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# 24-YEAR-OLD WOMAN WITH AN INTERNAL AUDITORY CANAL MASS

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## CLINICAL HISTORY

A 24-year-old woman with a 3 year history of multiple sclerosis, for which she was treated with beta interferon, was found to have a T2 intermediate mass (1.3 cm × 0.7 cm) within the left internal auditory canal (Figure 1) in a follow up MRI. There was no associated mass effect on the adjacent brain parenchyma. In addition, there were numerous T2 hyperintense lesions throughout the supratentorial white matter, consistent with the known history of multiple sclerosis. At the time, she did not have any related symptoms; there was no reported abnormality of hearing or balance, and no facial nerve dysfunction. On examination, visual fields and acuity were normal and cranial nerves II through XII were intact, with normal hearing on both sides. Motor-sensory skills were normal.

The patient underwent surgery (suboccipital approach) with an attempt to preserve hearing and facial nerve function. The surgical intervention was successful, with complete removal of the lesion and preservation of hearing and facial functions.

## NEUROPATHOLOGIC FINDINGS

At gross examination, the lesion was a nodular, well circumscribed mass measuring 1.1. × 0.9 cm. It was tan-white and firm. Histologic examination demonstrated a neoplasm composed of spindled cells with bland, tapered, and at times "wavy", nuclei. The tumor had a biphasic pattern. The predominant pattern is a fascicular growth pattern with focal nuclear palisading around nuclear free areas (Figure 2). The second, minor component shows concentric proliferation of neoplastic spindle cells around one or more axons, with formation of pseudo-onion bulbs (Figure 3). These proliferating cells have elongated to oval nuclei, delicate chromatin, and inconspicuous nucleoli. Mitoses and necrosis are not recognized in either component. Degenerative atypia with nuclear hyperchromasia is focally observed.

Immunohistochemical studies revealed the fascicular component is diffusely positive for S100 protein, while the area with the concentric proliferation of cells was positive for the perineurial markers claudin-1 (Figure 4a) and Glut-1. As expected, the axons surrounded by the concentric rings of cells were highlighted by neurofilament immunostaining (Figure 4b). In addition, antibody anti-CD34 was also focally positive in the tumor (Figure 4c). **What is the diagnosis?**

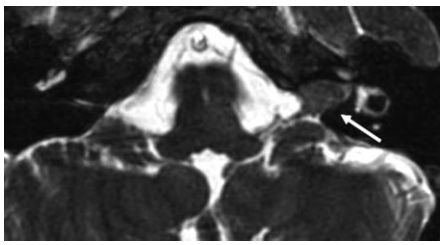

**Figure 1.**

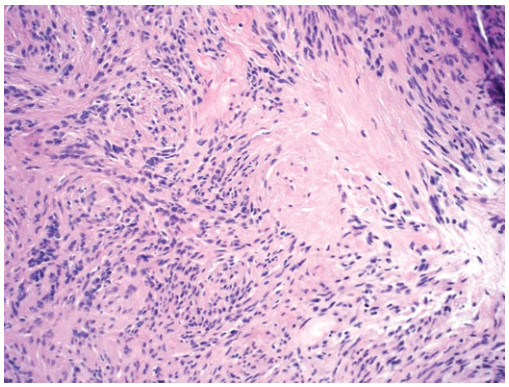

**Figure 2.**

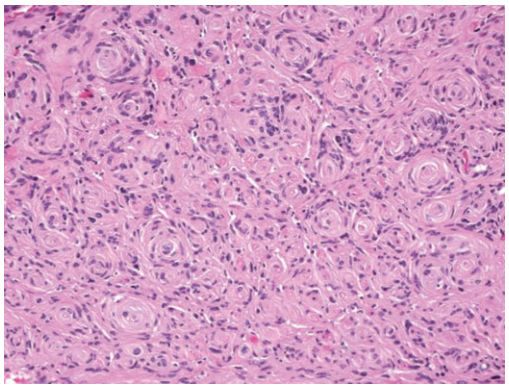

**Figure 3.**

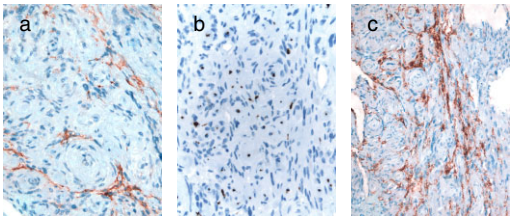


**Figure 4.**

## DIAGNOSIS

Hybrid peripheral nerve sheath tumor with schwannoma/perineurioma components.

