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# Intradural cystic schwannomas of the spine: A case-based systematic review of an unusual tumor

Alexis Paul Romain Terrapon [a,b], Martin N. Stienen [a], Anand Veeravagu [c], Michael Fehlings [d], Oliver Bozinov [a], Nader Hejrati [a,*]

[a] Department of Neurosurgery & Spine Center of Eastern Switzerland, Kantonsspital St.Gallen & Medical School of St.Gallen, St.Gallen, Switzerland
[b] Department of Neurosurgery, Bern University Hospital, Rosenbühlgasse 25, 3010 Bern, Switzerland
[c] Neurosurgery AI Lab & Department of Neurosurgery, Stanford University School of Medicine, Stanford, CA, USA
[d] Division of Neurosurgery, Department of Surgery, University of Toronto, Toronto, ON, Canada

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ABSTRACT

*Introduction:* Cystic schwannomas have only been reported in a few case reports/series. As a result, they may be misdiagnosed and a standardized management approach remains challenging to establish.
*Research question:* The aim of this study was to compile all reported cases of cystic schwannomas and analyze the perioperative course based on a systematic review of the literature with an additional two cases from the authors' experience.
*Material and methods:* We conducted a search of MEDLINE and CENTRAL databases for spinal intradural extramedullary cystic schwannomas, in accordance to the PRISMA statement. All title/abstracts were screened, and a full-text review of the remaining articles was conducted. The results were compiled in tables and summarized using means and standard deviation (SD), median and interquartile range, and percentage and 95% confidence intervals.
*Results:* We identified 263 articles, of which 35, which reported 54 cases, were included. Including our case-reports (n = 56), patients had a mean age of 47.7 years (SD ± 13.0 years) at presentation, 57% were males, and most lesions were lumbar (43%). The most common symptoms were pain (82%) and muscle weakness (68%) with 84% of patients showing neurological findings. 70% of patients showed a complete relief of symptoms after surgery and 96% reported improvement. Only four complications were reported.
*Discussion and conclusion:* Schwannomas should be considered in the differential diagnosis of intradural extramedullary cystic lesions. Patients typically present with subacute to chronic pain and/or neurologic changes. Surgical resection is the primary therapeutic modality and usually has a good to excellent outcome.

## 1. Introduction

Schwannomas (also called neurinomas, neuri-/neurolemmomas) are benign nerve sheath tumors consisting of clonal schwann cells (SC), which usually affect small peripheral nerves, but may also be located in the central nervous system, commonly arising from sensory nerve roots. Although usually sporadic, schwannomas may be associated with syndromes like neurofibromatosis type 2, schwannomatosis or Carney's complex and may be induced by external factors such as radiation (Hilton and Hanemann, 2014).

After meningiomas, spinal schwannomas are the second most common intradural extramedullary tumors with an estimated incidence of 0.24 per 100′000 persons in the United States. The incidence has been reported to be the highest among male Caucasians aged 65–75 years (Tish et al., 2019). Although schwannomas may exhibit cystic components, they rarely present as complete or predominantly cystic lesion and are often initially misdiagnosed as other possibly cystic lesions (arachnoid cyst, spinal arachnoid web, focal arachnoiditis or arachnopathy, ependymoma. meningioma) (Voglis et al., 2022). To date, a comprehensive systematic review of the literature has not been undertaken.

To increase the awareness for the existence and to better delineate

* Corresponding author. Department of Neurosurgery & Spine Center of Eastern Switzerland, Kantonsspital St.Gallen, Medical School of St.Gallen, Rorschacher Strasse 95, 9007, St.Gallen, Switzerland.
*E-mail address:* Nader.Hejrati@kssg.ch (N. Hejrati).

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the specific features of spinal intradural, extramedullary schwannoma with predominant or very large cystic components, we chose a systematic approach in reviewing the literature on all documented cases. By doing so, our goal was to provide guidance on diagnosis and therapeutic strategies for this rare entity. In addition, we add two cases which were treated in the author's centers.

## 2. Methods

This study was conducted and is reported in accordance to the Preferred Reporting Items for Systematic reviews and Meta-Analyses (PRISMA) statement (Page et al., 2021). The aim of the systematic review was to summarize all clinical reports of patients with intradural, largely/predominantly cystic, extramedullary schwannoma. Accordingly, we did not consider reports that did not provide clinical patient data, reports of tumors located mainly extradural or intramedullary, or reports of lesions that were predominantly solid without large cystic portions.

