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· 临床医学图像 ·

促纤维增生/结节型髓母细胞瘤

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Desmoplastic/nodular medulloblastoma

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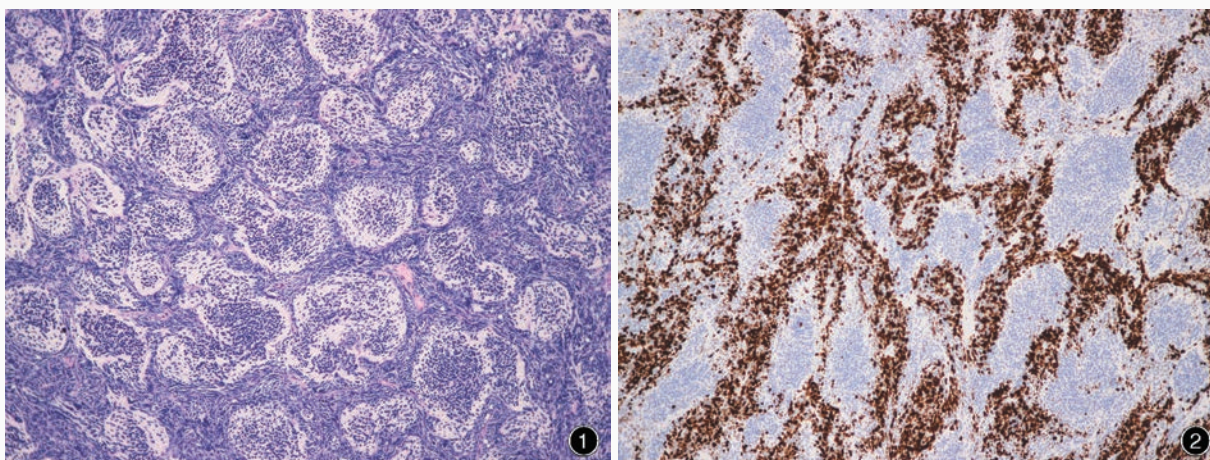


图1 光学显微镜观察显示,淡染的结节区周围包绕密集、未分化、染色质浓染的肿瘤细胞 HE染色 ×100 图2 光学显微镜观察显示,结节间Ki-67抗原标记指数明显增加 免疫组织化学染色(EnVision二步法) ×100

Figure 1 Optical microscopy findings showed pale nodular areas were surrounded by densely packed hyperchromatic undifferentiated tumor cells. HE staining ×100 Figure 2 Optical microscopy findings showed high Ki-67 labeling index in internodular regions. Immunohistochemical staining (EnVision) ×100

促纤维增生/结节型髓母细胞瘤是发生于小脑的中枢神经系统胚胎性肿瘤,以结节状、无网织纤维区周围包绕致密的生成网织纤维的分化较差的肿瘤细胞为特征,属WHO IV级。可发生于儿童、青少年和成人,主要位于小脑半球和中线结构。幼年发病者常与家族性痣样基底细胞癌综合征(亦称Gorlin综合征)相关。其特征性病理生理学机制是Shh信号转导通路病理性激活。肿瘤组织结节状、无网织纤维区(淡染岛)周围包绕密集、未分化、染色质浓染、中等多形性细胞核、高度增生的生成大量网织纤维的肿瘤细胞(图1);不同成熟程度的结节内肿瘤细胞嵌于神经毡样纤维基质中,结节内核分裂象低于结节间,无神经母细胞性“菊形团”样结构。必须同时具备促纤维增生和结节状两项特征方可明确诊断为促纤维增生/结节型髓母细胞瘤。免疫组织化学染色,结节内肿瘤细胞胞质表达突触素(Syn),胞核表达神经元核抗原(NeuN),结节间肿瘤细胞Ki-67抗原标记指数明显高于结节内(图2)。

(天津市环湖医院病理科阎晓玲供稿)