SECTION EDITOR: ENID GILBERT-BARNESS, MD

Pathological Case of the Month

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A 10-YEAR-OLD GIRL was seen with a temperature of 102°F and a 4-month history of abdominal distension without pain, nausea, or vomiting. Findings from physical examination demonstrated a large mass filling her entire abdominal cavity. A computed tomographic scan showed a heterogeneous, large, abdominal mass measuring 20 × 11 × 20 cm (**Figure 1**). Her white blood cell count was 10.8 × 10^9/L with no left shift, and levels for blood urea nitrogen were 12.1 mmol/L; creatine, 92 µmol/L; α_1-fetoprotein, 4.8 µg/L (reference range, <8.5 µg/L); and human chorionic gonadotropin assay, 3 IU/L (reference range, <5 IU/L). Antibiotics were administered for presumed urosepsis, and she underwent exploratory celiotomy. A large ovarian tumor was removed, measuring 29.0 × 19.5 × 10.5 cm and weighing 4.09 kg, and the capsule was intact. The cut surface was friable with multicystic areas (**Figure 2**). Findings from microscopic examination (**Figure 3** and **Figure 4**) of the tumor showed solid areas, and tubules were lined by columnar to cuboidal cells with prominent mitotic activity. The patient was discharged, and no recurrence of the tumor was seen at follow-up 1 year later.

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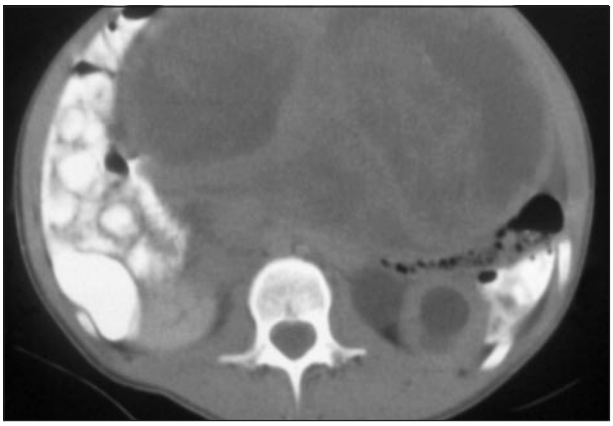
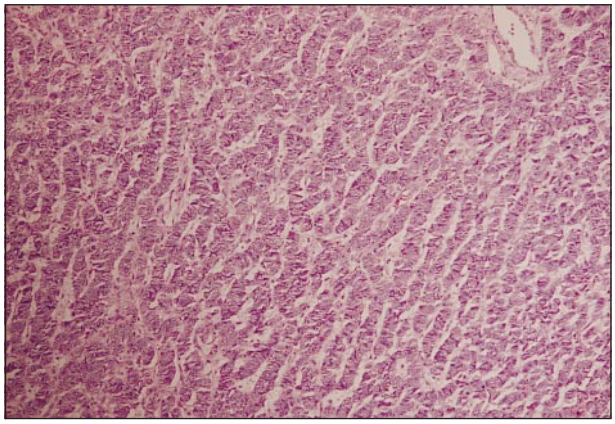


Figure 1.



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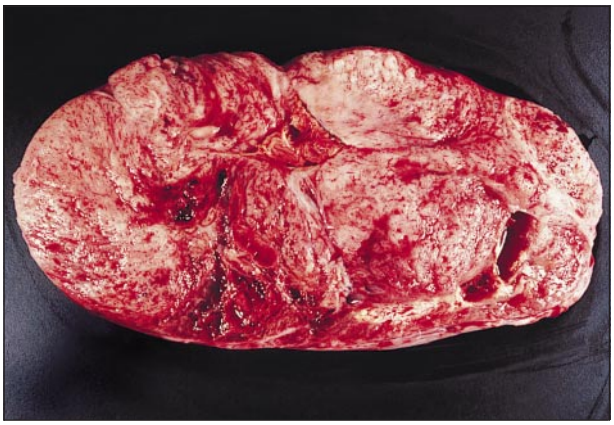

Figure 2.

