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1: [Pediatr Neurosurg.](#) 2009;45(1):61-8. Epub 2009 Mar 4.

**KARGER** Full Text

**CNS tumor 22 years after spinal neuroblastoma IV: diagnostic dilemma between recurrence and secondary malignancy.**

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We present the very unusual case of a young woman suffering from a brain tumor 22 years after a stage IV spinal neuroblastoma as an infant, demonstrating the difficulties of differentiating late neuroblastoma relapse from secondary supratentorial primitive neuroectodermal tumor (sPNET). Lacking specific immunohistochemical features, the first cerebral tumor at the age of 21 was regarded as sPNET, and we pursued a therapeutic approach consisting of neurosurgical resection as well as irradiation and high-dose alkylator-based chemotherapy according to the HIT2000 protocol. Two years later the patient suffered from a diffusely infiltrating local recurrence, changing its imaging appearance as well as its immunohistochemical characteristics, now revealing disseminated positivity for neuron-specific enolase and neural cell adhesion molecule. Moreover, the lack of PNET-specific translocations (EWS/FLI1 gene fusion) in both brain tumors as well as the development of hepatic metastases was more compatible with the diagnosis of a very late relapse 22 years after initial stage IV spinal neuroblastoma.

PMID: 19258732 [PubMed - in process]

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