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· 临床医学图像 ·

小脑发育不良性节细胞瘤/Lhermitte-Duclos 病

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Dysplastic cerebellar gangliocytoma/Lhermitte-Duclos disease

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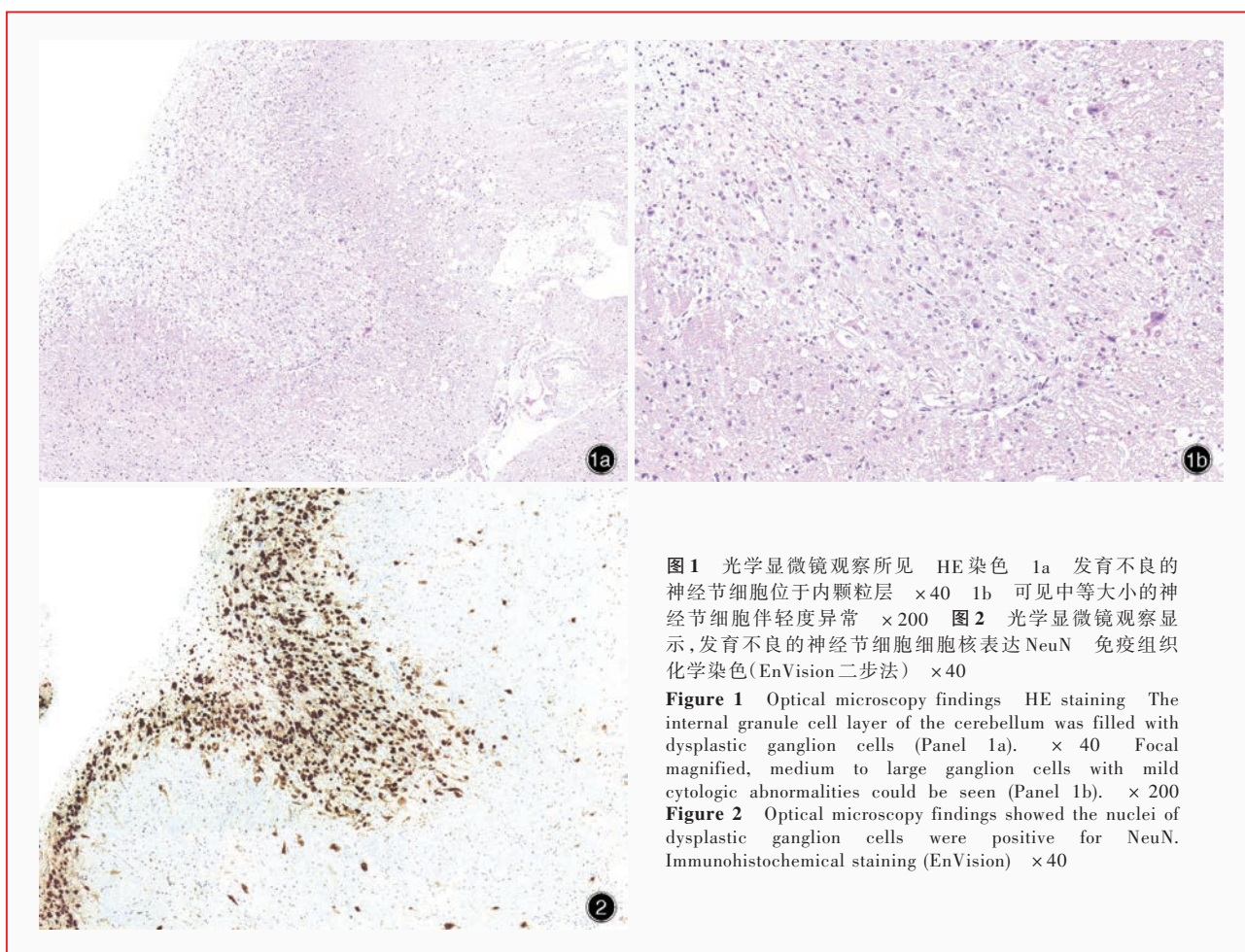


图1 光学显微镜观察所见 HE 染色 1a 发育不良的神经节细胞位于内颗粒层 $\times 40$ 1b 可见中等大小的神经节细胞伴轻度异常 $\times 200$ 图2 光学显微镜观察显示,发育不良的神经节细胞细胞核表达 NeuN 免疫组织化学染色(EnVision 二步法) $\times 40$

Figure 1 Optical microscopy findings HE staining The internal granule cell layer of the cerebellum was filled with dysplastic ganglion cells (Panel 1a). $\times 40$ Focal magnified, medium to large ganglion cells with mild cytologic abnormalities could be seen (Panel 1b). $\times 200$ **Figure 2** Optical microscopy findings showed the nuclei of dysplastic ganglion cells were positive for NeuN. Immunohistochemical staining (EnVision) $\times 40$

小脑发育不良性节细胞瘤是临床罕见的良性小脑占位性病变,亦称为 Lhermitte-Duclos 病(LDD),由发育不良的神经节细胞在原小脑结构层次中形成占位性病变,增大的神经节细胞主要位于内颗粒层,使小脑皮质增宽。小脑发育不良性节细胞瘤是常染色体显性遗传性疾病 Cowden 综合征在中枢神经系统的主要表现。目前尚未确定病变是肿瘤性还是错构性,如果呈肿瘤性相当于 WHO I 级。组织学形态可见弥漫性增厚的小脑分子层和颗粒层内大量异常增生的神经节细胞(图1),但原有结构仍相对保留;分子层外可见平行排列的异常有髓纤维;浦肯野细胞减少或消失,亦可见扩张血管和钙化灶。免疫组织化学染色,异常神经节细胞细胞核表达神经元核抗原(NeuN,图2)和突触素(Syn)、而不表达同源性磷酸酶-张力蛋白(PTEN),胶质纤维背景表达胶质纤维酸性蛋白(GFAP),神经纤维表达神经微丝蛋白(NF)。

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