Cervical Intramedullary Schwannoma with Syrinx: Case Report and Review of the Literature

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Abstract

Schwannoma is a nerve sheath tumor originating from the Schwann cell. It is benign in nature and it arises from anywhere where Schwann cells can be found. It is rarely found in the parenchyma of the spinal cord. Intramedullary schwannomas (or neurilemmomas) without evidence of neurofibromatosis are rare spinal cord tumors. Intramedullary schwannoma was first reported in 1932 by Penfield. Our patient presented with neck pain, gradually worsening, weakness in the right upper and lower limbs, numbness in both shoulders, and a decrease in the grasping strength of both hands over a 4-year period. A magnetic resonance imaging of the spine showed a heterogeneously enhancing mass in the cervical spinal cord extending from the C2 to T1 levels with associated hemorrhagic changes. Histologically, the tumor was found to be composed of bland spindle cells with blunt-ended and sometimes wavy nuclei admixed with hyalinized vasculature. Surrounding reactive spinal cord parenchyma with frequent Rosenthal fibers was also observed. Focal Verocay bodies were evident, and with immunohistochemistry, there was diffuse and strong positivity for S100, which is confirmatory for the diagnosis of schwannoma. We report a case of cervical intramedullary schwannoma presented with syringobulbia in a young adult.

Keywords

- intramedullary schwannomas
- syrinx
- spinal cord tumor
- cervical

Introduction

Schwannoma is a nerve sheath tumor originating from the Schwann cell. It is benign in nature and arises from anywhere where Schwann cells can be found.[1] It is rarely found in the parenchyma of the spinal cord.[2] Because Schwann cells do not exist in the central nervous system. Intramedullary schwannoma was first reported in 1932 by Penfield.[3] One of the theories to explain the development of the tumor in this location is that it arises from the small bundles of peripheral nerves in the periphery of vasculature within the spinal cord.[4,5] Other theorized origins are from anterior and posterior nerve roots that have extensions inside the spinal cord, or from metaplastic cells of the pia mater that may have differentiated into Schwann cells, or from neural crest cells that may have migrated to the spinal cord during fetal development.[4,6] Most of the reported spinal cord schwannomas are found to be extramedullary. They have also been observed in the extradural space (25%) and as a combination of intradural and extradural lesions (15%); rarely have they been reported to be intramedullary.[5,7] Intramedullary schwannoma accounts for almost 0.3 to 1.5% of all primary intraspinal lesions.[8] It commonly involves the cervical region (61%) and, to a lesser extent, the thoracic (29%), and lumber (10%) regions.[1] Infrequently, they are associated with syringomyelia.[4] To make a diagnosis of intramedullary schwannoma and to differentiate it from other neoplasms by imaging only is almost impossible.[4] We can suspect an

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intramedullary schwannoma in patients with neurofibromatosis because approximately 20% of cases are associated with intramedullary lesions particularly schwannoma.[1,9] We report a case of cervical intramedullary schwannoma occurring at the level between C2 and T1 and associated with syringobulbia.

Case Report

A 24-year-old male was admitted with complaints of neck pain, gradually worsening, weakness in the right upper and lower limbs, numbness in both shoulders, and a decrease in the grasping strength of both hands over a 4-year period. There was no history of urine or fecal incontinence. There was no history of trauma. He was not known to have any medical illness, and his family history was negative. On examination, he was well-built, conscious, and oriented. Glasgow Coma Scale was 15/15. The muscle power of the right upper and lower limbs was grade ⅘ with spasticity and hyperreflexia. Both left upper and lower limbs had normal power, tone, and reflexes. The patient also displayed a hemiplegic gate. At the time of presentation, vibration, light touch, and position sense were all normal. A diagnosis of a cervical spinal cord lesion had been made at another institution. He was referred to our hospital for surgical management. A magnetic resonance imaging (MRI) at the spine showed a heterogeneously enhancing mass in the cervical spinal cord extending from the C2 to T1 levels with associated hemorrhagic changes (►**Fig. 1A, B**). There was an associated syrinx extending from the medulla oblongata to the lower thoracic cord. The appearance of the tumor was suggestive of an ependymoma. There were no specific brain findings. The patient was then prepared for surgery. The patient underwent awake endotracheal intubation and was given general anesthesia. He was placed prone and approached posteriorly through laminoplasty. Under neurophysiological monitoring, the dura was opened, and an exophytic part of the tumor was found at the level of C3, where the tumor was grossly totally resected (piece meal) using the microscope and Omni (dissection and suction). Postoperative MRI showed gross total removal of the tumor (►**Fig. 1C**). In postoperative physical examination, the patient developed severe quadriparesis. Muscle power on the left side was ⅕ and on the right was ⅗. Upon histological examination, the tumor was found to be composed of bland spindle cells with blunt-ended and sometimes wavy nuclei admixed with hyalinized vasculature (►**Fig. 2A–C**). Surrounding reactive spinal cord parenchyma with frequent Rosenthal fibers was also observed. Focal Verocay bodies were evident, and with immunohistochemistry, there was diffuse and strong positivity for S100 (►**Fig. 2D**), which is confirmatory for the diagnosis of schwannoma.

