There is typically low resistance flow, (10, and 11), and Doppler imaging typically shows prominent peripheral vascularity and adjacent vessels (7, 11).

On CT the tumor appears as a well circumscribed mass with heterogeneous attenuation, often with thick, nodular enhancing tissue at the periphery and irregular central regions of hypoattenuation(12). On MRI the tumor typically appears as a heterogeneous mass with intermediate to high T2 signal with thick peripheral hypointense tissue thought to represent compressed ovarian tissue (10,12,and 13). On dynamic post contrast CT or MR imaging, the tumor shows early peripheral enhancement with progressive centripetal enhancement and a nodular or "pseudolobulation" appearance (10,12,13). Ascites is uncommon but may be present (7,11).

The prognosis for sclerosing stromal tumor of the ovary is good. The tumor is benign, and surgery is curative with improvement in menstrual abnormalities. The tumor does not typically demonstrate recurrence (7-10).

References:

1. Lund, DP, Murphy EU. Management of perforated appendicitis in children. *J. Pediatr Surg.* 1994; 29: 1130-34.

2. Garcia Pena BM et al. Ultrasonography and Limited Computed Tomography in the Diagnosis and Management of Appendicitis in Children. *JAMA.* 1999; 282(11):1041-46.

3. Sivit CJ. Diagnosis of acute appendicitis in children: spectrum of sonographic findings. *AJR* .1993. 161:147–152.

4. Rettenbacher T et al. Outer diameter of the vermiform appendix as a sign of acute appendicitis: evaluation at US. *Radiology*. 2001; 218:757–762

5. Kessler N et al. Appendicitis: evaluation of sensitivity, specificity, and predictive values of US, Doppler US, and laboratory findings. *Radiology.* 2004; 230:472–478

6. Goldin A B et al. Revised ultrasound criteria for appendicitis in children improve diagnostic accuracy. *Pediatr Radiol* (2011) 41:993–999).

7. Lee, MS et al. Ovarian Sclerosing Stromal Tumors. *J Ultrasound Med.* 20:413-17, 2001.

8. Chalvardjian A, Scully R. Sclerosing stromal tumor of the ovary. *Cancer. 1973;* 31: 664-670.

9. Saitch A, Tsutsumi Y, Osamura Y, Watanabe Y. Sclerosing stromal tumor of the ovary. Immunohistochemical and electron-microscopic demonstration of smooth-muscle differentiation. *Arch Pathol Lab Med.* 1989; 113:372-6.

10. Stylianidou A, et al. Sclerosing stromal tumor of the ovary: A case report and review of the literature. *European J Gynaecol Oncol*. 2001; 22:300-4.

11. Deval B et al. Sclerosing stromal tumor of the ovary: color Doppler findings. *Ultrasound Obstet Gynecol.* 2003. 22(5):531-4.

12. R. Matsubayashi et al.: Sclerosing stromal tumor of the ovary: radiologic findings. *Eur. Radiol*. 1999; 9: 1335-1338.

13. Jung, SE et al. CT and MRI findings of sex cord-stromal tumor of the ovary. *AJR*. 2005; 185(1):207-15.