



Genetic Steroid Disorders: Chapter 12. Genetics of Adrenal Tumors

Antonio M. Lerario, Thomas J. Giordano, Gary D. Hammer

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Adrenocortical tumors (ACT) are common neoplasms, with a prevalence that increases with age, reaching a peak of 6% after 60 years. Most are benign cortical adenomas (ACA). Their malignant counterparts, adrenocortical carcinomas (ACC), are rare and are usually associated with a dismal prognosis. The genetic basis of adrenocortical tumorigenesis is not completely understood, but is thought to be a multistep process. Over the past two decades many molecular aspects of ACT tumorigenesis have been uncovered, especially after the elucidation of the molecular basis of genetic syndromes of which ACTs are a feature. More recently, genome-wide expression profiles and animal models have provided new insights into the explanation of this complex process. Many of the key genes and pathways have been elucidated and are the current focus of therapeutic intervention. Integrated pangenomic and other global analyses will be done in the coming years and promise to advance our understanding of adrenocortical tumorigenesis to a higher level.

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