### 2.1. Literature search and terms

To retrieve the relevant literature, we conducted a structured search of the MEDLINE database using Pubmed on June 2, 2023 with the terms ((intradural*) OR (spinal*)) AND ((schwannoma*) OR (neurinoma*) OR (neurilem*) OR (neurolem*)) AND (cyst*) NOT (animals[mesh] NOT humans[mesh]) and retrieved 250 records. The same day, the CENTRAL database was searched with the terms (cyst*) AND ((schwannoma*) OR (schwannoma*) OR (neurinoma*) OR (neurilem*) OR (neurolem*)) and 13 records were found. One duplicate was excluded. We screened all titles/abstracts and 201 articles were excluded because they were not considered relevant (e.g.: irrelevant to the topic, not reporting clinical data, mostly solid or non-cystic lesions, mainly extradural lesions, intramedullary tumors) or not in English language. A full-text review of the available remaining records was then conducted, and references of key articles were screened for additional studies, but none was found. The detailed review process is illustrated in Fig. 1.

### 2.2. Data extraction and variables

The relevant data (name of first author, year of publication, type of study, number of reported cases, mean age, gender, spinal levels concerned, radiographic appearance of the tumor, patients' history, pathologic neurological status at admission, therapy, symptoms and neurological status at last follow-up, description of adverse events) was extracted from the full-text and/or abstract of all included manuscripts (n = 35) and summarized in Table 1. Items that were not provided by the

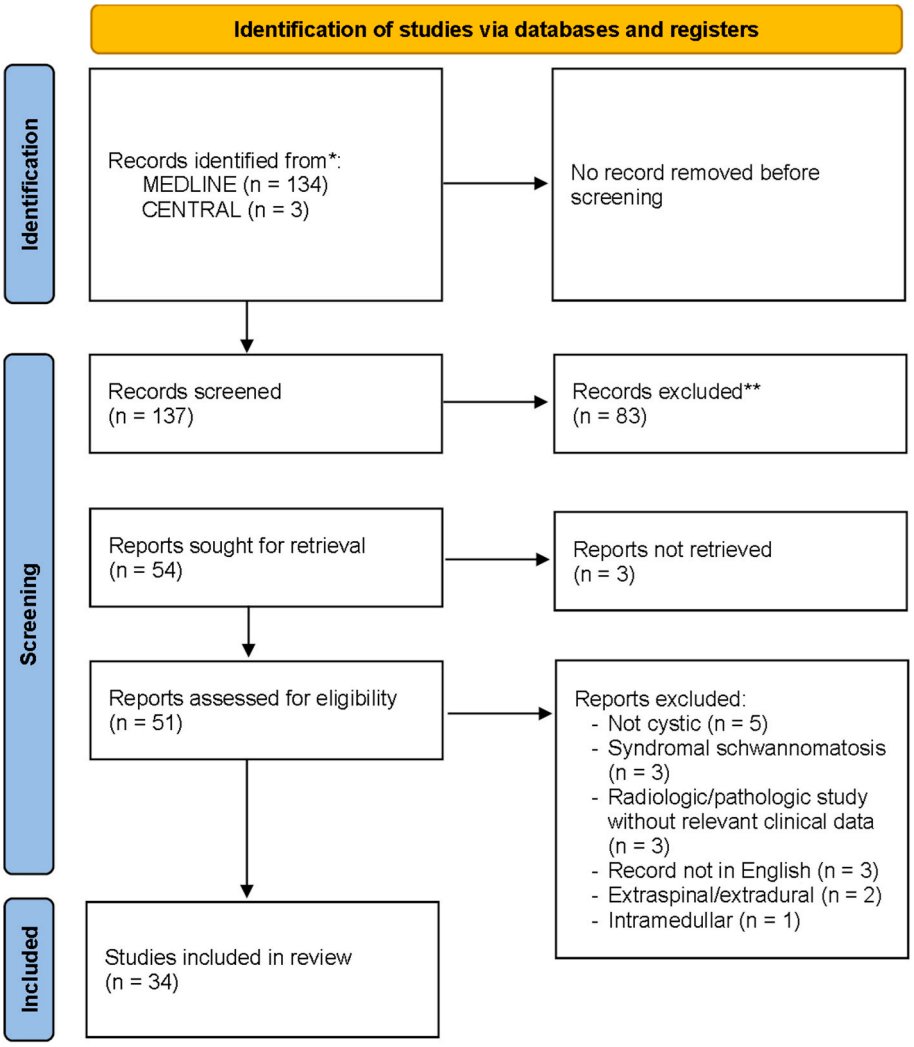


**Fig. 1.** Flow diagram depicting review process according to PRISMA (Page et al., 2021)
Additionally to the structured literature search, references of key articles were screened for additional studies, but none was found. It should be noted that one article was not accessible as full-text in English but still was included in the study because the relevant information could be obtained from the abstract (Sato et al., 1993).

