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. 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. 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. 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. Intramedullary schwannoma accounts for almost 0.3 to 1.5% of all primary intraspinal lesions. It commonly involves the cervical region (61%) and, to a lesser extent, the thoracic (29%), and lumbosacral (10%) regions. Infrequently, they are associated with syringomyelia. To make a diagnosis of intramedullary schwannoma and to differentiate it from other neoplasms by imaging only is almost impossible. We can suspect an
intramedullary schwannoma in patients with neurofibromatosis because approximately 20% of cases are associated with intramedullary lesions particularly schwannoma.\textsuperscript{1,6} 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 with spasticity and hyperreflexia. 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 C2 to T1 levels with associated hemorrhagic changes (\textsuperscript{▶}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 (\textsuperscript{▶}Fig. 1C). In postoperative physical examination, the patient developed severe quadriplegia. Muscle power on the left side was ⅙ 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 (\textsuperscript{▶}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 (\textsuperscript{▶}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.\textsuperscript{8} The age of patients ranges from 9 to 75 years (mean: 40.5 years).\textsuperscript{1} Intramedullary schwannoma is more frequently found in males than females (male:female = 3:1).\textsuperscript{10} The fourth decade of life is the mean age of onset of the symptoms.\textsuperscript{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.\textsuperscript{7} In some cases, it has been reported that muscular fasciculations were the first clinical manifestation.\textsuperscript{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).\textsuperscript{7} Intramedullary schwannoma has three
types based upon the lesion’s location in the cross-sectional area of the spinal cord: central, surfacing, and dumbbell.\textsuperscript{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.\textsuperscript{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.\textsuperscript{3,12,14,15} In only two cases of thesurfacing type, it was suggested that the origin is from the anterior nerve root.\textsuperscript{4,11} Intra- and extramedullary schwannomas origin is considered to be from the dorsal root entry zone, which gives it the “dumbbell” shape.\textsuperscript{15} According to Kyoshima et al, eight cases of intra- and extramedullary schwannomas have been reported including their case.\textsuperscript{15} Nearly 12% of intramedullary schwannoma patients are affected by neurofibroma.\textsuperscript{6} Cases of intramedullary schwannoma with neurofibromatosis that have been reported include five with neurofibromatosis type 1 and one with neurofibromatosis type 2.\textsuperscript{16,17} Also, Yang et al reported two cases with neurofibromatosis, and Lee et al reported one case but neither mentioned the type.\textsuperscript{18,19} Pediatric intramedullary schwannoma cases are rare.

To date, only seven pediatric intramedullary schwannomas have been reported in the literature.\textsuperscript{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.\textsuperscript{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, Binati 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.\textsuperscript{1–3,8,10,11,13,18,23–27} The cases reported between 2005 and 2014 are summarized in Table 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

