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·临床医学图像·

胼胝体发育不良

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Dysgenesis of corpus callosum

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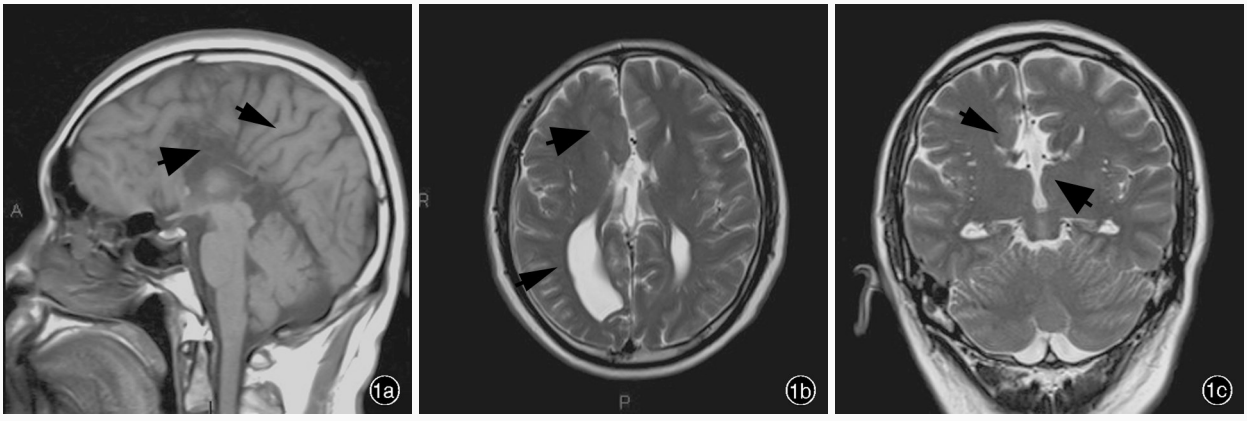

图1 男性患者，29岁。主因6天前癫痫大发作入院 1a 矢状位T_1WI显示胼胝体及扣带回缺如，第三脑室高位（粗箭头所示），大脑内侧面可见呈放射状排列的脑回指向第三脑室（细箭头所示） 1b 横断面T_2WI显示双侧侧脑室平行且间距增宽，双侧额角呈尖角样（粗箭头所示），右侧枕角扩张（细箭头所示） 1c 冠状位T_2WI显示类似于“麋鹿角”三叉样额角（粗箭头所示），以及呈“锁孔”样扩大的颞角，另可见呈纵向走行且致密的Probst纤维束（细箭头所示），半球间裂增宽

Figure 1 A 29-year-old male was admitted for major epilepsy 6 days ago. Sagittal T_1WI showed the absence of corpus callosum and cingulate gyrus, the high riding of the third ventricle (thick arrow indicates) and the radially arrayed gyri "pointing to" the third ventricle on the medial surface (thin arrow indicates, Panel 1a). Axial T_2WI revealed widely separated and parallel lateral ventricles. Bilateral pointed frontal horns (thick arrow indicates) and right dilated occipital horn (thin arrow indicates) were seen (Panel 1b). Coronal T_2WI showed trident-like frontal horns like "moose antler" (thick arrow indicates), keyhole-like temporal horn and compact longitudinally oriented Probst fasciculus (thin arrow indicates). The interhemispheric fissure was enlarged (Panel 1c).

胼胝体发育不良（dysgenesis of corpus callosum）为中枢神经系统常见先天性发育畸形，表现为胼胝体完全或部分节段缺如。在正常情况下，胼胝体纤维呈水平状排列。当胼胝体缺如时，来自大脑皮质的神经元轴突形成致密的由前向后纵向走行的Probst束，构成侧脑室内壁，因此双侧侧脑室内缘光滑且相互平行。胼胝体枕钳和压部发育不良可引起侧脑室后角、三角区和下角扩大，而且脑室系统由于失去胼胝体的支撑作用而出现双侧侧脑室分离、室间孔增宽，以及第三脑室顶向背侧抬高的征象。CT征象包括胼胝体完全或部分缺如；双侧侧脑室体部平行且间距增宽，额角窄小呈尖角样、枕角扩张；半球间裂增宽，并与第三脑室前方接近或相通；第三脑室扩大、上抬。MRI征象包括：（1）矢状位胼胝体全部或部分缺如（图1a）。（2）第三脑室高位，大脑半球内侧面脑沟随上移的第三脑室顶部呈放射状排列（图1a）。（3）双侧侧脑室体部平行且间距增宽，额角窄小呈尖角样，枕角扩张（图1b）。（4）冠状位颞角呈“锁孔”样扩张（图1c）。鉴别诊断：（1）广泛性脑白质疏松，可侵犯胼胝体而使纵裂增深增宽，需与胼胝体发育不良相鉴别。（2）脑积水，可引起胼胝体受牵拉而变薄，但不存在胼胝体某一节段的缺如。

（天津市环湖医院神经放射科韩彤供稿）