

# Concomitant Ependymoma and Schwannoma of the Lumbar Spine Without Neurofibromatosis

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**Abstract:** The coexistence of 2 or more different tumors of the central neural axis has been defined and is well known in patients with neurofibromatosis-2 (NF-2), an inherited autosomal dominant disorder caused by defects in chromosome 22. The presence of multiple histologically different spinal tumors in the absence of NF-2 is extremely rare, however. The authors describe a patient with backache, in whom an ependymoma and a schwannoma of the lumbar spine were found in the absence of NF-2.

**Key Words:** coexisting spinal tumors, neurinoma, neurofibromatosis, spinal ependymoma, spinal schwannoma

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After the definitive histopathologic diagnosis of our patient was made, my (TT) professor (MH) said to me: “this case is very rare because it is unusual to see two histopathologically different spinal lesions in the same patient without neurofibromatosis-2 (NF-2). Please search the pertaining literature and report the case.” A search of the English literature did not reveal any report demonstrating the coexistence of spinal ependymoma and schwannoma in the same patient in the absence of NF-2.

Most cases of 2 histologically different tumors have been reported in the head and, rarely, in the spine. Ependymomas are the most common primary spinal tumors of the lower spinal cord and have a benign clinical course when treated with complete surgical resection.<sup>1,2</sup> Spinal cord schwannomas are also rare tumors and account for 0.3% of all intraspinal tumors and 1.1% of all spinal schwannomas.<sup>3</sup>

We report on a patient in whom 2 intradural extramedullary lesions of the lumbar spine were found, an ependymoma and a schwannoma, in the absence of NF-2.

## CASE REPORT

### History

A 45-year-old woman was admitted to our department with a 2-month history of severe low back pain radiating to the right leg.

She was previously admitted to the neurology department with the complaint of headache, but magnetic resonance imaging (MRI) of the head showed nothing abnormal.

### Examination

The results of her clinical examination were normal, with no signs of NF. The results of the neurologic examination were also normal. MRI of the lumbar spine demonstrated an intradural extramedullary mass 0.5 cm to 1 cm in diameter at the level of the inferior end plate of the L2 vertebra. The mass appeared isointense on T1-weighted images and inhomogeneously hyperintense on T2-weighted images (Fig. 1A); it enhanced homogeneously with contrast administration (see Figs. 1B, C).

### Operation and Postoperative Course

An L2 laminectomy and dural opening were performed, and a small mass 2 mm in diameter was noted. The mass was a well-encapsulated lesion; it was dissected from the surrounding neural tissue and completely removed. The existence of this lesion was unexpected for us because it was absent on the preoperative MRI. In addition, the size of the lesion as seen under the microscope was not consistent with the size of the lesion demonstrated on preoperative MRI. We then looked at the L3 lamina and recognized a distinct lesion almost 0.5 cm in diameter, which was consistent with the appearance of the lesion on preoperative MRI. The second lesion, situated in an extramedullary position, was also excised completely. The pathologic findings were as follows. The first lesion was an ependymoma (Fig. 2A) and the second lesion was a schwannoma (World Health Organization [WHO] grade II; see Fig. 2B). The postoperative course of the patient was uneventful, and she was discharged from the hospital on the third postoperative day.

## DISCUSSION

The clinical, radiologic, and intraoperative findings demonstrated under the operative microscope in this case provided data that should be taken into account. First, the presence of 2 histologically distinct spinal tumors increased the possibility that this patient might have NF-2. Neither the clinical findings nor MRI of the head provided a clue suggestive of the presence of NF-2 in our patient, however. Therefore, it is possible to encounter in clinical practice the coexistence of 2 histopathologically distinct and rare tumors of the spinal cord in the same patient without NF-2. Second, the existence of conflict between the MRI and intraoperative findings in the same patient may come as a surprise. The ependymoma in this patient was not detected with a high-quality MRI study. We hypothesize that the lesion was not appreciated because it was composed of a thin layer of tumor cells that preoperative MRI was not able to detect.<sup>4</sup> Third, the concomitant presence of

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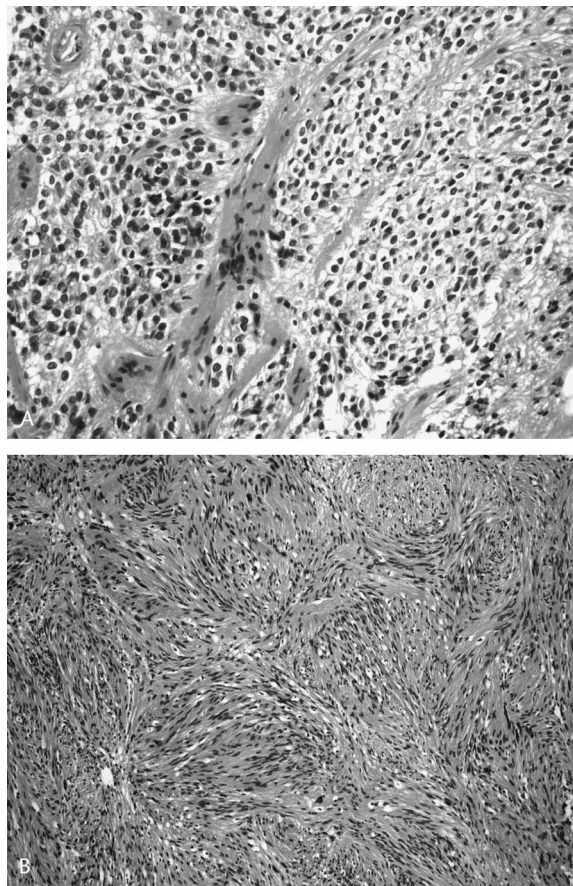


