Spinal extramedullary ependymoma

Yakar et al. reported the case of a patient with NF1 presenting with low back pain and gait disturbance. The patient had an spinal extramedullary ependymoma at the L2-3 vertebra level. This report is the first case of NF1 with spinal ependymoma localized in the lumbar region ¹⁾

Pomeraniec et al present a rare extramedullary ependymoma with diffuse spinal metastatic disease, and review the previous reports of extramedullary spinal ependymomas.

A 23-year-old man with a history of progressive neck, shoulder and arm pain, with sensory and motor symptoms in the C7 dermatome. MRI of the cervical spine demonstrated a ventral contrast-enhancing lesion with evidence of enhancement along the dura and spinal cord of the upper cervical spine, thoracic spine, and cauda equina. He underwent a tumor debulking procedure without complications. Following surgery, he received craniospinal radiation to treat the remaining tumor and diffuse leptomeningeal disease. The final pathology of the tumor revealed that is was a World Health Organization Grade III anaplastic ependymoma. At the 1 year follow-up, the patient had stable imaging and had returned to his preoperative functional status. Of the 19 reported patients with primary intradural, extramedullary spinal ependymomas, two had extradural components and seven had anaplastic grades. Only one tumor with an anaplastic grade resulted in metastatic disease, but without spinal recurrence.

This is the first report of an intradural, extramedullary spinal ependymoma with an anaplastic grade, presenting with concomitant diffuse, nodular leptomeningeal metastases involving the upper cervical spine, thoracic spine, conus medullaris, and cauda equina. Similar to the treatment of intramedullary ependymomas with metastases, this patient underwent an aggressive debulking procedure followed by radiation therapy to the entire neuroaxis ²⁾.

1)

Yakar H, Ertugrul B, Kaplan M. A rare tumor case in an adult patient with neurofibromatosis: Lumbar ependymoma. Niger J Clin Pract. 2022 Feb;25(2):197-199. doi: 10.4103/njcp.njcp_79_21. PMID: 35170447.

2)

Pomeraniec IJ, Dallapiazza RF, Sumner HM, Lopes MB, Shaffrey CI, Smith JS. Anaplastic extramedullary cervical ependymoma with leptomeningeal metastases. J Clin Neurosci. 2015 Dec;22(12):1871-6. Epub 2015 Oct 23. PubMed PMID: 26601808.

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