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Recurrent Schwannoma Postirradiation:

Histological Review Reveals Mixed Schwannoma and Meningioma

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Vestibular schwannomas (VS) are benign tumors that emerge from Schwann cells ensheathing the eighth cranial nerve.¹ Meningiomas arise from the meninges and are usually benign.² These tumors, occurring separately, are the 2 most common intracranial tumors.³ The occurrence of a meningioma and a schwannoma within the same tumor is very rare, with only a few cases reported.⁴ We present a unique case of a mixed schwannoma and meningioma in the left cerebellopontine angle (CPA). This case illustrates a postirradiation recurrent tumor in a 64-year-old woman. Immunostaining revealed that the tumor was a mixture of schwannoma and meningioma. We propose that the patient was probably mosaic for the *NF2* gene mutation, due to the presence of 2 other ipsilateral meningiomas, which qualifies her for the diagnosis of neurofibromatosis type 2 (NF2) by National Institutes of Health (NIH) criteria (1988). In addition, her daughter developed a unilateral vestibular schwannoma in her fourth decade. Interestingly, proliferating cell nuclear antigen (PCNA) and vascular endothelial growth factor (VEGF) staining were highly correlated, suggesting that VEGF expression induced tumor survival after irradiation.

Case History

A 50-year-old woman underwent subtotal resection of a left CPA tumor via retrosigmoid craniotomy at an outside facility. Within 2 years, the tumor regrew, and the patient underwent gamma knife radiosurgery at age 53, also done elsewhere. Following radiation, the tumor initially remained stable at 2 cm. At age 57, the patient developed left facial weakness. At age 61, she underwent left hypoglossal to facial nerve anastomosis to treat progressive facial weakness. Magnetic resonance imaging (MRI) after the procedure revealed that the patient's residual schwannoma was still stable at 2 cm.

At age 64, the patient underwent a follow-up MRI scan that revealed that her residual schwannoma had significantly increased in size, and 2 intracranial meningiomas had also developed: one in the posterior fossa and one in the midline prepontine area (Figure 1). At that time, the patient reported dizziness and loss of balance that began 3 months prior. It was recommended that she undergo surgical removal of the 3 tumors because of progressive tumor growth and symptoms, as well as to avoid malignancy because of her history of radiation therapy.

Preoperative audiogram results revealed a profound sensorineural hearing loss in the left ear. Preoperative MRI results revealed a large tumor in the left CPA whose cisternal component measured 24 × 16 mm. The tumor also filled the left internal auditory canal and a portion of the labyrinth (Figure 1).

Postoperative surgical findings included a typical appearing meningioma of the foramen magnum and left cerebellum, as well as a densely fibrotic recurrent tumor of the CPA,

which was diffusely adherent to the leptomeninges and to the surface of the cerebellar peduncle and brainstem, encasing the internal carotid and posterior communicating artery and lower cranial nerves. A subtotal resection of the CPA tumor was achieved, leaving tumor fragment adherent to the brainstem.

Histopathology

Tumor tissue was collected under St Vincent Hospital Institutional Review Board approval. The specimen was stained with hematoxylin and eosin (H&E), PCNA, S100, and VEGF. Breast cancer tissue was used as a positive control for PCNA, S100, and VEGF stains (not shown). Histopathological findings revealed a mixture of meningioma schwannoma within the tumor: schwannoma stained positive for S100 and meningioma was S100 negative (Figure 2G–I). The PCNA stains revealed that meningioma and schwannoma along the periphery were proliferating (Figure 2D–F). The VEGF stains correlated well with the PCNA stains: the areas that stained positive for PCNA also stained positive for VEGF (compare D–F and J–L), suggesting a role for VEGF in tumor regrowth and/or survival. Figure 2 reveals that the meningioma and schwannoma tissues are differentially proliferating (compare D–F with G–I).

Discussion

This is a unique case describing a postirradiation recurrent VS that was found upon histological review (ie, S100 staining) to also contain meningioma. Although she qualifies for a diagnosis of NF2, this patient is suspected of being mosaic for the *NF2* gene mutation because of multiple tumors arising on the left side (Figure 1) and a daughter with a unilateral VS.⁵ The VEGF and PCNA staining revealed that both meningioma and schwannoma were viable. The high degree of VEGF expression within the tumor is correlated to a high tumor proliferation rate, supporting a role for anti-VEGF therapy for NF2-related VS.

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Figure 1.

T1-weighted post contrast magnetic resonance imaging demonstrates a left cerebellopontine angle lesion consistent with meningioma (A); coronal view also demonstrates the foramen magnum lesion (B), and inferior axial view shows both ipsilateral meningiomas (C).

Figure 2.
Hematoxylin and eosin (A, B, C), Immunostaining: proliferating cell nuclear antigen (PCNA) (D, E, F), demonstrating proliferation (asterisk); S100 (G-I), meningioma negative (arrow), schwannoma positive (H, I); and vascular endothelial growth factor (VEGF) positivity where proliferating (J, K, L).