



## Intracranial paramedian hourglass-shaped dermoid associated with hereditary steatocystoma multiplex.

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Abstract: This is the first report to describe the coexistence of two rare diseases, intracranial paramedian hourglass-shaped dermoid and steatocystoma multiplex. A 46-year-old female with a history of steatocystoma multiplex, bradydactylies and kyphosis showed oscillopsia, ataxia and hemifacial spasm. MRI findings suggested a giant dermoid cyst extending from the left middle temporal fossa to the cerebellopontine angle, and this was confirmed surgically. We propose the involvement of some genetic factor or pathological process common to both disorders, steatocystoma multiplex and dermoid. From the clinical point of view, patients with cystic skin lesion should also be checked for intracranial lesion.

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