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

The mean duration was $39.3 \pm 36.0$ (mo: 3–120)
<table>
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<tr>
<th>Authors</th>
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<th>Duration of symptoms</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Karatay et al, 2014</td>
<td>30 F</td>
<td>Thoracolumbar T12–L1 “conus medullaris”</td>
<td>Back pain, walking disturbance, and numbness in both legs</td>
<td>2 mo</td>
<td>STR</td>
<td>Improved</td>
</tr>
<tr>
<td>Yang et al, 2014</td>
<td>17 M</td>
<td>Thoracic T6–T8</td>
<td>Right lower limb pain and numbness</td>
<td>1 y</td>
<td>STR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>31 M</td>
<td>Cervical C3–C4</td>
<td>Neck pain, bilateral upper limb numbness</td>
<td>1 y</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>34 M</td>
<td>Thoracic T12</td>
<td>Back pain and left lower limb weakness</td>
<td>4 y</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>35 M</td>
<td>Cervical C6</td>
<td>Neck pain and left lower limb weakness</td>
<td>3 y</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>38 M</td>
<td>Thoracic T11</td>
<td>Bilateral lower limb pain and numbness, and difficulty in urination</td>
<td>18 mo</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>39 M</td>
<td>Cervical C3–C5</td>
<td>Neck pain and bilateral lower limb weakness</td>
<td>1 y</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>40 M</td>
<td>Cervical C3</td>
<td>Right upper limb pain and numbness</td>
<td>2 mo</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>41 F</td>
<td>Cervical C4–C6</td>
<td>Neck pain and bilateral lower limb weakness</td>
<td>6 mo</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>42 M</td>
<td>Thoracic T10–</td>
<td>Bilateral lower limb numbness</td>
<td>2 y</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>44 M</td>
<td>T12</td>
<td>Right lower limb numbness and weakness</td>
<td>1 y</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>44 F</td>
<td>Thoracic T3</td>
<td>Thoracic and midback pain, and bilateral lower limb weakness</td>
<td>4 y</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>46 M</td>
<td>Cervical C5–C7</td>
<td>Back pain, and bilateral lower limb numbness and weakness</td>
<td>1 y</td>
<td>STR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>48 M</td>
<td>Thoracic T3–T5</td>
<td>Bilateral lower limb weakness, and numbness and difficulty in urination</td>
<td>12 y</td>
<td>GTR</td>
<td>No change</td>
</tr>
<tr>
<td></td>
<td>50 F</td>
<td>Thoracic T9–T10</td>
<td>Neck and back pain, and right lower limb weakness</td>
<td>2 y</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>52 M</td>
<td>Cervicothoracic C5–T1</td>
<td>Bilateral upper limb pain, bilateral lower limb weakness, and difficulty in urination</td>
<td>10 y</td>
<td>GTR</td>
<td>No change</td>
</tr>
<tr>
<td></td>
<td>56 F</td>
<td>Cervicothoracic C6–T4</td>
<td>Neck pain, and bilateral lower limb numbness and weakness</td>
<td>3 y</td>
<td>STR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>57 M</td>
<td>Cervical C5–C6</td>
<td>Neck and back pain, and right upper limb pain</td>
<td>6 mo</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>59 M</td>
<td>Cervical C4–C6</td>
<td>Right upper limb numbness and left lower limb pain</td>
<td>3 y</td>
<td>STR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>60 F</td>
<td>Cervical C1–C2</td>
<td>Bilateral lower limb pain and weakness</td>
<td>3 y</td>
<td>GTR</td>
<td>Improved</td>
</tr>
<tr>
<td></td>
<td>61 M</td>
<td>Thoracic T2–T3 Cervical C6–C7</td>
<td>Left upper limb pain and numbness</td>
<td>2 y</td>
<td>GTR</td>
<td>Improved</td>
</tr>
</tbody>
</table>

Note: ? indicates that the value is not mentioned in the original paper.

