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'Notch'ing up a role in the multisystem disease tuberous sclerosis complex

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Two independent teams of researchers have identified a role for enhanced activation of the signaling protein Notch in tumors characterized by inactivation of either the TSC1 or the TSC2 protein. As indicated by Warren Pear, at the University of Pennsylvania, Philadelphia, in an accompanying commentary, these data provide a rationale for testing whether Notch inhibitors are of benefit to those with TSC-associated tumors.

Tuberous sclerosis complex (TSC) is a multisystem disease characterized by the formation of benign tumors in multiple organs. It is caused by mutations in either the TSC1 or TSC2 gene. In the first study, Elizabeth Petri Henske, at Brigham and Women's Hospital, Boston, and Fabrice Roegiers, at Fox Chase Cancer Center, Philadelphia, found evidence of Notch signaling pathway activation in human angiomyolipomas, benign kidney tumors often found in patients with TSC, and in an angiomyolipoma-derived cell line.

Importantly, inhibition of Notch suppressed proliferation of TSC2-deficient rat cells in a xenograft model. These authors therefore conclude that TSC proteins regulate Notch activity and that Notch dysregulation may underlie some of the distinctive clinical and pathologic features of TSC.

Results presented in the second study, by Hongbing Zhang and colleagues, at the Chinese Academy of Medical Sciences and Peking Union Medical College, People's Republic of China, provide further

evidence that TSC proteins regulate Notch activity and that Notch overactivity contributes to the tumorigenic potential of cells deficient in either TSC1 or TSC2.

More information: www.jci.org/articles/view/4022 … 76V59G6WkmwhlzpC43Dd

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