expression in ACCs has not been reported. Neuroendocrine marker expression aids in confirmation of a neuroendocrine phenotype and CD117 positivity argues against a PLGA. This distinction is of significant therapeutic and prognostic significance.

The first-line treatment for ACC is surgical resection with local radiotherapy. ACC is a low-grade malignancy and long term survival can be expected. However, recurrences after ≥10 years have been observed, with the lung being the most commonly involved organ. Thus a careful follow-up is mandatory.

A significant proportion of ACC cases are characterized by a t(6;9)(q22-23;p23-24) translocation that results in MYB-NFIB fusion. MYB rearrangement is considered highly specific for ACC and therefore is a helpful adjunct in the distinction of ACC from other carcinomas in the lung. This distinction is important because of differences in prognosis and treatment. Moreover, therapies targeting MYB-activated pathways to treat ACC are also being explored.

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