Discussion

Schwannomas account for 30% of all intraspinal tumors, which are the commonest primary tumors of the spine.[8] The age of patients ranges from 9 to 75 years (mean: 40.5 years).[1] Intramedullary schwannoma is more frequently found in males than females (male:female = 3:1).[10] The fourth decade of life is the mean age of onset of the symptoms.[11] Pyramidal symptoms manifest most commonly and are followed by sensory disturbances and sphincter malfunction. This presentation is usually due to the slow compression of the spinal cord, which manifest as weakness, even though these tumors usually arise in the posterior portion of the spinal cord.[7] In some cases, it has been reported that muscular fasciculations were the first clinical manifestation.[10] The time between the beginning of the symptoms until diagnosis was almost always lengthy, with a mean of 28.2 months (range: 6 weeks to 12 years).[7] Intramedullary schwannoma has three

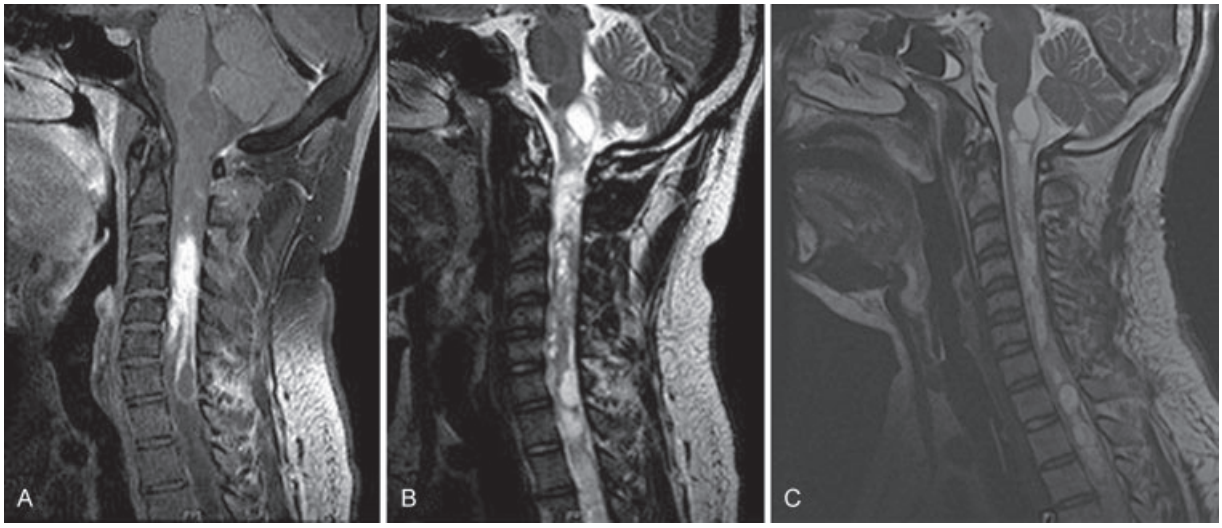

Fig. 1 (**A**) Contrast-enhanced T1-weighted magnetic resonance imaging (MRI) showing a heterogeneously enhanced mass lesion in the cervical spinal cord extending from C2 to T1 with syrinx extending from medulla oblongata to the lower thoracic cord. (**B**) T2-weighted MRI showing a heterogeneous hyperintense mass lesion in the cervical spinal cord extending from C2 to T1 with syrinx extending from medulla oblongata to the lower thoracic cord. (**C**) Postoperative T2-weighted MRI showing gross total removal of the tumor with regression of the syrinx.

