Paratesticular Yolk Sac Tumor

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A 17-year-old male patient, no past medical history, presented with two-month history of left groin swelling with gradual increase in size and now pain.

Clinical History:

A 17-year-old male patient, no past medical history, presented with two-month history of left groin swelling with gradual increase in size and now pain.

Laboratory Findings: Alpha Fetoprotein level was high. Quantitative Beta HCG was in normal range.

Ultrasound Findings:
Bilateral testes are WNL. Large heterogenous mass in left inquinal region in increase in vascularity, most likely tumor for clinical and laboratory correlation.

US shows large soft tissue mass at the site of swelling in left inquinal region in para-testicular region. It shows some internal vascularity. No calcification.
US shows heterogeneous hyper vascular mass in left inguinal region with some necrosis.

X-Ray Abdomen: Unremarkable.
CT chest, abdomen and pelvis was done for staging after laboratory. Findings: Large soft tissue mass at the level of the left perineum displacing the testicle anteriorly with enlarged lymph nodes. Rest of the study was within normal limit with no distant metastasis.
CT axial images show left paratesticular mass with areal of low density represent necrosis.
Coronal reconstruction.
CT shows left paratesticular mass with area of low density represent necrosis.

MRI: Left paratesticular mass shows T1 isointense with few bright signals as hemorrhagic component and shows heterogeneous enhancement.
MRI: T1 coronal: Left paratesticular mass shows T1 isointense with few bright signal as hemorrhagic component.
MRI: T2 coronal. MRI: Left paratesticular mass shows T2 mildly hyperintense with few bright signal as hemorrhagic component.
MRI: Axial T1 post contrast image shows heterogeneous enhancement of left paratesticular mass.

MRI: T1 Saggital post contrast: Left paratesticular mass shows heterogeneous enhancement.

Conclusion:
Left paratesticular mass, D/D germ cell tumors, Rhabdomyosarcoma or leiomyosarcoma.
Histopathology: Yolk Sac Tumor.
Definition:
Yolk sac tumors are those that resemble the yolk sac, allantois, and extraembryonic mesenchyme. A yolk sac tumor is a rare, malignant tumor of cells which line the yolk sac of the embryo. These cells normally become ovaries or testes; However, the tumor can also occur in areas such as in the brain or chest. The cause of a yolk sac tumor is unknown. It is most often found in children before the ages of one to two. Yolk sac tumors are also known as germ cell tumors, teratomas, embryonal carcinoma, or endodermal sinus tumors.
Epidemiology — Yolk sac tumors (YSTs) can be seen in males and females, involving the testis, ovary, and other sites, such as the mediastinum. Yolk sac tumors (YSTs) of the testis are observed in two forms or age groups: pure YST in young children and mixed type in adults.

Pure yolk sac tumor (YST) is the most common testicular neoplasm in prepubertal children, accounting for 80 percent of testicular germ cell tumors in this age group, with a median age of 1.5 years. In adults, yolk sac tumor (YST) presents as a component of mixed nonseminomatous germ cell tumor, with an age averaging 25 to 30 years. Yolk sac tumor (YST) components are present in 40 percent to 50 percent of nonseminomatous germ cell tumors in the adult testis.

In children, yolk sac tumors (YSTs) are more common in Asians than in white or black persons. In adults, these tumors are more common in white individuals than in other races.

Etiology — The etiology of yolk sac tumors (YSTs) is essentially unknown. It is speculated that hypermethylation of the RUNX3 gene promoter and overexpression of GATA-4, a transcription factor that regulates differentiation and function of yolk sac endoderm, may play important roles in the pathogenesis of yolk sac tumors (YSTs). However, these hypotheses have not been validated.

Location — Yolk sac tumors (YSTs) of the testis are located in the testis parenchyma.

Prepubertal Testicular and Paratesticular Tumors

Testicular tumors account for 1 percent to 2 percent of all pediatric tumors, with an incidence of 0.05 to 2 per 100,000 children. A bimodal age distribution is observed; one peak occurs in the first two years of life, and the second occurs in young adulthood.