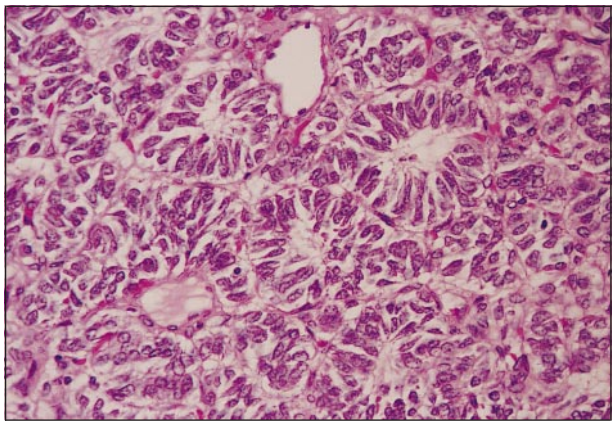

Figure 4.

Diagnosis and Discussion

Sertoli Cell Tumor

Figure 1. *Abdominal computed tomographic scan showing a heterogeneous mass.*

Figure 2. *The cut surface of the tumor was predominantly solid with an intact thick capsule and multifocal areas of hemorrhage, necrosis, and thrombosis.*

Figure 3. *The tumor was composed of well-differentiated tubules, which are cut longitudinally in this section (hematoxylin-eosin, original magnification ×100).*

Figure 4. *The tubules are lined by columnar neoplastic cells and are surrounded by scant fibrous stroma (hematoxylin-eosin, original magnification ×400).*

Sertoli cell tumors are classified as sex-cord stromal tumors and comprise fewer than 0.1% of ovarian tumors. They are included in the category of Sertoli-Leydig cell tumors even though they lack Leydig cells and account for only 4% of tumors in this generic category. These tumors have been reported in patients aged 7 to 79 years, with only a small fraction presenting in prepuberty.[1] The tumor has been referred to as arrhenoblastoma, tubular adenoma of Pick, and tubular androblastoma. Androblastoma and arrhenoblastoma denote the masculinizing potential of these tumors. Two thirds are hormonally active, with 70% secreting estrogen and 16% secreting androgens.[2]

Children with tumors producing functional hormones develop evidence of virilization, such as hirsutism, hoarsening of the voice, clitoral hypertrophy, and an increase in somatic growth. Postmenarchal girls can present with complaints of amenorrhea, hirsutism, acne, masculinization of the body habitus, and breast atrophy.[1] The differential diagnosis includes other causes of virilization: heterologous androgen administration, adrenal tumor or adrenal hyperplasia, intersex abnormalities, polycystic ovaries, or other ovarian neoplasms.[3]

To our knowledge, all pure Sertoli cell tumors reported in the English-language literature have been stage IA at the time of presentation.[4] These tumors tend to be quite large, with 60% of the patients presenting with a palpable abdominal mass. The tumors are usually unilateral and confined to the ovary.[4] On gross examination, the size ranges from 2½ to 28 cm in diameter.[5] The tumor is firm, solid, and the cut surface demonstrates a yellow, fleshy, and tan appearance. Microscopically, the cells may be cuboidal, columnar, or round with either clear granular or vacuolated, cytoplasm.[1] Three patterns are described: (1) tubular, (2) complex tubular, and (3) folliculoma lipidic of Lecene (lipid-rich).[6] Mitotic figures, if present, are usually observed in younger patients. Charcot-Böttcher crystals, parallel arrays of microfilaments forming a crystal-like lattice, are a feature of human Sertoli cells and have been identified in these tumors.[6] The survival rate for patients with Sertoli cell tumors is higher than 90%. The treatment of choice for stage IA tumors is surgical excision with no chemotherapy. Alpha$_1$-fetoprotein and Müllerian inhibiting substances have been used as biochemical markers.[5]

Accepted for publication April 30, 1999.

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Submissions

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