**Table 1**
Summary of the available evidence.

| Authors, year, study type | Number of patients, age, gender | Spinal levels | Radiographic features | History (duration) | Clinical examination | Therapy | Outcome (date of last follow-up) | Adverse Events | Comments |
|---|---|---|---|---|---|---|---|---|---|
| Terrapon et al., 2024, case report and systematic review | 2,<br>1: 44y, F<br>2: 35y, M | T7-11 | Septated cyst with hemorrhage and slight vertebral scalloping | Back pain (years) leg weakness (2m), difficulties walking (weeks), subjective voiding difficulties (1d) | Muscle weakness in lower extremities (4/5), hypoesthesia sub T12, decreased anal sphincter contraction, gait ataxia, hyperreflexia of lower extremities | Complete resection, laminoplasty T7-11 | Minimal ataxia, complete resolution of other symptoms (6m) | None | NA |
| | | L3 | Rim-enhancing intradural cystic lesion at the L3 level | Back and leg pain (3m), difficulties walking (2w), sensorimotor deficits left leg (2w) | Muscle weakness left leg (thigh flexion 3+/5, other groups 4/5), hypoesthesia L3 and L4 left, difficulties walking | Complete resection, laminectomy L3, undercutting L2 and L4 | Complete resolution of all symptoms (6m) | | |
| Bozyigit et al., 2022, case report | 1, 66y, F | T8-9 | Cyst with peripheral rim enhancement | Back pain with radiation to the left side (months) | No pathological findings | Complete resection, T8-9 laminectomy | NA | None | NA |
| Ursini et al., 2021, case report | 1, 23y, M | L1-S1 | Multilobulated lesion with cysts and vertebral scalloping | Progressive walking impairment (NA), weakness and pain in the lower limbs bilaterally (NA) | Hypoesthesia and paresthesia on the left side | Resection, vertebroplasty and T11-12-L1-S1 arthrodesis | Neurological improvement, still requiring walking aid (2y) | NA | NA |
| Samman et al., 2021, case report and literature review | 1, 80 y, M | T6-7 | Cyst with peripheral contrast enhancement and focal nodular wall thickening | Mid-thoracic back pain with bilateral radiation following the T6 dermatome (3m) | Up-going plantar reflex without weakness | Complete resection, T5-7 laminectomy | Complete resolution of symptoms (2m) | None | NA |
| Yu et al., 2020, case report and literature review | 1, 66y, M | L1-S3 | Multilobulated lesion with large cystic portion | Leg pain and paresthesia (2y with aggravation over 6m) | Motor weakness of lower extremities 4/5 | Partial resection, L1-S2 laminectomy | Complete recovery of voiding difficulties and leg weakness (2y) | Transient voiding difficulties | NA |
| Hamabe et al., 2019, case report | 1, 48y, F | L4-S1 initially, relocation L2-L4 after 6m | Entirely cystic | Dysuria and right leg pain (gradually worsening over 3w) | NA | 1. Initial conservative approach: diagnostic lumbar puncture with improvement of bladder function<br>2. Resection 8m after diagnosis | Complete resolution of symptoms (1m) | NA | NA |
| Gandhoke et al., 2018, case report | 1, 38y, M | C2-4 | Solid-cystic | Neck pain radiating to left upper extremity (8m) | Isolated hyperreflexia without deficits (on admission), severe neck pain and acute spastic quadriparesis 2/5 with C5 sensory level (<1d) | Complete resection, resection of the attached dorsal nerve root, partial C2-4 laminectomy | Complete resolution of symptoms (5d) | NA | Intratumoral hemorrhage diagnosed intra-operatively, which may potentially explain the clinical deterioration |
| Zimering et al., 2017, case report | 1, 36y, F | L1-2 (n = 1) and L5 (n = 2) | L1-2: heterogeneous cystic lesion (n = 1), L5-S1: | Low back pain (NA) with deterioration during pregnancy and following delivery, | No pathological findings | Complete resection of the two largest lesions, biopsy of smaller nerve rootlet and lumbar nerve | Complete resolution of symptoms, growth of remaining tumors (6m) | Transient voiding difficulties | Preoperative suspicion of myxopapillary ependymoma, intraoperatively, at least 6 small tumors |

(*continued on next page*)

