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Congenital extraventricular neurocytoma with atypical features in a 3-day-old neonate

Sho Matsunaga¹, Yusuke Kimura², Yukinori Akiyama², Masaki Yamamoto³, Nobuhiro Mikuni⁴

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Abstract

Extraventricular neurocytoma (EVN) is a rare neuronal tumor with a marked tendency towards ganglionic or glial differentiation. Although EVN commonly occurs in young adults, congenital cases are extremely rare, and standardized therapeutic strategies remain undetermined. The presence of atypical features such as increased mitotic activity on histological analysis is correlated with a higher rate of recurrence and poor prognosis. A 3-d-old infant with no abnormalities at birth presented with recurrent apnea and paroxysmal eye deviation on the upper right side. Magnetic resonance imaging revealed a large intracranial tumor located anterior to the brainstem. The patient underwent biopsy via craniotomy, leading to a histological diagnosis of EVN with atypical features. Despite the absence of adjuvant radiation therapy or chemotherapy, the lesion remained stable over 18 months, and the patient's growth was within normal limits. EVN is an important differential diagnosis of congenital brain tumors. Therapeutic strategies for congenital cases should be distinguished from those used for older children, and treatment decisions should be based on individual patient risks.

Keywords: Atypical features; Brain stem; Congenital brain tumor; Extraventricular neurocytoma; Neonate.

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