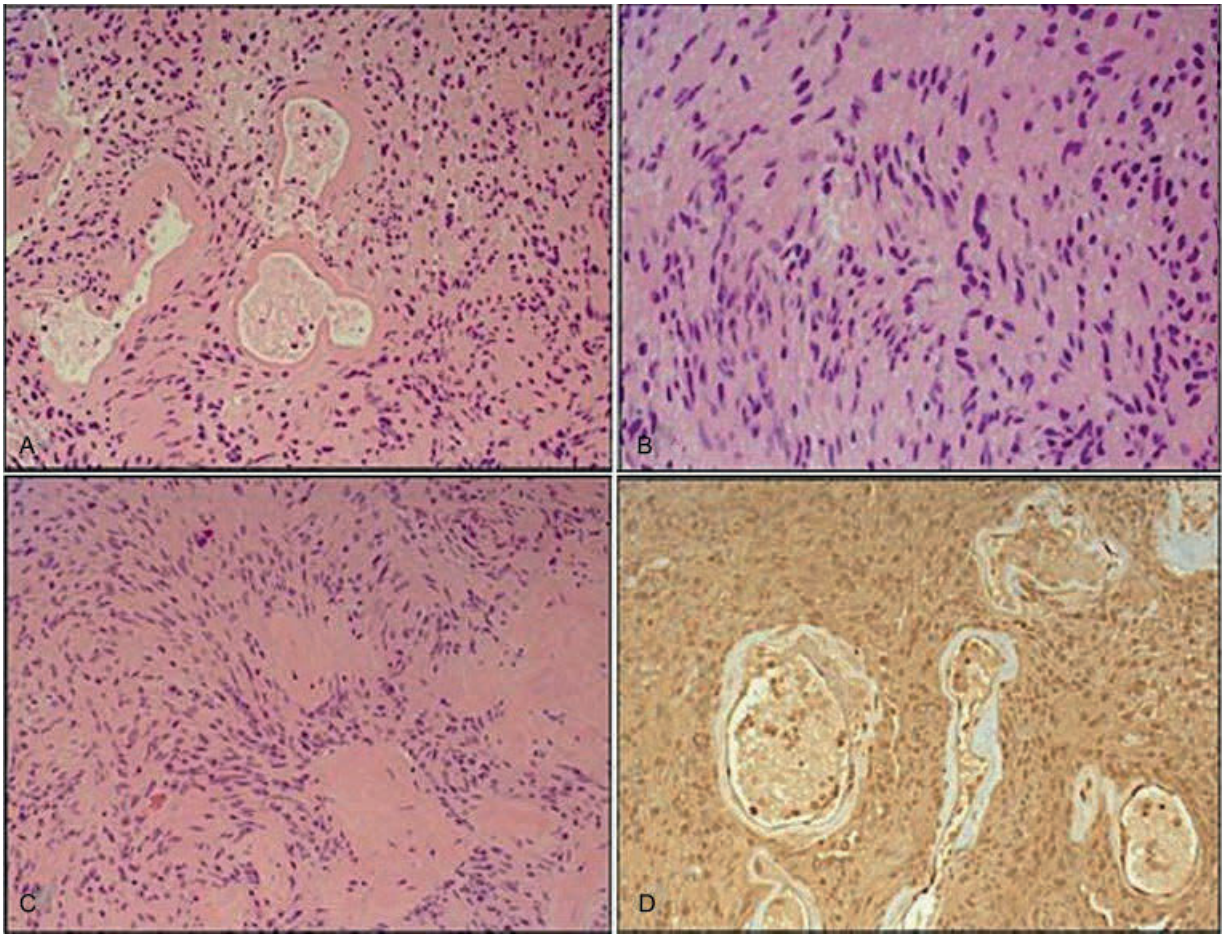

Fig. 2 (**A**) Benign appearing spindle cells and hyalinized vessels. (**B**) Tumor cells depicting blunt-ended and wavy nuclei. (**C**) Subtle palisading around nuclear free zones and more hyalinized vessels. (**D**) Positive immunoreactivity for S100 in nuclear and cytoplasmic patterns.

