Case Based Urology Learning Program

Resident’s Corner: UROLOGY

Case Number 14
Case Based Urology Learning Program

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A 3 month-old boy presents with a hard painless mass in the left testicle. He is otherwise healthy and has no other abnormal physical findings.

What are the most common benign and malignant testis tumors in prepubertal boys?
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The most common benign tumor (and the most common tumor overall) is a mature teratoma (while teratomas may exhibit malignant behavior in adolescents and adults, they are universally benign prior to puberty). The most common malignant tumor in prepubertal boys is a yolk sac tumor. Other malignant tumors are exceedingly rare.
What studies would you recommend for this patient?
What studies would you recommend for this patient?

An ultrasound is usually obtained in these patients. While it cannot definitively identify the lesion, it can confirm its intratesticular location and characterize the nature of the lesion which may suggest a particular diagnosis. A serum alpha-fetoprotein (AFP) level should also be obtained as this is elevated in 90% of patients with yolk sac tumor. Serum HCG is unnecessary as the testis tumors that occur prepubertally do not elaborate HCG.
The patient’s ultrasound shows a well-circumscribed intratesticular solid mass with some cysts. The serum AFP is 100 ng/mL (the laboratory normal range for males is <15 ng/mL).

What is the likely diagnosis?
What is the likely diagnosis?

This is probably a teratoma. Most laboratories report the normal adult AFP level as “normal” regardless of the patient’s age. However, serum AFP is normally very high at birth (in the vicinity of 50,000 ng/mL) and drops exponentially until it reaches the “normal” level of <15 ng/mL by 8 to 12 months of age. A value of 100 ng/mL in a 3 month old is not abnormal. The cysts seen on the ultrasound also support the diagnosis, although that finding is not definitive.
What is the appropriate management for this tumor?
What is the appropriate management for this tumor?

While orchiectomy has been the standard treatment for testicular tumors in patients of all ages, there is growing support for testis-sparing surgery in prepubertal patients likely to harbor a teratoma or other benign tumor (e.g., epidermoid cyst). This would include patients over one year of age with a normal AFP and infants less than a year of age with an appropriate age-adjusted AFP level. In either case an inguinal incision is performed with early atraumatic occlusion of the spermatic cord at the internal inguinal ring. The testis, within the tunica vaginalis is then delivered into the operative field. If the pre-operative studies suggest a yolk sac tumor or other malignancy then a radical orchiectomy without biopsy is appropriate. If testis-sparing is being considered as would be appropriate for this patient, the testis is draped off and the tunica vaginalis is opened. The tumor is excised or enucleated and a frozen section analysis undertaken. If the frozen section confirms a benign tumor, then the testicular tunics are closed with absorbable suture and returned to the scrotum. If the frozen section suggests a malignant histology or is equivocal then an orchiectomy is performed.
Testis-sparing surgery for a benign tumor – in following images note soft occlusion of the cord with a vessel loop through an inguinal approach, enucleation of the tumor, and closure of the testis with absorbable suture.
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The frozen section analysis and final pathology reveal a juvenile granulosa cell tumor.

What other studies and/or treatment are indicated?
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Juvenile granulose cell tumors typically occur in early infancy and, like mature teratomas, are benign. No metastatic evaluation nor oncological follow-up is required.
Had the initial AFP level been elevated even correcting for the patient’s age and the patient had undergone an orchiectomy revealing a yolk sac tumor,

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Since yolk sac tumor is a potentially malignant tumor, a metastatic evaluation is indicated. The most common sites for metastases are the retroperitoneal lymph nodes and the lungs. Therefore a CT of the abdomen and pelvis with a chest x-ray or chest CT should be obtained. The AFP level should also be followed post-operatively to ensure it normalizes at the expected rate.
What is the biological half-life of AFP?
What is the biological half-life of AFP?

Approximately 5 days.
If a patient with a yolk sac tumor has a normal chest x-ray, but enlarged retroperitoneal lymph nodes and a persistently elevated AFP level, what is the appropriate treatment?
What is the appropriate treatment?

Patients with evidence of persistent disease following orchiectomy, whether evidenced by lesions in the retroperitoneum or chest or by a persistently elevated AFP level are treated with 3-4 cycles of platinum-based multi-agent chemotherapy. Retroperitoneal lymph node dissection plays a very limited role being employed only for biopsy of equivocal nodes or excision of a persistent mass following chemotherapy. The cure rate for metastatic yolk sac tumor exceeds 95%.
If a patient with a yolk sac tumor has no evidence of metastatic disease and a normalizing AFP, what is the appropriate treatment?
What is the appropriate treatment?

Yolk sac tumor patients with stage 1 disease can be managed with observation by periodic physical exam, CT of the abdomen and pelvis, chest x-ray, and measurement of serum AFP. 15-20% of stage 1 patients can be expected to recur on observation and virtually all of those patients can be cured with the same chemotherapeutic regimen that is used for initial therapy of metastatic disease.
Selected Readings

Topic:

Pediatric Urology/Neoplasms/Embryology

Subtopics:

Pediatric Testicular Tumors