**Table 1** (*continued*)

| Authors, year, study type | Number of patients, age, gender | Spinal levels | Radiographic features | History (duration) | Clinical examination | Therapy | Outcome (date of last follow-up) | Adverse Events | Comments |
|---|---|---|---|---|---|---|---|---|---|
| | | | enhancing mass (n = 2) | unable to walk due to pain (since delivery) | | sheath tumors, T12-L2 laminectomy | | | were visualized along nerve rootlets, which were not resected |
| Kumar et al., 2017, case report | 1, 60y, M | T4-5 | Entirely cystic | Back pain with radiation along chest wall and bilateral lower limbs weakness (8m), urinary dribbling (2m) | Spastic paraparesis (left 0/5, right 3–4/5), hyperreflexia of lower limbs with up-going plantar reflex, T6 sensory level | Complete resection, thoracic laminectomy | Complete resolution of symptoms (2y and 9m) | NA | The lesion was attached to the T4 dorsal root |
| Attiah et al., 2016, case report | 1, 58y, F | S2-3 | Cystic mass | Lower back and left buttock pain (1y), occasional pain and tingling sensations in left leg (NA) | Loss of lumbar lordosis, hyperreflexia of lower extremities | Complete resection, S1-3 laminectomy | Pain improvement (1m) | None | Associated with S2 nerve root |
| Karaca et al., 2015, case report | 1, 21y, F | L5-S1 | Cystic mass | Back pain and walking difficulty (4–5m) | NA | Complete resection | Complete resolution of symptoms (1y) | NA | NA |
| Jenkins et al., 2015, case report and literature review | 1, 62y, M | L2-3 | Lesion with cystic changes | Pain and numbness in right L4 dermatome (NA), unable to walk because of pain (NA) | Weakness of right dorsiflexion 4/5 | Complete resection of tumor and nerve root, L2-3 laminectomy | Complete resolution of symptoms (11y) | NA | The lesion showed torsion and subsequent hemorrhage |
| Corley et al., 2014, case report | 1, 44y, F | L2-4 | Solid-cystic mass with heterogeneous enhancement with vertebral scalloping | New onset bilateral lower extremity weakness and numbness (NA), resection of intradural schwannoma (15y), external beam radiation for recurrence (10y) | Weakness in lower extremities, decreased reflexes and sensation, gait difficulties | Initial treatment with steroids, the patient refused surgery | NA | NA | Diagnosis was known because the lesion was a recurrence of a schwannoma resected 15 years before |
| Savardekar et al., 2014, case series | 6,1: 35y, M2: 45y, F3: 45y, M4: 39y, F5: 41y, M6: 25y, F | Thoracolumbar | Giant multilobulated cyst | Backache, motor deficit below L1 (NA) | NA | Complete resection, T11-L1 laminectomy | Symptoms improved (NA) | NA | NA |
| | | L2-4 | Unilobulated cyst with rim-enhancement | Backache, focal deficit at L3-4 level (NA) | Motor weakness knee and ankle right 3/5 and left 4/5, L2 sensory level | Complete resection, L2-4 laminectomy | Full recovery of motor and sensory function (6m) | None | |
| | | Thoracolumbar | Multilobulated cyst | Progressive motor deficit below L1 (NA) | NA | Complete resection, T11-L1 laminectomy | Symptoms resolved (NA) | NA | |
| | | Thoracal | Unilobulated cyst | Spastic paraparesis (NA) | NA | Complete resection, T2-3 hemilaminectomy | Paraparesis improved (NA) | NA | |
| | | Thoracal | Cyst with lobulations at lower end | Spastic paraparesis (NA) | NA | Complete resection, T3-5 hemilaminectomy | Motor function improved, spasticity resolved (NA) | NA | |
| | | T11-L5 | Giant multilobulated cyst | Flaccid paraparesis, bowel and bladder dysfunction (2mo) | Flaccid paraparesis 2/5 | First L3-4 hemilaminectomy and cyst drainage and biopsy. Second T11-L5 laminoplasty and complete resection | Partial improvement of all symptoms (3m) | None | |

