Choroid plexus (CP) tumors are brain neoplasms arising from CP epithelium. They occur most often in childhood and comprise 10-20% of all brain tumors in infants. While CP papilloma is benign, CP carcinoma is malignant and associated with poor prognosis. Previous studies have implicated Notch signaling in human CP tumors. However, the cell of origin and mechanisms underlying Notch-induced CP tumors remain unclear. We have developed novel mouse models of CP tumor caused by sustained expression of Notch 1 that recapitulate properties of human CP tumors with abnormal NOTCH signaling. While tumor cells in our models expressed CP markers Lmx1a and Otx2, the expression of aquaporin 1 and transthyretin, markers for differentiated CP epithelium, was absent or significantly reduced. Gene expression profiling revealed that tumor cell proliferation is accompanied by enhanced Sonic Hedgehog (Shh) signaling. Importantly, human CP tumors with abnormal NOTCH pathway activities also exhibited increased SHH signaling. Shh treatment stimulated the proliferation of Notch-induced CP tumor cells, while inhibition of Shh signaling suppressed tumor growth. Vertebrate Shh signaling is dependent on primary cilia. Unlike mature CP epithelial cells with clusters of primary cilia on cell surface, Notch-induced CP tumor cells possess a solitary primary cilium. Further analyses showed reduced expression of genes involved in multiciliate differentiation in tumor cells but not in epithelial cells. Lineage studies showed that Notch-induced CP tumor arose from mono-ciliated epithelial progenitors in roof plates characterized by elevated Notch signaling. Together, these results indicate that sustained activation of Notch signaling causes aberrant proliferation of epithelial progenitors, converting them into tumor-initiating cells that retain the ability to transduce Shh signals in the local environment and allows for Shh-driven proliferation. Our studies demonstrate Shh pathway is a potential therapeutic target for Notch-induced CP tumors.