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Malignant Idiopathic Intracranial Hypertension Revealed a Hidden Primary Spinal Leptomeningeal Medulloblastoma

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Abstract : Context: Frequently, the cause of raised intracranial pressure remains unresolved and rarely is related to spinal tumors, moreover less to spinal medulloblastoma without primary brain focus. Process: An 18-year-old woman had a 3-month history of headaches and impaired vision. Neurological examination revealed bilateral sixth cranial nerve palsies with bilateral papilloedema of grade III. No focal brain or spine lesion was found on imaging. Consecutive lumbar punctures showed high opening pressure and subsequent increasing protein level. The meningeal biopsy was negative. At one point, she developed an increasing headache, vomiting and back pain. Spine MRI showed diffuse nodular leptomeningeal enhancement with the largest nodule at T6–T7. Malignant cells were detected in cerebrospinal fluid. She underwent laminectomy with excisional biopsy, and pathology showed medulloblastoma WHO grade IV. Outcome: She was treated with chemotherapy and craniospinal irradiation and made a good recovery. Relevance: Primary spinal leptomeningeal medulloblastoma is extremely rare, especially without primary brain focus, but may cause increased intracranial pressure, even in the early microscopic phases, and it should be considered in the differential diagnosis if conventional and aggressive treatment of idiopathic intracranial hypertension fails. We assume that arachnoiditis from tumor seeding caused increased intracranial pressure. Appropriate neurosurgical intervention and surgical biopsy are mandated if a suspicious lesion is detected. Consider proper rescreening of the whole neuroaxis in refractory cases of intracranial hypertension.

Keywords : CNS infection, IIH, headache, primary spinal leptomeningeal medulloblastoma
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