



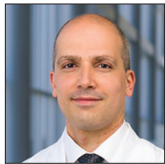
Image Report

Aggressively recurring cervical intramedullary anaplastic astrocytoma in a pregnant patient

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ABSTRACT

Background: Many patients with spinal juvenile pilocytic astrocytoma can experience prolonged remission after resection. However, some reports suggest that pregnancy may be associated with progression.

Case Description: The authors provide an image report highlighting a case of rapid and aggressive transformation of an intramedullary astrocytoma of the cervical spine in a pregnant patient. Over the course of 1 year, the lesion progressed from a juvenile pilocytic astrocytoma to an anaplastic astrocytoma. Genetic testing revealed mutations associated with aggressive behavior.

Conclusion: The case and associated imaging demonstrate the importance of close neurologic monitoring and counseling regarding risk of progression in pregnant patients with spinal gliomas.

Keywords: Anaplastic astrocytoma, FGFR1, Intramedullary, Pilocytic astrocytoma, SETD2, TACC1

INTRODUCTION

While gliomas account for ~80% of primary malignant brain tumors, primary glioma of the spine remains a rare finding.^[1] Median survival ranges from 10 to 17 months for high-grade gliomas of the spine,^[4] and progressive neurologic and respiratory compromise contribute to high rates of morbidity and mortality. It is suspected that metabolic and hormonal changes related to pregnancy can influence cranial glioma progression,^[4] but minimal evidence exists illustrating this phenomenon in spinal gliomas. As such, we present an image report focused on the rapid progression and malignant transformation of a low-grade cervical glioma in the setting of pregnancy.

CLINICAL IMAGE

A woman with a progressive left hemiparesis and diffuse paresthesias presented for outpatient neurosurgical evaluation. Magnetic resonance imaging (MRI) of the cervical spine revealed a peripherally enhancing intramedullary lesion spanning from the medulla to C5 [Figure 1a]. After extensive discussion of the risks and benefits, the patient consented for and underwent a suboccipital craniectomy with C1–C5 laminectomy and C2–C5 laminoplasty for tumor

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Figure 1: (a) Sagittal MRI T1 sequence after gadolinium administration showing a peripherally enhancing intramedullary spinal mass spanning from the brainstem to C5. (b) Postoperative MRI after the initial resection showing a sagittal T1 sequence after gadolinium administration and minimal residual disease at the level of C3. (c) Sagittal MRI after gadolinium administration showing significant recurrence of the tumor with diffuse enhancement and cyst formation compressing the brainstem, within less than a year after her first surgery. (d) Sagittal MRI T2 sequence showing diffuse intramedullary edema spanning from the brainstem to the thoracic T2 segment, and worsening swan-neck deformity. (e) Sagittal MRI T1 sequence with gadolinium administration after the second tumor resection surgery showing decompression of the cyst with minimal peripheral residual enhancing mass. (f) Lateral postoperative X-ray of the cervical spine showing posterolateral instrumentation with restitution of lordosis.

resection. Postoperatively, her left hemibody strength returned, and her MRI revealed evidence of near-total resection with minimal residual disease [Figure 1b]. Histopathologic examination of the lesion identified a World Health Organization (WHO) Grade 1 juvenile pilocytic astrocytoma, with an MIB-1 proliferation index of $<0.5\%$ and no evidence of mitotic activity. She was counseled regarding the importance of continued surveillance, and future imaging and clinic follow-up were planned. She became pregnant in the weeks following her surgery, and her pregnancy limited her ability to attend her scheduled follow-up appointments.

After delivery, she noted progressively worsening weakness in her left hemibody. Of note, this weakness progressed more rapidly compared to her initial presentation. She underwent an MRI of the cervical spine, which revealed extensive disease recurrence and a new large cystic component of the tumor that compressed her medulla [Figure 1c]. In addition, her imaging demonstrated satellite lesions throughout the subaxial cervical spine, significant edema spanning from the brainstem to the upper thoracic spine [Figure 1c and d], and a progressive swan-neck deformity. She underwent additional posterior cervical decompression and tumor debulking with cyst aspiration to relieve the compression

on the medulla. Notably, tumor debulking was intentionally conservative to prevent further neurologic decline. Given the planned extent of decompression and her progressive kyphosis, she also underwent C2–T2 posterolateral fusion for stabilization. Postoperatively, her neurologic function remained stable.

Her postoperative MRI revealed decreased tumor size, medullary decompression, and improved cervical alignment [Figure 1e and f]. Histopathologic examination revealed malignant transformation to a WHO Grade III anaplastic astrocytoma, along with a markedly increased MIB-1 index (25%) and very high mitotic activity. Next-generation sequencing identified fusion of *FGFR1* and *TACC1* genes with constitutive activation of *FGFR1*, as well as loss of function variant in the tumor suppressor gene *SETD2*. These changes can be seen in glioblastoma multiforme.^[2,3,6] She is currently being assessed for her eligibility to participate in ongoing clinical trials.

DISCUSSION

Case reports implicate pregnancy in the rapid transformation of gliomas.^[4,5] Proposed mechanisms underlying this association include stimulation of glial cell growth from

increased levels of estrogen, progesterone, and placental growth factor,^[4] promotion of neovascularization from pregnancy-induced hemodynamic changes, and an association between tumor growth and fluid retention during pregnancy. Existing literature also suggests that genetics may influence the association between pregnancy and malignant transformation. Further examination of this phenomenon is warranted.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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