AMSER Case of the Month November 2022

27 y/o male presents with bilateral testicular fullness and right sided scrotal pain

Parker Sage Anderson, MS4
University of Utah School of Medicine

Jeffrey D. Olpin, MD
University of Utah Health

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Patient Presentation

- HPI: 27 y/o male presents with bilateral testicular fullness, and right sided testicular pain. He first noticed that several years ago his testes were “lumpy and bumpy.” The fullness has become particularly noticeable and right sided pain has worsened over past several months. He has a history of classic congenital adrenal hyperplasia, diagnosed at age 3 weeks. He has been receiving steroid supplementation but admits to poor medication compliance in his teens. He also reports a history of hypogonadism with azoospermia for which he recently began receiving beta HCG injections.
- PE: Vital signs WNL. Right testicle is enlarged, painful with irregular contour. Left testicle is small, non-painful. Left testicle is firm throughout with multiple bumpy hard lesions.

Pertinent Labs

- CBC unremarkable
- Serum tumor markers (Alpha fetoprotein, LDH) within normal limits
- Serum beta HCG elevated, but received beta HCG injection previous day

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Procedure	Appropriateness Category	Relative Radiation Level
US duplex Doppler scrotum	Usually Appropriate	O
US scrotum	Usually Appropriate	O
MRI pelvis (scrotum) without and with IV contrast	May Be Appropriate	O
MRI pelvis (scrotum) without IV contrast	May Be Appropriate	O
CT abdomen and pelvis with IV contrast	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without IV contrast	Usually Not Appropriate	☢☢☢
CT pelvis with IV contrast	Usually Not Appropriate	☢☢☢
CT pelvis without IV contrast	Usually Not Appropriate	☢☢☢
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	☢☢☢☢
CT pelvis without and with IV contrast	Usually Not Appropriate	☢☢☢☢
MRI abdomen and pelvis without and with IV contrast	Usually Not Appropriate	O
MRI abdomen and pelvis without IV contrast	Usually Not Appropriate	O
Nuclear medicine scan scrotum	Usually Not Appropriate	☢☢☢

This imaging modality was initially ordered

This imaging was subsequently ordered for further characterization

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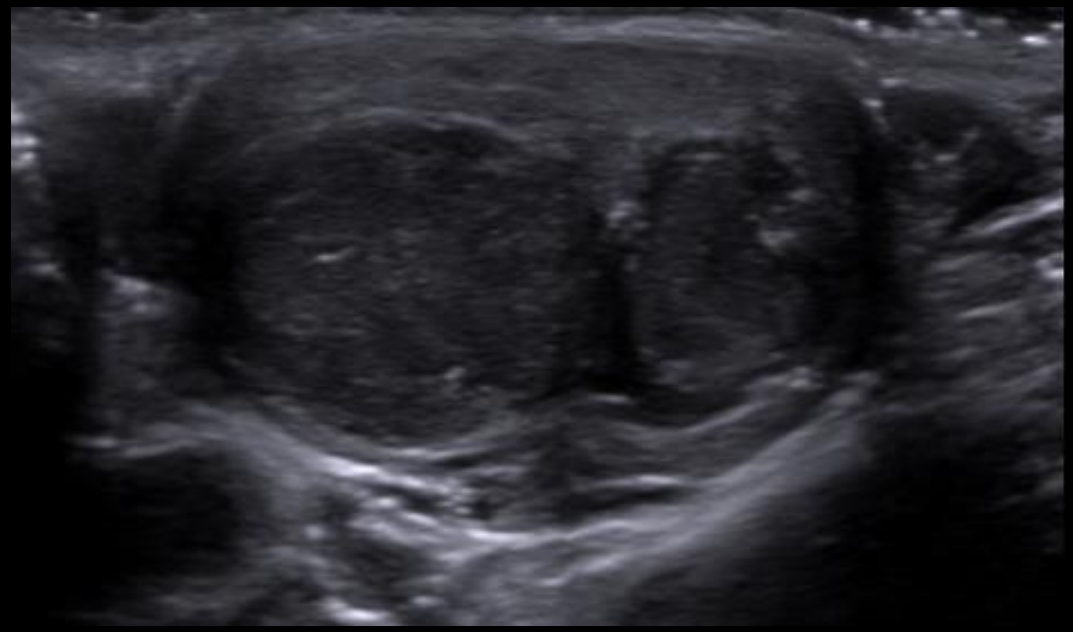
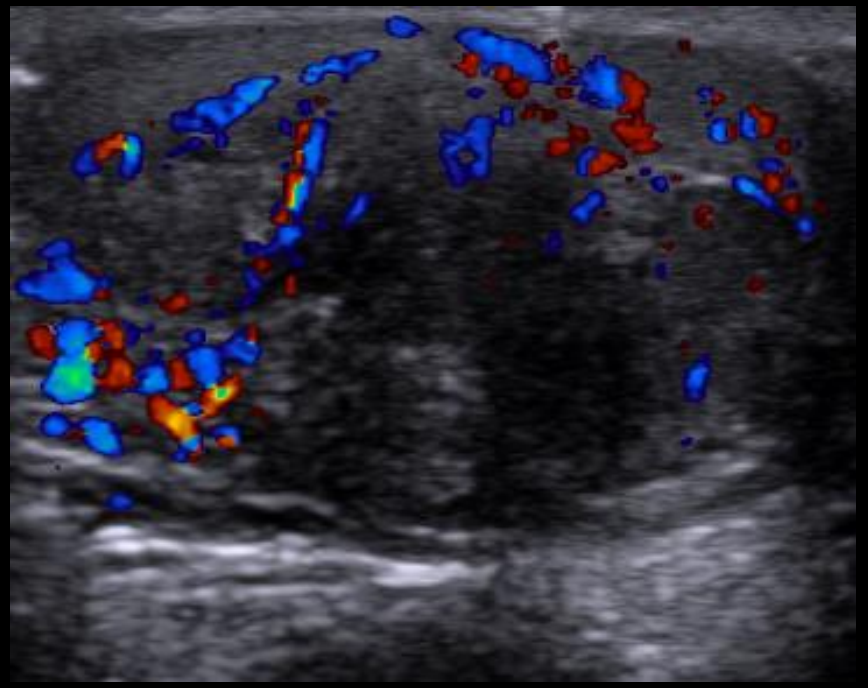
Findings (unlabeled)