types based upon the lesion's location in the cross-sectional area of the spinal cord: central, surfacing, and dumbbell.[12,13] The specific type may give a clue as to the origin of these tumors. In the central type, the lesion is found in the parenchyma, which supports the hypothesis that it originates from the perivascular nerve plexus or ectopic Schwann cells.[13] When the lesion is in the peripheral margin of the cord and attached to the pia mater, it named surfacing type, and this type suggests an origin from the Schwann cells of the posterior nerve roots or from conversion of pial cells.[3,12,14,15] In only two cases of the surfacing type, it was suggested that the origin is from the anterior nerve root.[4,13] Intra- and extramedullary schwannomas origin is considered to be from the dorsal root entry zone, which gives it the "dumbbell" shape.[15] According to Kyoshima et al, eight cases of intra- and extramedullary schwannomas have been reported including their case.[13] Nearly 12% of intramedullary schwannoma patients are affected by neurofibroma.[6] Cases of intramedullary schwannoma with neurofibromatosis that have been reported include five with neurofibromatosis type 1 and one with neurofibromatosis type 2.[16,17] Also, Yang et al reported two cases with neurofibromatosis, and Lee et al reported one case but neither mentioned the type.[18,19] Pediatric intramedullary schwannoma cases are rare.

To date, only seven pediatric intramedullary schwannomas have been reported in the literature.[1,11,20–24] After reviewing these cases, we found that the age ranged from 8 to 15 years. All seven cases presented with sensory and motor deficits (7/7; 100%). Pain was present in three (42.8%) and genitourinary functional disturbance also 3 (42.8%). Cervical lesions were found in four (57%), thoracic in two (28.5), and one case involved the C6 to T1 levels. All seven cases underwent complete surgical resection, except for one who had a subtotal resection and an adjuvant radiotherapy.[11] Partial or complete recovery was achieved in the majority of the cases postoperatively. To our knowledge, several authors did a literature review of intramedullary schwannomas since 1931. In 1986, Ross et al reviewed 25 cases, in 1991, Herregodts et al reviewed 36 cases, in 1999, Binatli et al, reviewed 57 cases, in 2002, Darwish et al reviewed 49 cases, and in 2005, Kim et al found that a total of 69 cases had been reported. We have found that 48 cases were reported since 2005, which brings the total number of cases to 118, including our case.[1–3,8,10,11,13,18,19,23–37] The cases reported between 2005 and 2014 are summarized in ►**Tables 1** and **2**. Lee et al studied 10 cases of intramedullary schwannomas that were diagnosed in their hospital from 1995 to 2010 and they found that 7 of them were in the lumber region and 3 in the cervical regions.

