A Case of Multiple Neurofibroma/Schwannoma Hybrid Tumors of the Facial Nerve

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Schwannomas and neurofibromas are benign peripheral nerve sheath tumors (BPNSTs) that may be implicated in neurofibromatoses such as neurofibromatosis type 1, neurofibromatosis type 2 (NF2), and the more recently described schwannomatosis.1 When multiple schwannomas present in a single patient, their role in one of the aforementioned syndromes becomes much more likely.2 The latter of these syndromes, schwannomatosis, involves multiple schwannomas of peripheral and cranial nerves with a predilection toward nerves of the head and neck region.1,3 Neurofibroma/schwannoma hybrids are rare tumors that have been implicated in both schwannomatosis and neurofibromatosis and favor a syndromic rather than sporadic diagnosis.2 Up to 71% of definitively diagnosed patients with schwannomatosis have at least 1 hybrid tumor, and 61% of all peripheral nerve tumors have been shown to demonstrate hybrid pathology.1 There has yet to be a report of multiple neurofibroma/schwannoma hybrid tumors of the facial nerve (CN VII). Here we describe a case in which a 49-year-old man had such a presentation. Approval for this report was attained from the Capital Health Research Ethics Board.

Case Report

This man presented to the QEII Health Sciences Center with a history of increasing sensory loss and weakness. Guillain-Barré syndrome was diagnosed, and upon admission to neurology, it was incidentally discovered on magnetic resonance imaging that he had multiple right-sided parotid masses, the largest of which measured 2.0 × 1.6 cm (Figure 1). Examination by otolaryngology–head and neck surgery demonstrated masses in his right parotid area that were nontender to palpation, although they did cause him prandial pain. Facial nerve function was normal. Fine-needle aspirate (FNA) was performed and demonstrated a possible benign mesenchymal tumor, pleomorphic adenoma, or myoepithelial neoplasm. A right superficial parotidectomy was undertaken, and 4 individual neurogenic tumors were removed, all of which originated from CN VII. There were 2 separate tumors on the main trunk and pes region, but the largest was removed from the temporal branch. The final tumor originated from the lower division. Pathology

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Figure 1. Nonenhanced, T2-weighted, axial magnetic resonance imaging of the head demonstrating multiple right-sided parotid masses initially reported as lymph nodes but subsequently identified as hybrid neurofibroma/schwannoma tumors.
identified these tumors as benign peripheral nerve sheath tumors most consistent with hybrid neurofibromas/schwannomas (Figure 2).

Discussion

Cases in which multiple hybrid neurofibromas/schwannomas present raise the clinical suspicion of the presence of one of the neurofibromatoses. In our patient, the lack of vestibular nerve involvement, the absence of a first-degree relative with NF2, and the lack of other NF2 stigmata or associated NF2 mutations suggest the presence of schwannomatosis.

Pathologically, these hybrid tumors are defined as having components of neurofibromas and schwannomas, thus differentiating them from other BPNSTs. The schwannoma-like part of the tumor usually contains Verocay bodies with Schwann cells demonstrating nuclear palisading. The neurofibroma-like part may demonstrate abundant fibroblasts, collagen, and myxoid change with an elongated and wavy appearance, as demonstrated in Figure 2. Immunohistochemistry demonstrates positivity for both S100 protein in the Schwann cells and neurofilament in residual entrapped axons. The presence of axons within the tumor is a key differentiating feature from pure schwannoma, which lacks axons or fibroblasts.

The median age of diagnosis for schwannomatosis is 40 years and has not been shown to decrease survival. Pain is the most common presenting symptom (57%), followed by the presence of a mass (41%), although focal numbness and weakness have also been described. Correlating with this diagnosis, our patient had right-sided prandial pain and a progressively growing right-sided facial mass.

Our case is unique in that there were multiple hybrid neurofibromas/schwannomas involving various branches of CNVII in the parotid area. Although a single neurofibroma/schwannoma hybrid tumor of CNVII has been reported, there has yet to be a report of multiple such tumors. This presentation poses challenges in diagnosis as the differential is expanded to include common parotid masses as demonstrated by the FNA described herein. Multiple cranial nerve tumors in close proximity also add further intricacy to surgical management, thus increasing the risk of complications. With this in mind, surgical excision is the only definitive management for these BPNSTs. Radiation is reserved for schwannomas not amenable to surgery, and an effective chemotherapeutic agent has yet to be developed. Although seemingly rare, recurrence rates and potential for malignant transformation are as yet unidentified for hybrid tumors.

In conclusion, parotid mass and prandial pain may be a presentation of multiple hybrid neurofibromas/schwannomas of the facial nerve. This presentation garners unique diagnostic and surgical considerations and may represent an underlying schwannomatosis.

Author Contributions

Scott Murray, gathered patient information including chart review for this case report, prepared initial manuscript for case report and contributed to further revisions, edited final manuscript to meet criteria set out by the journal for final submission; Martin J. Bullock, provided pathological analysis and photomicrographs of tumor specimens for this report, contributed to manuscript revisions and final approval of manuscript; S. Mark Taylor, surgeon involved with this case performing the surgery herein, conceived case report, contributed to manuscript revisions, and provided final approval of manuscript.

Disclosures

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