## DISCUSSION

Benign peripheral nerve sheath tumors are classically divided into schwannomas, neurofibromas, and perineuriomas (8). Their diagnosis is usually straightforward, given the unique morphological and immunohistochemical characteristics of each type. In recent years, peculiar tumors with hybrid features of these entities have been described. Histologically, most of these tumors revealed a biphasic pattern, showing an abrupt transition between the two components.

Since the first report of an hybrid peripheral nerve sheath tumor in 1998 (2), a few cases have been described, with the great majority arising in peripheral nerves of the trunk and extremities (2, 4, 5, 7–9). Uncommon locations, such as colon, nasal cavity, orbit and lymph node, have also been reported (1, 3, 6, 10). However, there is no report to date, of an intracranial hybrid tumor arising from a cranial nerve. We report an unusual case of a hybrid peripheral nerve sheath tumor arising from the VIIIth cranial nerve. The tumor shows a hybrid, biphasic appearance, with a predominantly schwannoma component, and a minor perineurioma component. This hybrid nature is confirmed and highlighted by immunohistochemical staining profiles, with S100 positivity of the schwannoma component and immunostaining of the perineurioma component for claudin-1, Glut-1 and CD34.

Hybrid peripheral nerve sheath tumors usually have a benign clinical course with only exceptional recurrences, similar to conventional benign schwannomas and perineuriomas. No association with neurofibromatosis has been identified.

The histological differential diagnosis includes spindle cell lesions such as schwannomas, perineuriomas, neurofibromas and low-grade malignant peripheral nerve sheath tumor. Conventional schwannomas and perineuriomas are composed of a single cell type (Schwann cells and perineurial cells, respectively). Schwannomas are characterized by loose and dense areas (Antoni A and Antoni B) and often have Verocay body formations. The conventional intraneural perineurioma consists of whorled tight proliferations of cells around one or more axons. Neurofibromas often have collagen bundles and myxoid background and are composed of axons and multiple cell types intimately mixed within the tumor. The diagnosis of a hybrid peripheral nerve sheath tumor is made in the presence of two distinct histological areas and is confirmed by different patterns of immunostaining of the two components. As in schwannomas, degenerative nuclear atypia should not be confused with malignant transformation (MPNST) and is seen in the absence of any other anaplastic features (mitosis, necrosis, increased cellularity). The pathogenetic basis of the dual differentiation seen in hybrid lesions is poorly understood.

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## ABSTRACT


Benign peripheral nerve sheath tumors are divided into schwannomas, neurofibromas and perineuriomas. In recent years, tumors with hybrid features, composed of multiple, discrete areas of different histological types, were described. These tumors may represent a diagnostic challenge. A 24-year-old woman with multiple sclerosis was found to have a 1.3 cm TV × 0.7 cm AP T2 intermediate lesion within the left internal auditory canal. Gross examination revealed a tan-white, well circumscribed mass. Histologic examination demonstrated a well demarcated, cellular, solid neoplasm with a biphasic pattern. Most of the tumor was composed of spindle cells arranged in fascicles with focal Verocay body formation and diffuse S100 positivity. A second, minor area showed concentric proliferation of neoplastic spindle cells around one or more axons. Tumor cells in this area were positive for perineurial markers, claudin-1 and Glut-1, and focally immunopositive for CD34. We present here a case of a benign peripheral nerve sheath tumor with histological and immunohistochemical features consistent with a dual pattern of differentiation of schwannoma and perineurioma, in the VIIIth cranial nerve. This is, to our knowledge, the first case of a hybrid perineurioma/schwannoma reported in a cranial nerve.