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Malignant Ovarian Steroid Cell Tumor: A Very Rare Tumor in Children

Fatemeh Malek*, Atbin Latifi, Saeed Habibi

Pediatric Congenital Hematologic Disorders Research Center, Research Institute for Children's Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran

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**Corresponding author:*
Fatemeh Malek,
Pediatric Congenital Hematologic Disorders Research Center, Research Institute for Children's Health, Shahid Beheshti University of Medical Sciences, Tehran, Iran

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A previously healthy 8-year-old girl was referred with abdominal pain for the past three months. Physical examination was only suspicious for an ill-defined mass palpable deeply in the flank area. An ultrasound examination of the abdomen and pelvis had shown a pelvic mass for which the patient was referred. Blood hormone tests and alpha-fetoprotein were within the normal range.

Abdominal ultrasonography revealed a 3×70×20 mm tumor originating from the right ovary. CT-scan of abdomen and pelvis revealed a well-defined hypodense right adnexal mass measuring 35×70×28 mm with heterogeneous enhancement (Figure 1). The patient underwent right salpingo-oophorectomy. Pathological examination reported a malignant ovarian tumor with intact capsule compatible with ovarian steroid cell tumor. The patients consent was obtained.



Figure 1: A and B) The computed tomography of pelvis revealed a well-defined 35×50×28 mm heterogenous mass in right adnexa.

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Ovarian tumors occur with an incidence of 2.6/100,000 women per year, of which 10-20% are malignant. They represent 3% of cancers in girls under the age of 15.[1] According to the World Health Organization, ovarian cancers are classified into three main groups with respect to their origin including: epithelial cells, germ cells or sex-cord stromal tumors.[1] Ovarian sex cord-stromal tumors which are different from epithelial and germ cell tumors, comprise approximately 7% of ovarian tumors overall and approximately 15% of ovarian tumors in children. According to their cell of origin, sex cord-stromal tumors of ovary have been divided into three subtypes: stromal luteomas, Leydig cell tumors, and steroid cell tumors (SCT).[2] SCT of the ovary account for 0.1% of all ovarian tumors.[3] Steroid cell tumors, not otherwise specified (SCT-NOS) cannot be categorized as either stromal luteomas or Leydig cell tumors.[4] SCT-NOS is reported to account for approximately 60% of steroid cell tumors, 25%–45% of which are clinically malignant.[5]

There are 13 cases of pediatric SCT-NOS reported until the time of this report, which all of them were benign except a 4-year-old girl who was presented with abdominal distention, virilization and hirsutism due to testosterone secretion from the functioning tumor.[2]

The most important factor which should be defined in steroid cell tumors of the ovary is whether the tumor has malignant features or not. There are 4 pathological features in favor of malignancy in SCT including tumor diameter >7 cm, mitotic figures per 10 high-power fields ≥2, necrosis and hemorrhage. Jiang and colleagues reported tumor embolus not only as a marker of malignancy, but also as a poor prognostic indicator of this disease.[6] The ovarian SCT-NOS in our patient was reported to be malignant due to tumor size, necrosis and high-grade nuclear atypia. It is noteworthy that most SCTs in children are functioning. Most cases of pediatric SCT-NOS were diagnosed due to various symptoms which depended on hormones secreted from the steroid cell tumor. Testosterone production is seen in SCT-NOS, with hirsutism and virilization as the most common symptoms.[2] The ovarian tumor in our patient was non-functioning and she had no signs of virilization on physical examination, since the hormone profile was within the normal range.

At imaging, steroid cell tumors frequently appear as unilateral solid tumors. Small cystic areas or necrosis has been reported. These tumors are commonly small when they cause virilization, but larger tumors have a lobulated, solid appearance.[5]

SCT-NOS is a rare type of an ovarian neoplasm in children and adults; however, should be considered in patients who present with hirsutism and elevated testosterone levels. Malignant SCT- NOS, should be managed with surgical removal followed by combination chemotherapy. There are no standard chemotherapy guidelines for treatment of SCT. PVB (cisplatin, vincristine, and bleomycin) or BEP (bleomycin, etoposide, and cisplatin) has been recommended by some authors.[7]

Conflict of Interest: None declared.

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