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**Table 1** (*continued*)

| Authors, year, study type | Number of patients, age, gender | Spinal levels | Radiographic features | History (duration) | Clinical examination | Therapy | Outcome (date of last follow-up) | Adverse Events | Comments |
|---|---|---|---|---|---|---|---|---|---|
| Netra et al., 2014, case series | 12, mean age: 52.75y,7M/5F | Cervical (n = 3), lumbar (n = 8), thoracolumbar (T6-L1, n = 1), mean number of segments: 2.5 | Cystic lesions without septation | Cervical: neck pain, paraparesis, radiating pain, myelopathy (NA) Lumbar: radicular pain, motor weakness, myelopathy, non-specific back pain, long-term history of vague flank pain, bladder or bowel incontinence, radiating pain in the bilateral lower limbs (NA) | NA | All surgery, not further specified | All patients achieved complete relief (6m to 2y) | NA | NA |
| Wu et al., 2013, case report | 2, 1: 62y, M 2: 63y, M | L2-3 | Both entirely cystic with rim enhancement | Pain and motor weakness in both lower extremities (1y) | Bilateral leg weakness 4/5 with hyperreflexia | Complete resection, laminectomy L2-3 | Significant pain relief (2y) | NA | NA |
| | | L4-5 | | Right leg radicular pain (6m) | Right leg motor weakness 3/5 with absent knee and ankle reflex | Complete resection, L3-5 laminectomy | Complete resolution of symptoms (2y) | | |
| Albert et al., 2012, case report | 1, 50y, M | C2-5 | Cystic mass | Neck and bilateral arm pain, paresthesia (2y) | Left sided weakness of arm, loss of reflexes and reduced sensation along left ulnar forearm and hand | Complete resection, C2-5 laminectomy | Complete resolution of symptoms (18m) | NA | Old focal hemorrhage was found |
| Hsieh et al., 2011, case report | 1, 51y, F | L4-5 | Cystic mass with rim enhancement | Lower back pain followed by bilateral radiating pain in lower legs (4m), L3-4 fusion (9y) | Weakness in left lower leg 4/5, dysesthesia in L4-S1 dermatome | Complete resection, L4-5 laminectomy | Complete resolution of symptoms (NA) | NA | Intratumoral hematoma was found |
| Wilkinson et al., 2010, case report | 1, 38y, F | L3-S2 (cystic part L5-S2) | Solid mass with contrast enhancement above cystic collection with rim enhancement, vertebral scalloping | Low back pain (>10y), increase in back pain and right-sided leg pain (1m) | Weakness of right hamstring, tibialis anterior, and gastrocnemius 3–4/5, numbness in L4-S1 dermatome right, absent right ankle reflex | Complete resection, L3-5 laminectomy | No back pain, other symptoms not reported, new neurologic deficits (see AEs) (21m) | Worsening of right foot drop 1/5, asymptomatic pseudomeningocele that subsided spontaneously over three weeks, bladder dysfunctions | Very large solid part |
| Vikram et al., 2010, case report | 1, 40y, M | C2-7 | Cyst with rim enhancement | Progressive neck pain with discomfort while using hand for fine motor activities (NA), gait disturbance (NA) | Quadriparesis 4/5 with increased tone and hyperreflexia | Complete resection, C2-7 right hemilaminectomy | Complete resolution of symptoms (NA) | NA | Attached to C2 posterior root |
| Santhosh et al., 2009, case series | 2, 1: 25y, F 2: 65y, F | L5-S1 | Large multilobulated cyst with vertebral scalloping, fluid levels, and rim enhancement | Low back pain with radiation in both lower limbs (9m) | Weakness of left hip abduction 4/5, sluggish bilateral knee jerks | Surgery | NA | NA | 3 cases are reported but one was excluded because mostly extraspinal |
| | | L1-S1 | Large multilobulated cyst with vertebral scalloping, fluid | Progressive weakness of both lower limbs (1y), bedridden (NA), urinary retention (NA) | Weakness in hip flexors/extensors 4/5, abductors/adductors 2/5, knee flexors/ | Surgery | | | |

(*continued on next page*)

