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A Contemporary Review of Molecular Therapeutic Targets for Adenoid Cystic Carcinoma

Simple Summary: Adenoid cystic carcinoma (ACC) is a salivary malignancy known for slow growth, a propensity for perineural spread, local recurrence following resection, and indolent distant metastases. Current treatments in recurrent/metastatic (R/M) ACC are generally of limited impact and often palliative in nature. Herein, we review the preclinical and clinical literature on molecular alterations in ACC with the potential for targeted therapeutics. We further review other molecular targets of ongoing investigation and active clinical trials for patients with ACC, offering a contemporary summary and insight into future therapeutic strategies.
Abstract: ACC is a rare malignant tumor of the salivary glands. In this contemporary review, we explore advances in identification of targetable alterations and clinical trials testing these druggable targets. A search of relevant articles and abstracts from national meetings and three databases, including PubMed, Medline, and Web of Science, was performed. Following keyword search analysis and double peer review of abstracts to ensure appropriate fit, a total of 55 manuscripts were included in this review detailing advances in molecular targets for ACC. The most researched pathway associated with ACC is the MYB–NFIB translocation, found to lead to dysregulation of critical cellular pathways and thought to be a fundamental driver in a subset of ACC disease pathogenesis. Other notable molecular targets that have been studied include the cKIT receptor, the EGFR pathway, and NOTCH1, all with limited efficacy in clinical trials. The ongoing investigation of molecular abnormalities underpinning ACC that may be responsible for carcinogenesis is critical to identifying and developing novel targeted therapies.