Primary Gliosarcoma of the Optic Nerve: A Unique Adult Optic Pathway Glioma.

Cimino PJ, Sychev YV, Gonzalez-Cuyar LF, Mudumbai RC, Keene CD.

Abstract

A 90-year-old woman presented with 1-year history of right-sided progressive proptosis, neovascular glaucoma, blindness, and worsening ocular pain. No funduscopic examination was possible because of a corneal opacity. Head CT scan without contrast demonstrated a heterogeneous 4.1 cm (anterior-posterior) by 1.7 cm (transverse) cylindrical mass arising in the right optic nerve and extending from the retrobulbar globe to the optic canal. She underwent palliative enucleation with subtotal resection of the orbital optic nerve and tumor. Pathological examination showed effacement of the optic nerve by an infiltrative high-grade glial neoplasm with biphasic sarcomeric differentiation. Invasion into the uvea and retina was present. The neoplasm was negative for melan-A, HMB45, tyrosinase, synaptophysin, smooth muscle actin, and epithelial membrane antigen. The glioma had strongly intense, but patchy immunopositivity for glial fibrillary acidic protein. Multiple foci of neoplastic cells had pericellular reticulin staining. The overall features were diagnostic of a gliosarcoma (World Health Organization grade IV) of the optic nerve. Postoperative MRI demonstrated postsurgical changes and residual gliosarcoma with extension into the optic chiasm. The patient died 2 and a half months after her enucleation surgery at her nursing home. Autopsy was unavailable due to the caregiver wishes, making a definitive cause of death unknown. Gliosarcoma is a rare variant of glioblastoma, and this is the first documented case presenting as a primary neoplasm of the optic nerve.

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