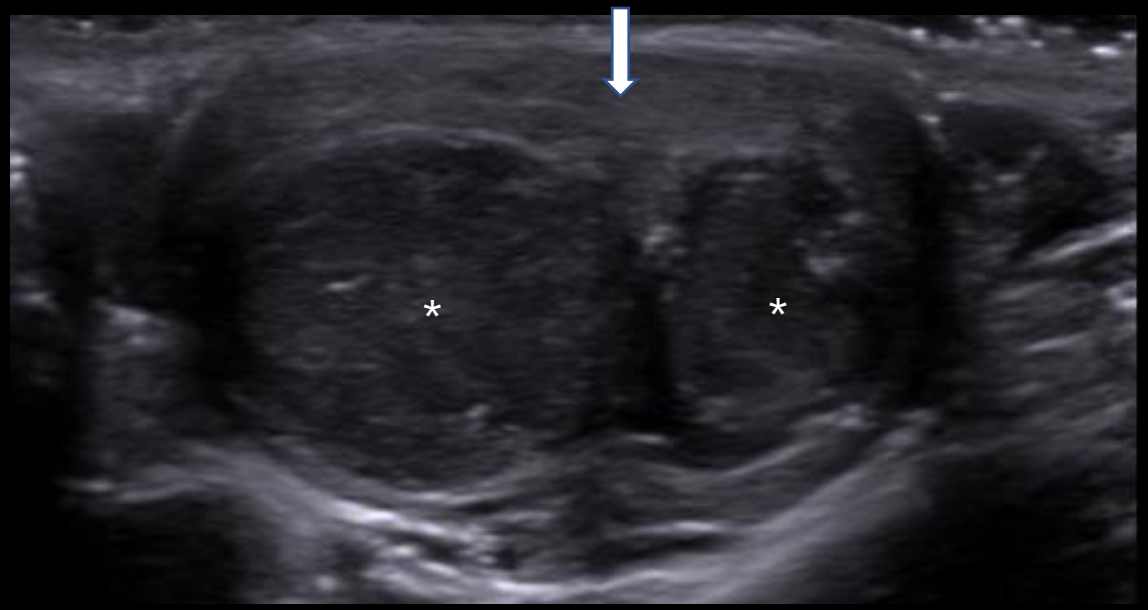


US R testis, transverse



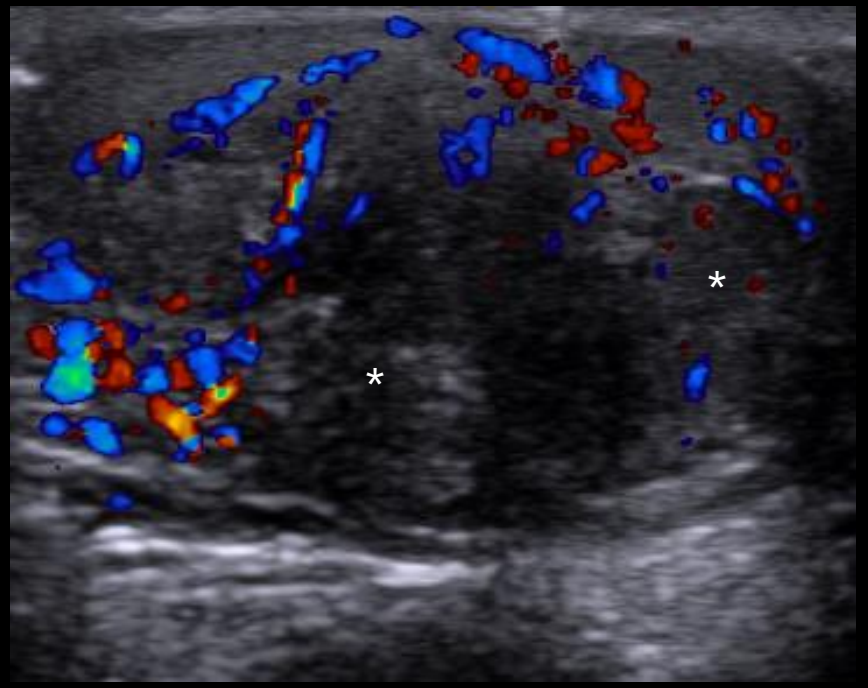
US w/ doppler R testis, transverse

Findings (labeled)



US Right testis, transverse

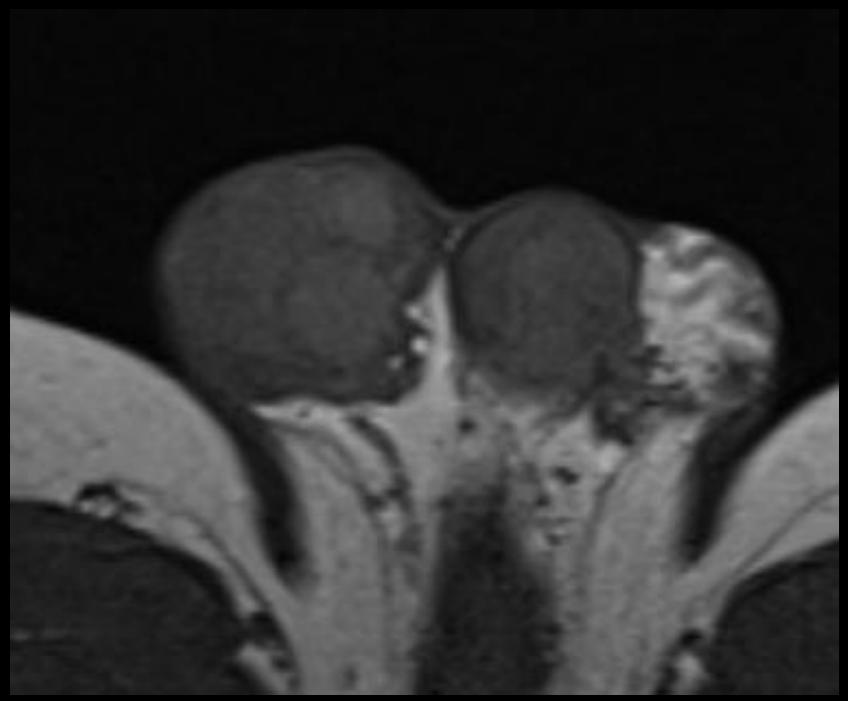
Right testis shows multiple hypoechoic masses (*) with surrounding normal testicular parenchyma (arrow)