Table 1 Summary of intramedullary schwannomas cases from 2005 to 2014

Authors	Age (y) and sex	Location	Initial symptoms	Duration of symptoms	Treatment	Outcome
Kim et al, 2005	72 M	Thoracic T8–T9	Left leg weakness and loss of sensation on the right side	10 mo	GTR	Improved
Kyoshima et al, 2005	54 M	Thoracic T9–10	Numbness in the left foot and rectovesical dysfunction	4 y	GTR	Improved
Shenoy and Raja et al, 2005	29 M	Cervical C4–C7	Interscapular pain that radiated to the upper limbs	3 y	GTR	Improved
Kahilogullari et al, 2005	34 F	Thoracolumbar T12–L2 “conus medullaris”	Pain around her waist and in her legs, and numbness	7 mo	STR	Improved
Ho et al, 2006	45 M	Cervical C5–C6	Incidental	Incidental	GTR	Improved
Ozawa et al, 2006	65 F	Cervical C2–C4	Numbness of the left hand and paresthesia of the left leg	2 y	GTR	Improved
Mukerji et al, 2007	8 M	Cervical C5–C7	Weakness in all limbs	Sudden	GTR	Improved
Hida et al, 2008	41 M	Cervical C1–C2	Dysesthesia of all four limbs	6 mo	Partial resection then GTR	Improved
	30 M	Cervical C5–C7	Decrease in the grasping strength of the left hand	?	Partial resection then GTR	Improved
Hayashi et al, 2009	78 F	Thoracolumbar T11–L1	Pain and numbness in both legs	20 y	GTR	Improved
Ohtonari et al, 2009	29 M	Thoracolumbar T12–L1 “conus medullaris”	Bladder dysfunction, sexual impotence, and paresthesia in the buttocks	8 mo	STR	Improved
Kim et al, 2009	11 F	Thoracic T5–T6	Weakness of the lower limbs, back pain, and urge incontinence	9 mo	STR + RT	Improved
Nicácio et al, 2009	40 M	Cervical C4–C6	Spastic tetraparesis and sphincterian disturbances	2 y	STR	Improved
Lyle et al, 2010	Neonate	Thoracic T2 to the thecal sac	?	?	?	?
Ryu et al, 2011	68 M	Thoracic T5–T6	Walking disturbance and decreased sensation	17 mo	GTR	Improved
Vij et al, 2011	25 M	Thoracic T10–T11	Low back pain radiating to right lower limb, and bilateral weakness and numbness	3 y	GTR	Improved
Li et al, 2013	42 M	Thoracic T3–T4	Zonesthesia in the right side of the chest, and weakness and numbness of the bilateral lower limbs	1.5 y	GTR	Improved
Eljebbouri et al, 2013	10 M	Thoracic T7–T9	Weakness of the lower limbs associated with bladder and bowel incontinence	Sudden	GTR	Improved
Lee et al, 2013	19 F	Thoracic T6–T8	Gait disturbance with motor deficit (nine cases) associated with sensory disturbance (six cases,) and difficulty in urination and toileting (two cases); one patient presented with weakness in the left upper limb	The mean duration was 39.3 ± 36.0 (mo: 3–120)	GTR	Improved
	37 F	Thoracic T9–T10			GTR	Improved
	39 F	Cervical C4–C7			GTR	Improved
	41 F	Cervical C5–C6			GTR	Improved
	42 M	Thoracic T7–T8			STR	Improved
	44 M	Thoracic T8–T9			GTR	Improved
	46 F	Thoracic T1–T2			STR	Improved
	49 F	Cervical C5–C7			STR	Improved
	60 M	Thoracic T7–T10			GTR	Improved
	78 M	Thoracic T10–T11			GTR	Improved

Table 1 (*Continued*)

Authors	Age (y) and sex	Location	Initial symptoms	Duration of symptoms	Treatment	Outcome
Karatay et al, 2014	30 F	Thoracolumbar T12–L1 "conus medullaris"	Back pain, walking disturbance, and numbness in both legs	2 mo	STR	Improved
Yang et al, 2014	17 M	Thoracic T6–T8	Right lower limb pain and numbness	1 y	STR	Improved
	31 M	Cervical C3–C4	Neck pain, bilateral upper limb numbness	1 y	GTR	Improved
	34 M	Thoracic T12	Back pain and left lower limb weakness	4 y	GTR	Improved
	35 M	Cervical C6	Neck pain and left lower limb weakness	3 y	GTR	Improved
	38 M	Thoracic T11	Bilateral lower limb pain and numbness, and difficulty in urination	18 mo	GTR	Improved
	39 M	Cervical C3–C5	Neck pain and bilateral lower limb weakness	1 y	GTR	Improved
	40 M	Cervical C3	Right upper limb pain and numbness	2 mo	GTR	Improved
	41 F	Cervical C4–C6	Neck pain and bilateral lower limb weakness	6 mo	GTR	Improved
	42 M	Thoracic T10–	Bilateral lower limb numbness	2 y	GTR	Improved
	44 M	T12	Right lower limb numbness and weakness	1 y	GTR	Improved
	44 F	Thoracic T3	Thoracic and midback pain, and bilateral lower limb weakness	4 y	GTR	Improved
	46 M	Cervical C5–C7	Back pain, and bilateral lower limb numbness and weakness	1 y	STR	Improved
	48 M	Thoracic T3–T5	Bilateral lower limb weakness, and numbness and difficulty in urination	12 y	GTR	No change
	50 F	Thoracic T9–T10	Neck and back pain, and right lower limb weakness	2 y	GTR	Improved
	52 M	Cervicothoracic C5–T1	Bilateral upper limb pain, bilateral lower limb weakness, and difficulty in urination	10 y	GTR	No change
	56 F	Cervicothoracic C6–T4	Neck pain, and bilateral lower limb numbness and weakness	3 y	STR	Improved
	57 M	Cervical C5–C6	Neck and back pain, and right upper limb pain	6 mo	GTR	Improved
	59 M	Cervical C4–C6	Right upper limb numbness and left lower limb pain	3 y	STR	Improved
	60 F	Cervical C1–C2	Bilateral lower limb pain and weakness	3 y	GTR	Improved
	61 M	Thoracic T2–T3 Cervical C6–C7	Left upper limb pain and numbness	2 y	GTR	Improved

Abbreviations: GTR, gross total resection; STR, subtotal resection.
Note: ? indicates that the value is not mentioned in the original paper.

