DEAR EDITOR,

Neurocutaneous syndromes include numerous diseases which develop manifestations of central and peripheral nervous system and cutaneous manifestations, and are alternatively named as neurophacomatosis. Representative diseases include neurofibrosis type 1 & 2, tuberous sclerosis, von Hippel-Lindau disease, Wyburn-Mason syndrome, Sturge-Weber syndrome, neurocutaneous melanosis, and so on. The causative genes of those diseases mostly belong to tumor suppressor genes, and the gene products function as inhibitors of tumor development. Among neurocutaneous syndromes, von Hippel-Lindau disease is a unique multitumor syndrome which develops central nervous system hemangioblastoma, renal cell carcinoma, pheochromocytoma, pancreatic cysts, and so on. The causative gene product, VHL protein, functions as not only a tumor suppressor which inhibits hypoxia-inducible factor 1α and regulates mRNA elongation under normoxic condition, but also an inducer of neuronal differentiation of neural stem cells. Like this, the causative gene products of neurocutaneous syndromes might have bifunctions of tumor suppressor and inducer of neuronal differentiation.

Regards,

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