**FIGURE 1.** A, Sagittal T2-weighted image shows inhomogeneously hyperintense mass at the L2 level. Sagittal (B) and axial (C) postenhanced images of the same lesion demonstrate homogeneous enhancement.

schwannoma and ependymoma, which are derived from different cell types, is extremely rare in the absence of NF-2, as in our patient. Our explanation is mainly dependent on the hypothesis provided by Heuschling et al<sup>5</sup> that the ependymoma may have developed in response to the microtrauma and irritation produced by the schwannoma.

Since 1975, there have been 10 case reports on 2 histologically different spinal tumors in the same patient without evidence of NF.<sup>6-15</sup> Table 1 provides a summary of these reports found in the literature on the coexistence of distinct spinal tumors in the absence of NF.

Meningiomas and schwannomas represent approximately 50% of all spinal tumors; however, the occurrence of these tumors without clinical signs of NF is rare. There are 4 such cases reported in the literature.<sup>9,10,12,13</sup> The coexistence of these tumors may be coincidental or might be attributed to a common origin of the Schwann cells and the meningocytes.<sup>13,16</sup> Epidural cavernoma associated with a vertebral angioma at the thoracic level was also reported.<sup>7</sup> A common benign tumor of the spine, osteoblastoma, and a rare tumor of the bone, heman-gioendothelioma, have been demonstrated in the posterior elements of 2 separate thoracic vertebrae in the same patient without evidence of NF.<sup>8</sup> The first case with the coexistence of different spinal tumors in the absence of NF was reported by Hockley<sup>6</sup> in 1975. The author described an extraordinary case of breast carcinoma that had metastasized to a spinal meningioma located at the cervical spine. In a recent article, Glasauer et al<sup>15</sup> reported on a single patient harboring extradural metastatic adenocarcinoma of unknown origin and intradural pigmented schwannoma at 2 separate thoracic levels. This occurrence was considered to be coincidental. Another interesting binary case was reported by Shen and Lee,<sup>14</sup> who had a female patient in whom spinal intradural meningioma was found together with an intramedullary ependymoma. They



**FIGURE 2.** Photomicrographs show ependymoma (WHO grade II) with characteristic perivascular pseudorosettes (A) and a schwannoma with alternating arrangements of spindle nuclei and cytoplasmic processes (B).

**TABLE 1.** Summary of the Patients Reported in the Literature Who Harbored Histologically Distinct Spinal Tumors in the Absence of Neurofibromatosis

Reference No.	Author(s)	Age (y)/Sex	Spinal Level	Tumor Type
6	Hockley, 1975	72/F	C2	ID meningioma
			C2	ID metastasis
7	Lechevailier et al, 1979	27/M	T8	Ver angioma
			T8	ED cavernoma
8	Lange et al, 1986	19/M	T10	Ver hemendot
			T12	Ver osteoblastoma
9	Angiari et al, 1987	61/M	L2–3	ID meningioma
			L5	ID schwannoma
10	Araszkiwicz et al, 1988	22/F	T8–9	ID meningioma
			L2	ID schwannoma
11	Rabieal and Padilla, 1988	20/M	C3–6	IM hemblast
			C3–6	IM astrocytoma
12	Pagni et al, 1989	46/F	T10	ID meningioma
			T11	ID schwannoma
13	Dorizzi et al, 1992	70/F	C7	ID schwannoma
			T2–3	ID meningioma
14	Shen and Lee, 1992	67/F	C6–7	IM ependymoma
			T12–L1	ID meningioma
15	Glasauer et al, 1999	72/M	T10–11	ID schwannoma
			T12	ED metastasis
	Present case	45/F	L2	ID schwannoma
			L2	ID ependymoma

ED indicates extradural; F, female; Hemblast, hemangioblastoma; Hemendot, hemangioendothelioma; ID, intradural; IM, intramedullary; M, male; Ver, vertebral.

reported the simultaneous occurrence of more than 1 spinal tumor in the same patient to be at a rate between 1% and 4% of all spinal tumors. The presence of astrocytoma of the spinal cord and hemangioblastoma of the brainstem in the same patient is also extraordinary without evidence of NF.<sup>11</sup>

### CONCLUSION

The occurrence of 2 or more histologically different spine or spinal cord lesions in the same patient without evidence

of NF is extremely rare. We add 1 more case to the list of these rare tumors, and the lesson that we learned from the limited number of such cases is that although the coexistence of schwannoma and meningioma can be explained partly by their common origin from the same mesenchymal cell, there is currently no explanation for the simultaneous existence of different spinal tumors other than simple coincidence.

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