**Table 1** (*continued*)

| Authors, year, study type | Number of patients, age, gender | Spinal levels | Radiographic features | History (duration) | Clinical examination | Therapy | Outcome (date of last follow-up) | Adverse Events | Comments |
|---|---|---|---|---|---|---|---|---|---|
| | | | levels, rim/septal enhancement | | extensors 4/5, dorsiflexors 1/5, plantar flexors 3/5, absent reflexes, impairment of vibration sense, poor anal tone | | | | |
| Kasliwal et al., 2008, case report | 1, 70y, M | L1-2 | Entirely cystic with rim-enhancement | Low back pain (6m) | No pathological findings | Complete resection, L1-2 laminectomy | No new neurologic deficit (NA) | None | NA |
| Jaiswal et al., 2008, case report | 1, 30y, M | L3-S2 | Cyst with septation, heterogeneous rim enhancement, and vertebral scalloping | Progressive low back pain with radiation in both lower limbs (2y) | Absent bilateral ankle reflexes | Complete resection, L3-S2 laminectomy | Complete resolution of symptoms (2y) | NA | NA |
| Akhaddar et al., 2008, case report | 1, 50y, M | L4 | Cyst with small posterior nodular lesion, rim and nodular enhancement | Low back pain (4y), right leg pain (recent) | No pathological findings | Complete resection, L3-5 laminectomy | Complete resolution of symptoms (2y) | None | Attached to right L4 root |
| Saiful Azli et al., 2007, case report | 1, 54y, M | L3-5 | Cyst with heterogeneous enhancement and fluid levels | Progressive low back pain (2y), worsening with radiation in right leg (3m) | Positive straight leg raising test right | Complete resection, L3-5 laminectomy | Complete resolution of symptoms (NA) | None | Possibly attached to L4 |
| Karataş et al., 2007, case report | 1, 27y, F | T11-12 | Cyst with rim enhancement | Back pain and walking difficulty (7m) | Paraparesis 4/5, T12 sensory level | Complete resection, T11-12 laminectomy | Complete resolution of symptoms (3 m) | NA | NA |
| Kobayashi et al., 2007, case report | 1, 32y, M | L5-S1 | Solid-cystic with enhancement | Lower back pain (1.5y) with radiation in right leg (NA), numbness of lateral side of right foot (3m) | Positive straight leg raising test right, hypesthesia in S1 dermatome right | Complete resection | Improvement of hypesthesia, complete relief of other symptoms (3y) | None | NA |
| Borges et al., 2005, case report | 1, 55y, M | L4-5 | Almost entirely cystic lesion with rim enhancement | Back pain with radiation in lower limbs (1y) | No pathological findings | Complete resection | Complete resolution of symptoms (12 m) | None | NA |
| Alkhani and Al-Zahrani, 2005, case report | 1, 20y, M | Two lesions, 1: T12 2: L1 | Two cysts with rim enhancement | Progressive paraparesis and paresthesia of lower-extremities (4m) | Paraparesis 4+/5, slight hyperreflexia with up-going plantar reflex | Complete resection of both lesions, T12-L1 laminectomy | Improvement of symptoms (4m) | None | MRI-features of lesion 2 were suggestive for an inflammatory or infectious process |
| Thomé et al., 2004, case report | 1, 47y, F | T6-7 | Cyst with rim enhancement as well as soli extradural lesion (hemangioma) | Progressive back pain with radiation to left costal arch (1y), deterioration with sensory deficits, paraparesis, ataxia, urinary retention (2w) | Paraparesis 4/5 and T9 sensory level | Complete resection, T6-7 laminectomy | Minimal ataxia, complete relief of other symptoms (1y) | None | The schwannoma was neighboring an epidural cavernous hemangioma, separated with dura |
| Kagaya et al., 2000, case report | 1, 57y, F | L3 - S1 | Solid lesion multiple cysts | Low back pain (10y), weakness and numbness both lower extremities since lumbar anesthesia (9y) | Gait disorders (NA), sever urinary retention (NA), constipation (NA), hypoesthesia L3-S1 (NA), weakness of | Partial resection from posterior, excision of tumor from vertebral bodies from anterior, laminectomy L3-5, posterior fusion from L2 | (Almost) complete relief of low back pain, great decrease of hypoesthesia, mild recovery of motor, bladder and | None | NA |

(*continued on next page*)

