Diffuse Leptomeningeal Glioneuronal Tumor Involving Circle of Willis And Spinal Nerve Roots

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A 14-year-old girl had headache and intermittent crying and screaming for four months and upper back pain for ten days. On examination, she was confused and had bilateral lagophthalmos and hearing loss, neck rigidity, and decreased deep tendon reflexes in the limbs. CSF analyses showed an elevated level of protein (1628 mg/L), a decreased level of glucose (0.934 mmol/L) and a normal nucleated cell count, with increased intracranial pressure (>330 mmH₂O). Following brain and spine MRI (Figure 1) and brain CTA (Figure 2), she had a brain biopsy. A diagnosis of diffuse
leptomeningeal glioneuronal tumor (DLGNT) was pathologically confirmed (eFigure 1). Her parents abandoned treatment, she died two weeks later.

DLGNT is a newly defined entity in the 2016 WHO classification of CNS tumors[1]. Involvements of cerebral arteries and spinal nerve roots haven't been reported in DLGNT. We speculate that the thinning of Circle of Willis is resulted from tumor mass compression and tumor cell infiltration via leptomeninges or Virchow-Robin spaces[2].

Reference


Figure 1. The findings of brain and spine MRI

Gadolinium-enhanced T1WI shows two enhancing nodules near the proximal C4 segment of bilateral internal carotid arteries (arrows; A), extensive meningeal enhancement in the brain and spinal cord (arrows; B), and enhancement in the cervical nerve roots (arrows; C, D).
Figure 2. The findings of brain CTA

Brain CTA shows thinning of the Circle of Willis (arrows; A, B). CTP maps reveal markedly decreased perfusion in local regions of the right cerebral hemisphere (arrows; C, D).
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