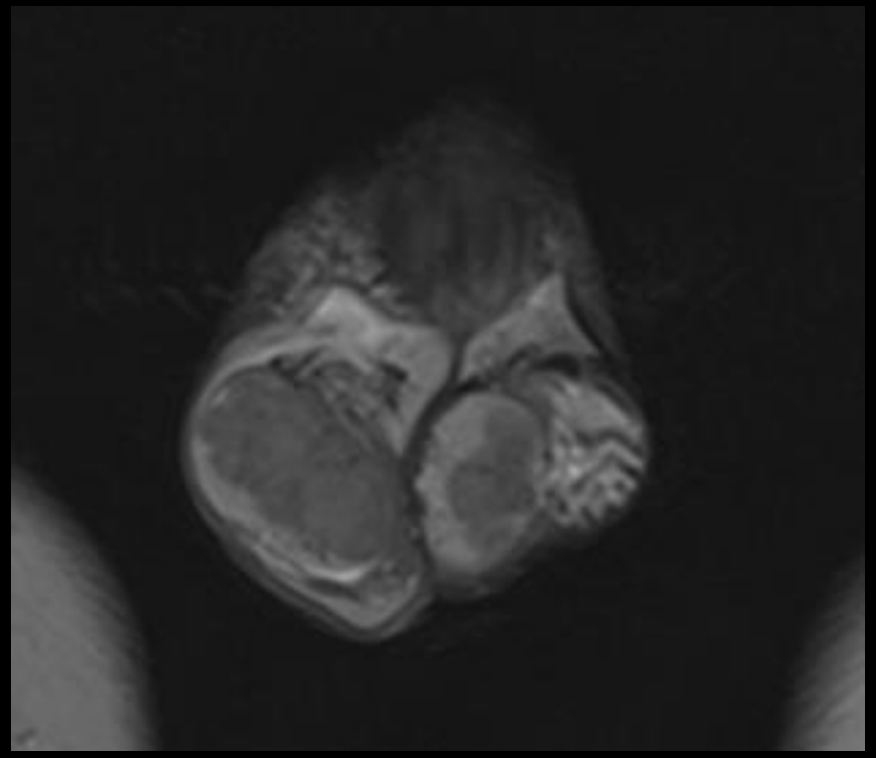
US w/ doppler Right testis, transverse

Right testis with color doppler interrogation with multiple hypoechoic masses (*) with vascular flow peripherally but little vascular flow internally

Findings: (unlabeled)