**Table 1** (*continued*)

| Authors, year, study type | Number of patients, age, gender | Spinal levels | Radiographic features | History (duration) | Clinical examination | Therapy | Outcome (date of last follow-up) | Adverse Events | Comments |
|---|---|---|---|---|---|---|---|---|---|
| | | | | | ankle dorsiflexion, ankle plantar flexion, toes extension, and toes flexion M2-4 (NA) | to pelvis, L3-S1 intervertebral body fusion | bowel function, walk with cane (3y4m) | | |
| Guida, 1997, case report | 1, 65y, F | L1/2 - caudalmost spinal canal | Cystic mass lesion | Deconditioning after prolonged hospitalization for perforated duodenal ulcer (6w) | Marked paresis of lower extremities (NA), saddle anesthesia (NA), bladder and bowel dysfunction (NA) | Laminectomy and resection | Minimal improvement of motor strength and bowel-bladder dysfunction (NA), walk with leg braces and walker | NA | NA |
| Shiono et al., 1995, case series | 2,<br>1: 40y, M<br>2: 51y, M | Both lumbar | Both cysts with rim enhancement | Low back pain with radiation in left leg (2mo)<br>Progressive bilateral thigh numbness (NA) | NA | Both surgery | NA | NA | One of the lesions showed signs of bleeding |
| Sato et al., 1993, case report | 1, 45y, F | C4-5 | Solid-cystic with rim enhancement | Cervical pain, motor weakness of right upper extremity, numbness of right fingers (NA) | NA | Resection, C4-5 laminectomy | NA | None | No full-text available in English, data extracted from the abstract |
| Shen et al., 1992, case report | 1, 41y, M | T12-L1 | Cyst with rim enhancement | Low back pain with radiation in both legs (2y), paraparesis and bladder/bowel dysfunction (3m) | T12 sensory level, muscle atrophy, areflexia of lower limbs | Complete resection, laminectomy | NA | NA | Attached to nerve root of cauda equina |
| Hisaoka et al., 1991, case report | 1, 52y, M | T10-L1 | Cyst with rim enhancement | Pain and numbness of left buttock with radiation in foot (3y), bilateral paralysis of lower extremities and unable to walk (NA) | NA | Partial resection | Improvement of pain and hypesthesia, aggravation of paralysis left (NA) | Deterioration of motor weakness | The tumor was diagnosed in 1987 and the patient received surgery in 1989 because of the new motor deficits because of hemorrhage in the tumor |

authors were reported with "NA". Relevant items for the section "History", "Examination", and "Outcome" were duration of symptoms, pain localization, motor weakness (documented with grade from 0 to 5), sensory changes, reflexes (deep tendon reflexes and/or pathologic reflexes), sphincter function, gait disturbance, other signs of ataxia/myelopathy. Funding sources and conflicts of interest were not reviewed.

### 2.3. Statistical analysis & data availability

This review was not registered and there is no study protocol available. Since only case reports/small case series were available, we did not conduct any risk of bias assessment. Statistical analyses were performed using R (R: A Language and Environment for Statistical Computing, 2019). The two additional cases reported in this article were included in all analyses, as well as in relevant figures and tables. Means are presented accompanied by standard deviation (SD), medians by interquartile range (IQR), and percentages by 95% confidence intervals (CI). The data used and produced in this study can be provided upon reasonable request (list of PMID, data extracted from included studies and used for all analyses).

## 3. Results

### 3.1. Epidemiology and basic features

A total of 35 manuscripts were found which reported on 54 cases (56 cases including ours) of (largely) cystic, intradural extramedullary schwannomas (Table 1). Mean age at presentation was 47.7 years (SD ± 13.0 years, Fig. 2) and 32/56 of patients were males (57.1%, CI: 44.1–69.2). Most patients had lumbar (24/56, 42.9%, 30.4–55.9) or lumbosacral (10/56, 17.9%, 5.4–30.9) lesions, the other results are summarized in Table 2. The exact location was known for 40 lesions and is illustrated (red and blue) in Fig. 3 along with the distribution of lesions among all spinal segments (black). In all but one case, treatment was surgical. In the only case treated conservatively, an operation was indicated but refused by the patient (Corley et al., 2014).

**Table 2**
Localization of cystic, intradural extramedullary schwannomas.

| Level | Number of cases | Percentage | 95% CI |
|---|---|---|---|
| Cervical | 7 | 12.5% | 0–25.6 |
| Thoracic | 9 | 16.1% | 3.6–29.1 |
| Thoracolumbar | 5 | 8.9% | 0–22.0 |
| Lumbar | 24 | 42.9% | 30.4–55.9 |
| Lumbosacral | 10 | 17.9% | 5.4–30.9 |
| Sacral | 1 | 1.8% | 0–14.8 |

### 3.2. Signs and symptoms

The most common symptoms at admission were pain (36/44, 81.8%, CI 68.0–90.5) and muscle weakness (30/44, 68.2%, CI 53.4–80.0); other results are listed in Table 3. 37/44 of patients showed at least one abnormality in neurological status (84.1%, CI 70.6–92.1) ranging from abnormal reflexes to spastic paraparesis (Tables 1 and 3).

### 3.3. Complications and outcome

Complications occurred following four surgeries (4/19, 21.1%, CI 8.5–43.3). These complications were transient voiding difficulties (n = 2), worsening of foot drop with asymptomatic pseudomeningocele and bladder dysfunctions (n = 1), and deterioration of motor weakness (n = 1). The vast majority of patients for whom follow-up data were available showed an improvement in symptoms following surgery (45/47, 95.7%, 85.8–98.8). The median follow-up time was 52.2 weeks (IQR 84.8). The two remaining patients showed postoperative improvements of their pain scores but had worsening muscle weakness. The majority of patients showed a complete relief of symptoms (33/47, 70.2%, 56.0–81.3; Tables 1 and 3).