Table 2 Summary of MRI and histopathological findings in intramedullary schwannoma cases from 2005 to 2014

Authors	T1-weighted image	T2-weighted image	Gadolinium enhancement	Cysts, peritumoral edema, or syringomyelia	Preoperative diagnosis	Histopathological diagnosis
Kim et al, 2005	Isointense	Hyperintense	Homogeneous; well demarcated	–, +, –	?	Schwannoma, Antoni type A and B, + S100 protein
Kyoshima et al, 2005	Hypo to isointense	Hypointense	Homogeneous; well demarcated	–, –, –	Intradural extramedullary tumor	Schwannoma
Shenoy and Raja, 2005	Hypo to isointense	Hyperintense	Ringlike peripheral enhancement	–, –, +	?	Schwannoma, Antoni type A and B, + S100 protein
Kahilogullari et al, 2005	?	?	Heterogeneous	–, –, –	?	Schwannoma, Antoni type A, + S100 protein
Ho et al, 2006	Isointense	Hyperintense	Homogeneous; well demarcated	–, –, –	Extramedullary tumor	Schwannoma, Antoni type A and B
Ozawa et al, 2006	Hypointense	Hyperintense	Homogeneous; well demarcated	–, +, –	Astrocytoma	Schwannoma, Antoni type A and B
Mukerji et al, 2007	?	?	?	–, +, –	Astrocytoma	?
Hida et al, 2008	Hypointense [2]?	Iso- to hyperintense ?	Heterogeneous; well demarcated Homogeneous; well demarcated	–, +, – –, –, –	Schwannoma ?	? ?
Hayashi et al, 2009	Hypointense	Isointense	Heterogeneous	–, –, – “calcification”	?	Ancient schwannoma, Antoni type A, + S100 protein + S100 protein and GFAP
Ohtonari et al, 2009	Isointense	?	Homogeneous; well demarcated	+, –, –	?	Schwannoma, Antoni type A, + S100 protein
Kim et al, 2009	Hypointense	Hyperintense	?	–, –, +	Ependymoma	Schwannoma, Antoni type A and B, + S100 protein
Nicácio et al, 2009	Hypointense	Hyperintense	Heterogeneous; well demarcated	–, –, +	?	Schwannoma, Antoni type A
Lyle et al, 2010	?	?	?	?	?	Congenital schwannoma, Antoni type A and B.

Table 2 (*Continued*)

Authors	T1-weighted image	T2-weighted image	Gadolinium enhancement	Cysts, peritumoral edema, or syringomyelia	Preoperative diagnosis	Histopathological diagnosis
Ryu et al, 2011	Hypointense	Hyperintense	Heterogeneous; well demarcated	–, +, –	?	Schwannoma, Antoni type A, + S100 protein and silver stain
Vij et al, 2011	Hypointense	Isointense	Homogeneous; well demarcated	–, –, –	Ependymoma	Schwannoma, Antoni type A, with cysticercus parasite cyst
Li et al, 2013	Isointense	Hypointense	Heterogeneous; well demarcated	–, –, –	Astrocytoma or ependymoma	Schwannoma?
Eljebbouri et al, 2013	?	Hyperintense	Heterogeneous	+ –, –, –	Astrocytoma	Schwannoma, Antoni type A and B.
Lee et al, 2013	Hyperintense in three cases; hypointense in four cases; and isointense in three cases	Hyperintense in five cases and hypointense in five cases	Six cases with a homogenous, well-enhanced mass with sharp demarcation, two cases with a heterogeneous enhanced mass, and two cases with peripheral enhancement	Peritumoral edema (seven cases,) and tumor cysts (eight cases)	Ependymomas in four cases, astrocytomas in three cases, and hemangioblastoma, lymphoma, and metastasis in one case each	All Schwannomas, Antoni type A and B, + S100 protein
Karatay et al, 2014	Hypointense	Hyperintense	Homogeneous; well demarcated	–, –, +	?	Schwannoma, Antoni type A and B, + S100 protein
Yang et al, 2014	Isointense	Isointense	Heterogeneous; well demarcated	–, +, –	Astrocytoma	All schwannomas, Antoni type A and B
	Isointense	Isointense	Heterogeneous; well demarcated	–, –, –	Ependymoma	
	Isointense	Iso- to hyperintense	Heterogeneous; well demarcated	+, –, +	Schwannoma	
	Hyperintense	Hyperintense	Homogeneous; well demarcated	+, –, +	Ependymoma	
	Isointense	Isointense	Homogeneous; well demarcated	–, –, +	Ependymoma	
	Isointense	Isointense	Homogeneous; well demarcated	–, –, –	Ependymoma	
	Isointense	Hyperintense	Heterogeneous; well demarcated	–, –, +	Ependymoma	
	Hyperintense	Hyperintense	Homogeneous; well demarcated	–, –, +	Ependymoma	