Abbreviations: GTR, gross total resection; STR, subtotal resection.
<table>
<thead>
<tr>
<th>Authors</th>
<th>T1-weighted image</th>
<th>T2-weighted image</th>
<th>Gadolinium enhancement</th>
<th>Cysts, peritumoral edema, or syringomyelia</th>
<th>Preoperative diagnosis</th>
<th>Histopathological diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kim et al, 2005</td>
<td>Isointense</td>
<td>Hyperintense</td>
<td>Homogeneous; well demarcated</td>
<td>–, +, –</td>
<td>?</td>
<td>Schwannoma, Antoni type A and B, + S100 protein</td>
</tr>
<tr>
<td>Kyoshima et al, 2005</td>
<td>Hypo to isointense</td>
<td>Hypointense</td>
<td>Homogeneous; well demarcated</td>
<td>–, –, –</td>
<td>Intradural extramedullary tumor</td>
<td>Schwannoma</td>
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<tr>
<td>Shenoy and Raja, 2005</td>
<td>Hypo to isointense</td>
<td>Hyperintense</td>
<td>Ringlike peripheral enhancement</td>
<td>–, –, +</td>
<td>?</td>
<td>Schwannoma, Antoni type A and B, + S100 protein</td>
</tr>
<tr>
<td>Ho et al, 2006</td>
<td>Isointense</td>
<td>Hyperintense</td>
<td>Homogeneous; well demarcated</td>
<td>–, –, –</td>
<td>Extramedullary tumor</td>
<td>Schwannoma, Antoni type A and B</td>
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<tr>
<td>Ozawa et al, 2006</td>
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<td>Hyperintense</td>
<td>Homogeneous; well demarcated</td>
<td>–, +, –</td>
<td>Astrocytoma</td>
<td>Schwannoma, Antoni type A and B</td>
</tr>
<tr>
<td>Hida et al, 2008</td>
<td>Hypointense</td>
<td>Iso- to hyperintense</td>
<td>Heterogeneous; well demarcated</td>
<td>–, +, –</td>
<td>Schwannoma</td>
<td>?</td>
</tr>
<tr>
<td>Hayashi et al, 2009</td>
<td>Hypointense</td>
<td>Isointense</td>
<td>Heterogeneous</td>
<td>–, –, –</td>
<td>?</td>
<td>Ancient schwannoma, Antoni type A, + S100 protein + S100 protein and GFAP</td>
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<tr>
<td>Ohtonari et al, 2009</td>
<td>Isointense</td>
<td>?</td>
<td>Homogeneous; well demarcated</td>
<td>+, –, –</td>
<td>?</td>
<td>Schwannoma, Antoni type A, + S100 protein</td>
</tr>
<tr>
<td>Kim et al, 2009</td>
<td>Hypointense</td>
<td>Hyperintense</td>
<td>?</td>
<td>–, –, +</td>
<td>Ependymoma</td>
<td>Schwannoma, Antoni type A and B, + S100 protein</td>
</tr>
<tr>
<td>Nicácio et al, 2009</td>
<td>Hypointense</td>
<td>Hyperintense</td>
<td>Heterogeneous; well demarcated</td>
<td>–, –, +</td>
<td>?</td>
<td>Schwannoma, Antoni type A</td>
</tr>
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</table>
Table 2 (Continued)

<table>
<thead>
<tr>
<th>Authors</th>
<th>T1-weighted image</th>
<th>T2-weighted image</th>
<th>Gadolinium enhancement</th>
<th>Cysts, peritumoral edema, or syringomyelia</th>
<th>Preoperative diagnosis</th>
<th>Histopathological diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ryu et al, 2011</td>
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<td>Eljebbouri et al, 2013</td>
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<td>Lee et al, 2013</td>
<td>Hyperintense in three cases; hypointense in four cases; and iso-intense in three cases</td>
<td>Hyperintense in five cases and hypointense in five cases</td>
<td>Six cases with a homogenous, well-enhanced mass with sharp demarcation, two cases with a heterogeneous enhanced mass, and two cases with peripheral enhancement</td>
<td>Peritumoral edema (seven cases,) and tumor cysts (eight cases)</td>
<td>Ependymomas in four cases, astrocytomas in three cases, and hemangioblastoma, lymphoma, and metastasis in one case each</td>
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(Continued)
## Table 2 (Continued)

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<th>T2-weighted image</th>
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<th>Histopathological diagnosis</th>
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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
Intramedullary schwannomas are commonly reviewed 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. Wu et al reviewed the data of seven patients with intramedullary schwannoma and ependymoma from 2000 to 2013, including 7 cases that were reported by Wu et al in 2011, but with a longer follow-up period. They found that there was a significant difference between the MRI findings and the Antoni classification. The new WHO classification of tumors has three types of schwannomas: plexiform, cellular, and melanotic. Complete excision of intramedullary schwannoma is usually achievable, and postoperative outcomes have been good. However, there were reports of death in five cases and recurrence in two. Onthonari 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 lumbar 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 lumbar level. 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.

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.
References


