Merlin

(also known as schwannomin).

Neurofibromatosis type 2 (NF2), a multiple neoplasia syndrome, is a manifestation of an impaired expression of the merlin protein, exerting inhibitory effects on cell proliferation signals due to abnormalities of the NF2 gene located on chromosome 22.

The NF2 gene product Merlin is a protein containing ezrin, radixin, and moesin domains; it is a member of the 4.1 protein superfamily associated with the membrane cytoskeleton and also interacts with cell surface molecules.

This protein is made in the nervous system, particularly in specialized cells that wrap around and insulate nerves (Schwann cells).

To carry out these tasks, merlin associates with the internal framework that supports the cell (the cytoskeleton). Merlin also functions as a tumor suppressor protein, which prevents cells from growing and dividing too fast or in an uncontrolled way.

This protein is produced in the nervous system, particularly in Schwann cells, which surround and insulate nerve cells (neurons) in the brain and spinal cord.

Although its exact function is unknown, this protein is likely also involved in controlling cell movement, cell shape, and communication between cells. Mutations in the NF2 gene lead to the production of a nonfunctional version of the merlin protein that cannot regulate the growth and division of cells. Research suggests that the loss of merlin allows cells, especially Schwann cells, to multiply too frequently and form the tumors characteristic of neurofibromatosis type 2.

The mammalian Hippo cascade, a downstream signaling cascade of merlin, inactivates the Yes-associated protein (YAP). Yes-associated protein is activated by loss of the NF2 gene and functions as an oncogene in meningioma cells; however, the factors controlling YAP expression, phosphorylation, and subcellular localization in meningiomas have not been fully elucidated. Tanahashi et al. demonstrate that merlin expression is heterogeneous in 1 NF2 gene-negative and 3 NF2 gene-positive World Health Organization grade I meningiomas. In the NF2 gene-positive meningiomas, regions with low levels of merlin (tumor rims) had greater numbers of cells with nuclear YAP versus regions with high merlin levels (tumor cores). Merlin expression and YAP phosphorylation were also affected by cell density in the IOMM-Lee and HKBMM human meningioma cell lines; nuclear localization of YAP was regulated by cell density and extracellular matrix (ECM) stiffness in IOMM-Lee cells. These results suggest that cell density and ECM stiffness may contribute to the heterogeneous loss of merlin and increased nuclear YAP expression in human meningiomas [1)].

Merlin immunohistochemistry

Merlin immunohistochemistry.

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Tanahashi K, Natsume A, Ohka F, Motomura K, Alim A, Tanaka I, Senga T, Harada I, Fukuyama R, Sumiyoshi N, Sekido Y, Wakabayashi T. Activation of Yes-Associated Protein in Low-Grade Meningiomas Is Regulated by Merlin, Cell Density, and Extracellular Matrix Stiffness. J Neuropathol Exp Neurol. 2015 Jun 5. [Epub ahead of print] PubMed PMID: 26049897.

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