(*Continued*)

Table 2 (*Continued*)

Authors	T1-weighted image	T2-weighted image	Gadolinium enhancement	Cysts, peritumoral edema, or syringomyelia	Preoperative diagnosis	Histopathological diagnosis
	Iso- to hypointense	Iso- to hyperintense	Heterogeneous; well demarcated	–, –, +	Ependymoma	
	Isointense	Isointense	Homogeneous; well demarcated	+, –, –	Schwannoma	
	Isointense	Iso- to hyperintense	Heterogeneous; well demarcated	–, +, –	Ependymoma	
	Iso- to hypointense	Hyperintense	Heterogeneous	+, +, –	Ependymoma	
	Isointense	Hyperintense	Homogeneous; well demarcated	–, –, +	Astrocytoma	
	Hypointense	Iso- to hyperintense	Homogeneous; well demarcated	+, +, –	Ependymoma	
	Iso- to hypointense	Hyperintense	Heterogeneous; well demarcated	+, +, –	Ependymoma	
	Hypointense	Hyperintense	Heterogeneous	+, +, –	Ependymoma	
	Iso- to hypointense	Hyperintense	Heterogeneous; well demarcated	+, +, –	Astrocytoma	
	Isointense	Isointense	Homogeneous; well demarcated	–, –, +	Ependymoma	
	Isointense	Isointense	Heterogeneous	–, –, +	Ependymoma	
	Hypointense	Iso- to hyperintense	Circular; well demarcated	+, –, –	Ependymoma	

Abbreviations: GFAP, glial fibrillary acidic protein; MRI, magnetic resonance imaging.
Note: ? indicates that the value is not mentioned in the original paper.
+ indicates that the value exists in the images
− indicates that the value does not exist in the images