Pediatric prepubertal testicular tumors are dramatically different from adult neoplasms. Germ-cell tumors account for only 60 percent to 77 percent of testicular tumors in children but account for 95 percent of testicular tumors in adults. Adult germ-cell tumors with malignant potential, such as seminoma and embryonal carcinoma, are not present in prepubertal patients. Teratomas, which are uniformly benign in children, are often malignant in adults.

The most common germ-cell tumors are teratomas and yolk-sac tumors, which account for about 62 percent and 26 percent of testis tumors, respectively.[3] Some series report that teratomas, which most believe are vastly underreported because of their benign nature, may account for almost 50 percent of prepubertal testicular tumors. However, in tumor registries, yolk-sac tumors are more common than teratomas, perhaps reflecting a reporting bias. Gonadal stromal tumors are significantly less common than germ-cell tumors (ie, tumors of non-germ-cell origin) and primarily include juvenile granulosa-cell tumors, Leydig-cell tumors, and Sertoli-cell tumors.

The vast majority (85 percent) of yolk-sac tumors in children present as clinical stage I disease, compared with 35 percent in adults. Alpha-fetoprotein (AFP) can be used as a reliable tumor marker because levels are increased in more than 90 percent of yolk-sac tumors. Therefore, patients can be safely managed with observation after orchiectomy followed by chemotherapy for recurrent tumors.[6] Retroperitoneal lymph node dissection is reserved for children with persistent retroperitoneal lymphadenopathy or increased serum tumor markers after orchiectomy and chemotherapy.

Epidemiology — International; The worldwide incidence of prepubertal testicular and paratesticular tumors is similar to that in the United States.

Mortality/Morbidity — One death occurs per every 10 million cases per year.

Race — Testicular germ cell tumors appear to have a 1.4-fold greater incidence in Asian/Pacific Islanders than in white or black children. There seems to be no discernible difference in the incidence of testicular tumors between black and white boys aged 0 to 14 years.[12]

Sex — Testicular tumors occur in boys and men.

Age — The incidence of pediatric testicular tumors peaks in children aged 2 to 4 years. Most yolk-sac tumors occur in children younger than 2 years.

History — About 85 percent of children with testicular tumors present with painless scrotal swelling. A few present with a hydrocele, scrotal pain, or a history of trauma, any of which probably alerts the child to the presence of a painless and enlarged testicle.

A hard mass may be palpable on physical examination. However, normal physical findings are not sufficient to exclude a tumor.

About 10 percent to 25 percent of patients with a malignant tumors present with a hydrocele.

Physical — Physical examination usually reveals a painless scrotal swelling with a hard mass or associated hydrocele. Some hormonally active tumors may appear in association with precocious puberty or gynecomastia.

Differentials

• Hydrocele and Hernia in Children
Paratesticular Yolk Sac Tumor
Published on Diagnostic Imaging (http://www.diagnosticimaging.com)

• Varicocele in Adolescents

Laboratory Studies
Obtain a serum AFP level before treating a testicular mass. AFP levels are elevated in 80 percent of patients with yolk-sac carcinomas and serve as a tumor marker. The half-life of AFP is about five days, and levels should return to normal (< 20 ng/mL) within one month after complete removal of the tumor. AFP levels are usually elevated in neonates (approximately 50,000 ng/mL) and drop to 10,000 ng/mL by age 2 weeks and to 300 ng/mL by age 2 months; therefore, age-specific values should be used. Persistently elevated AFP levels after surgery suggest tumor metastases or recurrence. Liver dysfunction can also cause false-positive elevations of AFP levels. Serum testosterone levels may be elevated in Leydig-cell tumors. Gonadoblastoma may elevate levels of beta-HCG.

Staging
The intergroup staging system for testicular germ cell tumors is as follows:
• Stage I - Limited to the testis and completely resected (Eighty-five percent of children < 4 y present with stage I disease, whereas only 35% of adults do.[13] )
• Stage II - Removed by transscrotal orchiectomy, involvement of scrotum or spermatic cord, persistently elevated markers
• Stage III - Retroperitoneal lymph node involvement (≤2 cm, no visceral or extra-abdominal involvement)
• Stage IV - Distant metastases

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