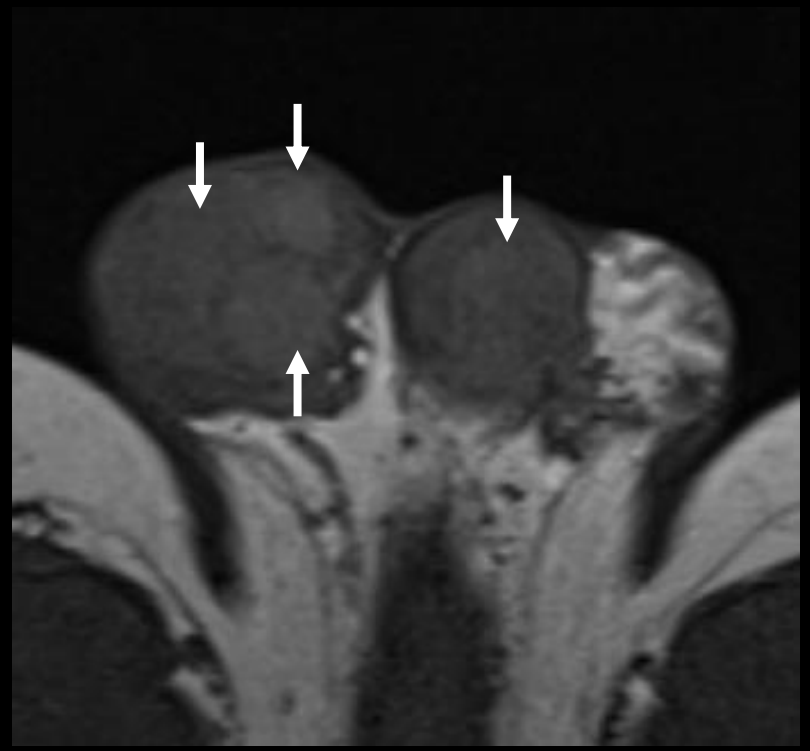


Axial T1-weighted image of bilateral testes

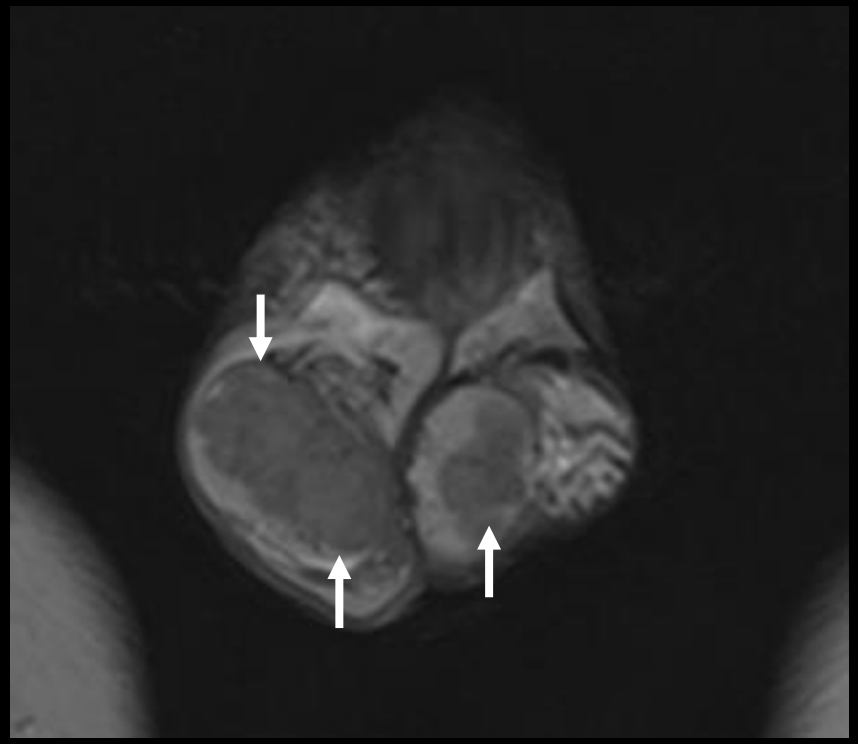


Coronal T2-weighted image of bilateral testes

Findings: (labeled)



Axial T1-weighted image demonstrates subtle masses within both testes with asymmetric enlargement of the right testis



Coronal T2-weighted image demonstrates bilateral masses that are hypointense relative to the surrounding testicular parenchyma

Final Dx:

Testicular Adrenal Rest Tumors

Testicular Adrenal Rest Tumors (TARTs)

- Testicular adrenal rest tumors are benign, frequently multiple and bilateral. These tumors occur in male patients with congenital adrenal hyperplasia (CAH) that are exposed to chronically elevated levels of ACTH. Untreated TARTs can disrupt normal testicular parenchyma with resultant infertility.
- Etiology: One theory suggests that TARTs are derived from ectopic intratesticular remnants of adrenal cortex. Another suggests that they are derived from undifferentiated stem cells which differentiate and grow under the influence of elevated ACTH.

Testicular Adrenal Rest Tumors (TARTs)

- Clinical Presentation: Often asymptomatic. Patients may present with testicular pain and infertility secondary to azoospermia due to obstruction of the rete testis.
- Differential diagnosis: Germ cell tumors, Leydig cell hyperplasia, epidermoid cysts

Testicular Adrenal Rest Tumors (TARTs)

Diagnosis:

- Often discovered through neonatal ultrasound screening programs in patients with CAH. Doppler ultrasound demonstrates hypoechoic testicular masses which are poorly vascularized and often bilateral. Ultrasound is often sufficient for establishing the diagnosis in a patient with a compatible history.

Management:

- Medical: Maintain glucocorticoid replacement. Supraphysiologic doses of glucocorticoids may also be effective in decreasing TART size and pain.
- Surgical: Partial vs. total orchiectomy can control long-term growth and pain in patients with pain refractory to hormone therapy.

Outcome & Significance

Outcome:

- Given the patient's intractible right testicular pain, he underwent radical orchiectomy. Pain subsided after orchiectomy, and he resumed hormone replacement for his CAH.

Significance:

- Testicular adrenal rest tumors are benign tumors generally seen in males with congenital adrenal hyperplasia. They typically present as hypoechoic lesions on ultrasound, which is the preferred initial imaging modality. They are often misdiagnosed as primary testicular tumors. Knowledge of this disease is necessary to recognize, diagnose and prevent unnecessary, often radical treatment.

References:

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