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·病例报告·

克拉屈滨治疗成人朗格汉斯细胞组织细胞增生症一例

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患者,男,73岁。入院前2年无明显诱因出现双眼视力进行性下降,未予重视。此后逐渐出现双眼球突出,于入院前1个月左眼失明,右眼光感,无发热、皮疹、骨痛。当地医院颅脑MRI平扫+增强:双侧眶内外侧部病变,性质待定,考虑炎性假瘤可能性大。行眼部手术,左眼眶内肿物病理:组织弥漫成片,浸润横纹肌,细胞体积较大,胞质丰富、淡染,核卵圆形、椭圆形或不规则形,核仁部分可见,难见核沟,可见核分裂,伴大量嗜酸性粒细胞、淋巴细胞浸润;免疫组化:CD68(部分+),S-100(部分+),CD20(部分+),CD79α(部分+),CD3(部分+),CD5(部分+),Ki-67(约20%+),CD1a(部分+),CD163(+),CD34(-),CK(-)。考虑朗格汉斯细胞组织细胞增生症。

入院查体:双眼球突出,眼睑水肿,粗测双眼视力:左眼失明,右眼光感,双侧下颌可触及单个黄豆大小淋巴结,质韧,无触痛,与周围组织无粘连,胸骨无压痛。PET/CT:双侧泪腺区及眶内软组织、右侧胸膜及全身多发淋巴结FDG代谢异常增高,结合病理考虑符合朗格汉斯细胞组织细胞增生症代谢改变。头颅MRI:①左侧眶内软组织(不规则形长T1等

稍短T2信号,大小约2.0 cm×0.9 cm),双侧眼直肌增粗,左侧显著,结合病史,考虑为朗格汉斯细胞组织增生表现;②左侧晶状体变薄。外周血细胞形态示:嗜酸性粒细胞10%。血常规、肝肾功能检查正常。根据患者临床表现、辅助检查、外院病理结果及累及部位,诊断:朗格汉斯细胞组织细胞增生症(多部位、多系统)。

予克拉屈滨9 mg/d×3 d化疗,化疗后观察血常规及肝功能等,未见明显不良反应,此后给予10 mg/d×5 d,每月1次,共计5个疗程,化疗3个疗程后眼球突出及眼睑水肿情况逐渐改善,视力较前缓慢改善。复查全身PET/CT:双侧泪腺区及眶内软组织及全身多发淋巴结FDG代谢异常增高,结合病理考虑符合朗格汉斯细胞组织细胞增生症治疗后改变,与治疗前PET/CT相比双侧泪腺区及眶内软组织大小及代谢程度相仿,全身多发淋巴结体积减小、数目减少。此后患者规律复查,病情平稳,未见新发病灶。

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