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Management of Adrenocortical Carcinoma (ACC)

Ekta Shah*

Department of Oncology, Children's National Medical Center, Washington, United States

Introduction

Adrenocortical carcinoma (ACC) is seen in 3–10% of the population, with the majority of tumors appearing as benign tumours. The majority of ACC instances are caused by a random accumulation of mutations over time. However, research suggests that a propensity to a variety of genetic alterations may play a role. Several molecular alterations, including inactivation of tumor suppressor genes and activation of a variety of different oncogenes, DNA mutations, and epigenetic modifications, have been linked to the development of ACC in recent decades. Because of excellent clinical results, the European Network of Study of Adrenal Tumors (ENSAT) has become the most extensively used staging system for ACC. A complete history, with special emphasis to the history of symptoms of hormonal excess and family history of suspected genetic effect, is taken at the time of presentation. It is followed by a complete physical examination to determine whether or not you have ACC.

ACC management is unusual in that it encompasses both oncologic and endocrine problems. Treatment guidelines, with the exception of one experiment, are based on retrospective research and non-randomized trials, resulting in a grade II to grade IV level of evidence. The future of ACC care is personalized treatment, which includes finding the actionable target in each patient. The ACC knowledge base is growing because to fundamental science and clinical studies done by organizations throughout the world, including COMITE in France, ENSAT in Europe, the TCGA project, and the American Australian Asian Adrenal Alliance (A5). The goal of future research should be to achieve unequivocal molecular and clinical uniformity. The treatment techniques that are recommended should be recorded in the future.

Adrenocortical carcinomas (ACCs) are a rare, fatal cancer with a dismal overall survival rate [1]. The majority of ACC instances are caused by a random accumulation of mutations over time. However, studies reveal that a propensity to certain genetic alterations may have a role in the diagnosis, particularly in youngsters. In addition, ACC is frequently seen in children as part of a wider genetic condition such as LiFraumeni syndrome, Beckwith-Wiedemann syndrome, Neurofibromatosis-1 (NF1), Carney Complex, or Werner syndrome [2]. Other etiological risk factors include the use of contraception in women under the age of 25 (due to increased estrogen exposure) and males who smoke cigarettes [3]. Additionally, during pregnancy, a relative rise in ACC has been seen, which might be attributable to increased oestrogen exposure [4]. In around 10% to 20% of adult instances of ACC, a secondary malignancy develops.

Epidemiology

According to data conducted by the National Institute of Health (NIH) Office of Rare Diseases, there are less than 200,000 ACC instances in the United States (US) and ACC deaths account for 0.2 percent of all deaths each year. ACC affects 0.72 million people in the United States and 0.5 to 2 million people globally [1]. South and Southeastern Brazil have a greater than 10- to 15-fold increase in cases of ACCs due to a higher prevalence of tumour protein p53 (TP53) germline mutations of the tumour suppressor gene allele R175H and R337H alleles due to a founder effect, with approximately 78 percent of children and 13 percent of adults.

Genetic predisposition

Several biochemical alterations, including the inactivation of tumour suppressor genes and the activation of a slew of various oncogenes, DNA mutations, and epigenetic modifications, have been linked to the development of ACC in recent decades. Large-scale alterations in gene expression occur at the genome level, as do chromosomal abnormalities such as chromosomal gains, losses, and heterozygozity; DNA methylation; and dysregulation of micro ribonucleic acid (miRNA), resulting in overexpression of these sequences. TP53 and the melanocortin 2 receptor, commonly known as the adrenal corticotropin hormone receptor, are two tumour suppressor genes implicated (ACTH-R). TP53 is a critical regulator of cell proliferation that is found on the 17p13 chromosome. Dysregulation of the TP53 gene is linked to a variety of malignancies. TP53 is responsible for both germline and somatic mutations that cause ACC in patients [5].

Differential diagnosis

ACTH independent cortisol-producing adenomas and ACC are the most common differentials for hypercortisolism. Bilateral adrenal hyperplasia or adrenocortical autoantibodies can cause mineralocorticoid excess. Polycystic ovarian illness, ovarian hyperthecosis, and congenital adrenal hyperplasia are all examples of hyperandrogenemia. The National Institutes of Health (NIH) has developed standards for assessing adrenal mass that has been detected by chance. ACA, myelolipoma, adrenal metastases of another tumour, pheochromocytoma, adrenal cyst, ganglioneuroma, sarcoma, and lymphoma are the most common differential diagnoses for adrenal tumours larger than 4 cm. Cystic ACC, benign cysts such as bronchogenic or retroperitoneal cysts, and cystic pheochromocytoma are all examples of adrenal cysts. Biochemically, adrenal pheochromocytoma can be distinguished. Large adrenal tumours are further assessed by surgical excision, with the exception of adrenal lymphoma [6].

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***Corresponding author:** Ekta Shah, Department of Oncology, Children's National Medical Center, Washington, United States, E-mail: ekta_shah21@hotmail.com

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