Testicular Adrenal Rest Tumors

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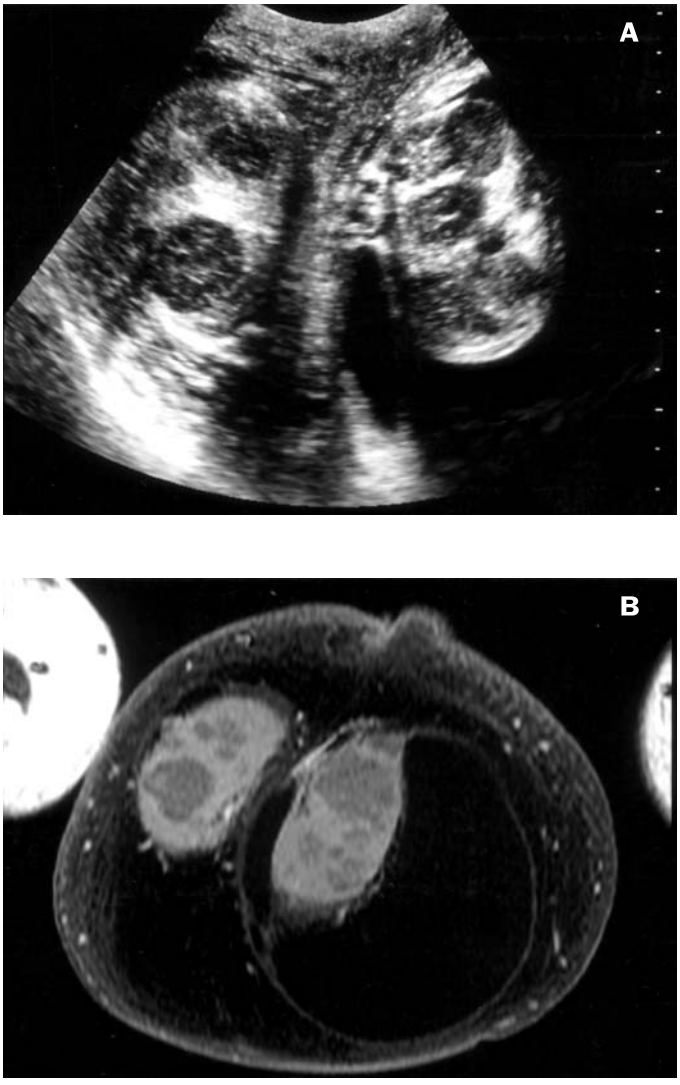
A 16 year old boy with congenital adrenal hyperplasia was referred for ultrasound evaluation of massive scrotal swelling. The sonogram showed multiple solid intratesticular masses bilaterally, varying in size up to 3 cm in diameter. Both testes were markedly enlarged and there was obvious scrotal edema. A large left hydrocele was also demonstrated [Figure A]. The sonograms from the study performed 8 years previously were retrieved and found to demonstrate similar masses. On magnetic resonance imaging the lesions were slightly hyperintense on T1-weighted and isointense on T2-weighted images, but were more clearly visualized as hypointense foci following intravenous gadolinium [Figure B]. The patient was referred to the pediatric endocrine clinic for further management and treatment, with follow-up sonography recommended.

Congenital adrenal hyperplasia is an autosomal-recessive condition caused by deficiency of 21-hydroxylase. Due to embryologic approximation between the adrenal glands and the gonads, adrenocortical cells may become included in testicular tissue. In patients with congenital adrenal hyperplasia who are untreated or inadequately treated with hormonal therapy, adrenal tissue within the testes is stimulated to enlarge, forming nodules of tissue [1]. These nodules of tissue, also known as adrenal rest tumors, may occur in other conditions with uncontrolled ACTH secretion such as Addison's disease, Cushing's disease and Nelson's syndrome. In a study of 42 patients with congenital adrenal hyperplasia, 12 were found to have testicular lesions [1]. The clinical presentation is usually in the second decade, with bilateral enlarged testes. In many patients the masses are asymptomatic.

The sonographic findings of hypoechoic, multifocal bilateral masses are often found near the mediastinum of the testes [2]. Color Doppler shows that most lesions are normovascular, but hypervascularity may be seen in some lesions [3]. On MRI the adrenal rest tumors are usually isointense or slightly hyperintense on T1- weighted and hypointense on T2-weighted images. Following intravenous gadolinium most lesions demonstrate enhancement, although in a minority of cases, as in our patient, the



Adrenal rest tumors in a 16 year old boy with a history of congenital adrenal hyperplasia.
[A] Ultrasonographic image in the transverse plane showing bilateral solid hypoechoic multifocal testicular lesions and left hydrocele. **[B]** Gadolinium-enhanced T1-weighted image, showing hypointense tumors compared with the testicular tissue.

lesions may appear hypointense relative to normal testicular tissue. The diagnosis and follow-up of testicular adrenal rest tumors, however, should be conducted by ultrasound which is more available and less costly than MRI [4].

In patients with congenital adrenal hyperplasia receiving hormonal therapy, the lesions are usually stable or decrease in size on follow-up sonography. In some cases the adrenal rest tumors may disappear. However, the lesions may occasionally increase rapidly in size, possibly due to poor compliance with therapy, raising the possibility of synchronous malignant testicular tumors, such as lymphoma [5]. In these cases, patients should first be treated with higher doses of glucocorticoids with repeated sonography 1 month later to assess the response [1]. When the adrenal rest tumor is unresponsive to steroid therapy, surgical treatment should be considered, preferably by a testis-sparing procedure. In that case, MRI is recommended because of its optimal visualization of the lesion's margins [3].

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