ABSTRACT

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H3K27M-mutant glioma in thoracic spinal cord and conus medullaris with pilocytic astrocytoma morphology: case report and review of the literature.

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BACKGROUND: The H3K27M-mutant spinal cord gliomas are very aggressive with a dismal prognosis, very few cases have been reported in the thoracic spinal cord and conus medullaris, and it is extremely rare with morphological features of pilocytic astrocytoma.

CASE PRESENTATION: A 20-year-old man presented with thoracolumbar pain, progressive paraparesis, and urinary incontinence. Magnetic resonance imaging revealed an intramedullary solid-cystic lesion from D9 to conus medullaris. Subtotal resection was performed, restricted by the indistinct margins and the decline of the motor evoked potential during the surgery. Pathologic findings revealed a pilocytic astrocytoma with anaplastic features. However, a further assessment determined a diffuse midline glioma H3K27M-mutant, and adjuvant chemoradiotherapy was administered. After seven months of progression-free survival, the paraparesis worsened; at twelve months of follow-up, the patient developed paraplegia, and at 24 months the patient remains alive without any neurologic functions distal to the tumor and he is still under adjuvant treatment.

CONCLUSIONS: The H3K27M-mutant spinal cord glioma is a very infrequent tumor with a wide variety of histological presentations even as indolent as pilocytic astrocytoma, which should be considered in spinal cord tumors, especially if there are clinical, histological, or radiological data that suggest aggressiveness. On the other hand, the fast progression led to the loss of complete neurological function distal to the tumor, in spinal tumors could explain a not so poor prognosis as it is in functionally and vital structures.

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