Pilomyxoid astrocytoma of the cervical spinal cord in a child with rapid progression into glioblastoma: case report and literature review.

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Abstract

INTRODUCTION: Pilomyxoid astrocytoma (PMA) is a recently described glial tumor with similarities to pilocytic astrocytomas, yet with distinct histopathological characteristics and a more aggressive behavior. It occurs predominantly in the hypothalamic/chiasmatic region. Only four patients with spinal cord PMA have been reported in the pediatric population. The 2007 WHO Working Group recognized PMA as a new variant and recommended an assignment to WHO grade II.

OBJECTIVE: The purpose of this paper was to report a rare location, address the aggressive behavior and rapid progression, and based on the specific patient, to review the literature and discuss current treatment strategies.

CASE PRESENTATION: A 12-year-old girl presented with motor and sensory deficits of the left side as well as gait disturbance. Imaging revealed an intramedullary tumor extending from C2 to C7. The patient improved impressively after surgical resection. Histopathological findings were consistent with PMA. Three months later, the patient presented with rapid neurological deterioration. Histopathology after the second operation was consistent with glioblastoma. The outcome was fatal 12 months after initial diagnosis, despite adjuvant therapy. CONCLUSIONS: This is the fifth pediatric spinal cord PMA in literature. Furthermore, it is the only documented patient with rapid recurrence and progression within 3 months into a glioblastoma. The question of a sampling error affecting initial pathology is raised. Based on contemporary literature data, we discuss the further treatment options, as there are no guidelines yet. Efforts towards registries should be encouraged, as the documentation of PMA might lead to more evidence based treatment strategies.

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