The primary radiological images suggested the diagnosis of ependymoma in four cases, astrocytoma in three cases, and hemangioblastoma, lymphoma, and metastasis in the other three cases. The T1-weighted MRI images revealed the lesion with hyperintensity in three cases, hypointensity in four cases, and an isointensity in three cases. The T2-weighted MRI images revealed the lesion with hyperintensity in five cases and with hypointensity in five cases. The T1-weighted MRI with contrast resulted in homogenous, well-enhanced tumors which were well-demarcated in six cases, a heterogeneous enhanced tumor in two cases, and peripheral enhancement in two cases. Lesions were accompanied by perilesional edema in seven cases and cysts in eight cases. Of 365 patients with a diagnosis of spinal cord schwannomas included in the study, only 10 (2.7%) had intramedullary schwannomas. The first symptom was gait abnormality with motor deficit, followed by sensory deficit and urinary symptoms. Histologically, all tumors showed Antoni A and B areas, and mitotic figures were hardly found. In the immunohistochemical tests, all tumor cells were positive for S100 protein but negative for glial fibrillary acidic protein. This study supports the theory of the nerve root origin of intramedullary schwannoma because half of the tumors had an attachment to the dorsal rootlets.[18] Wu et al reviewed the data of seven patients with diagnosed intramedullary schwannoma treated at their hospital from 2003 to 2010 and compared them with patients with ependymoma and astrocytoma from the same period. They found that there was a significant difference in the incidence of somatic and root pain as the first symptoms between intramedullary schwannoma and ependymoma ($p = 0.005$) and between intramedullary schwannoma and astrocytoma ($p = 0.019$), but not between ependymoma and astrocytoma ($p = 0.175$). MRI analysis showed isointense or low-intense tumors on T1-weighted images and high intense or mixed on T2-weighted images. There were four cases associated with tumor cysts and three cases with syringomyelia above the tumor. Contrast enhancement was homogenous in four cases, heterogeneous in two, and circular in one. All lesions were well-demarcated. But they did not find a significant difference on MRI between intramedullary schwannoma and the gliomas. The mean postoperative follow-up period was 56 months, with no neurologic deterioration or recurrence.[38] Yang et al analyzed 20 cases of intramedullary schwannoma that were diagnosed in their hospital from 2000 to 2013, including 7 cases that were reported by Wu et al in 2011, but with a longer follow-up period (►**Table 1**). They also encountered 1,320 patients with intramedullary lesions (ependymomas, astrocytomas, and hemangioblastomas) and 1,723 patients with intraspinal schwannomas during the same period. Intramedullary schwannomas accounted for 1.49% of all intramedullary lesions (total = 1,320) and 1.16% of all intraspinal schwannomas (total = 1,723).[19] Intramedullary schwannomas are commonly found on MRI T1-weighted images as iso- or hypointense lesions, and a little hyperintense on T2-weighted images.[7] The lesions are most commonly well marginated, with some edema.[7] With contrast enhancement, almost all cases show homogeneous or nodular enhanced tumors.[7] Ependymomas, astrocytomas, hemangioblastomas, and metastasis are all intramedullary lesions that are contrast-enhanced. Therefore, they are included in the differential diagnosis.[7,39] Commonly, they show vague lesion margins and are accompanied by edema and cysts.[40–42] On the contrary, intramedullary schwannomas usually reveal a greatly enhanced lesion with well-demarcated margins.[4,7] Most ependymomas are centrally located because they grow from the central canal, with a symmetrical expansion of the spinal cord.[41] Astrocytomas originate from the astrocyte, which can be found anywhere in the spinal cord, and therefore their growth patterns tend to be different.[41] Hemangioblastomas have a prominent enhancement and may have a flow void on MRI.[42] A review of 20 cases by Ozawa et al concluded that 10 cases showed hypointensity to isointensity signals on T1-weighted MRI and slightly hyperintensity signals in 5 cases. T2-weighted MRI revealed seven cases with hyperintensity signals and five cases with hypointensity signals. Contrast-enhanced T1-weighted MRI resulted in well-defined lesion margins in 15 cases. Homogenous enhancement was documented in 65% of cases. Perilesional edema was mentioned in 10 cases.[30] Balériaux concluded that there is no correlation between the MRI findings and the Antoni classification.[39] The Antoni A type is characterized by the presence of condensed wavy bundles of cells with rod or ovoid nuclei, and palisading arrangement of the cells can be seen.[1] The Antoni B type has large and loosely organized hypodense cells with polymorphism.[1] The new WHO classification of tumors has three types of schwannomas: plexiform, cellular, and melanotic.[11] Complete excision of intramedullary schwannoma is usually achievable, and postoperative outcomes have been good.[28] However, there were reports of death in five cases and recurrence in two.[1,43] Ohtonari et al analyzed the resectability of 39 cases of intramedullary schwannomas of the spinal cord at each level and found 5 subtotal excision and 17 total excision cases at the cervical level, 4 and 6 at the thoracic level, 2 and 5 at the lumber level. All cases with total excision showed no neurologic deterioration. Two cases worsened after subtotal excision. One was at the cervical level and the other was at the lumber level.[2] Recurrence after subtotal excision of intramedullary schwannomas was reported in two cases after 5 and 3 years follow-up. Even though the final histological diagnosis of the two cases were schwannomas, subtotal removal was done because frozen sections during the operation suggested astrocytoma.[43]

Conclusion

Intramedullary schwannomas are benign and slowly progressive lesions. The definitive diagnosis can be made by pathology. It is difficult to differentiate intramedullary schwannoma from other intramedullary lesions by MRI only. When gross total resection is usually needed but cannot be done, subtotal resection of the tumor is recommended. A good clinical outcome after surgery can be anticipated.

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