## 4. Case presentations

### 4.1. Case 1

A 44-year-old female with a six-year history of L4/5-disc herniation was referred by the general practitioner because of worsening back pain

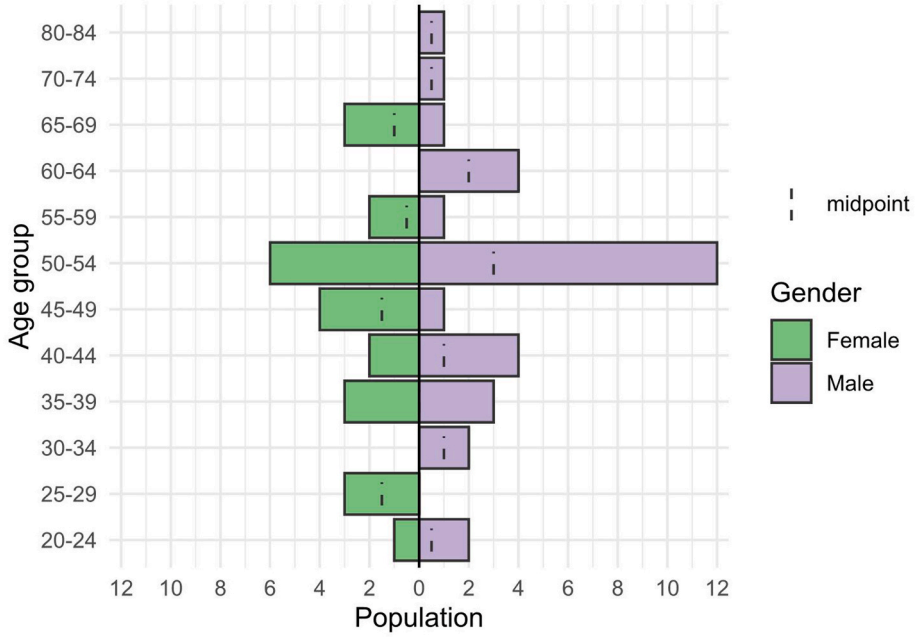


**Fig. 2.** Age and gender distribution of all patients included in the literature review
All patients were included in figure (54 patients from the literature, 2 patients from our centers). The most prevalent gender-age group was male aged 50 to 54, followed by female aged 50 to 54.

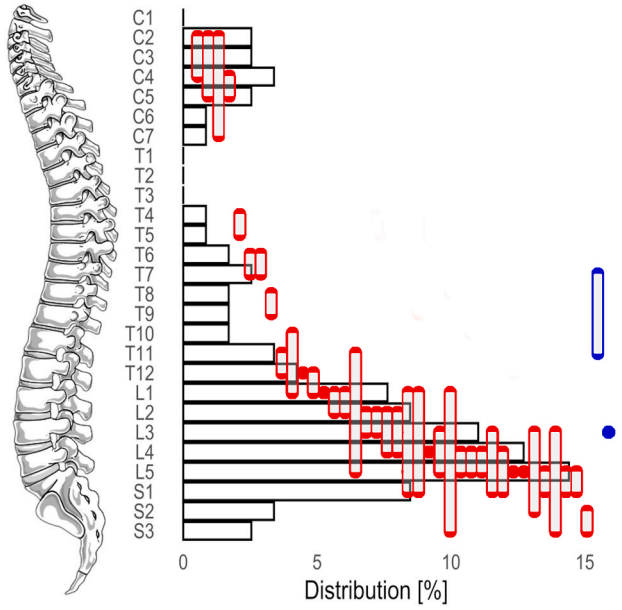


**Fig. 3.** Segmental distribution of cystic, intradural extramedullary schwannomas reported in the literature
All lesions for which the location was exactly known (40 lesions from 37 patients) were included in the figure. The histogram (black) provides the distribution of schwannomas among each spinal segments (percentage of the total). The position of each included lesion is illustrated by the red bars (e.g., the first lesion on the left extends from C2 to C4). Monosegmental lesions were illustrated by a dot. Additionally, the lesions described in our case-reports are shown in blue. The Figure was partly generated using Servier Medical Art (vertebral column on the left, transformed into monochrome), provided by Servier, licensed under a Creative Commons Attribution 3.0 unported license. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

**Table 3**
Symptoms on admission, outcome and adverse events following surgery of patients with cystic, intradural extramedullary schwannomas.

| | Number of cases | Percentage | 95% CI |
|---|---|---|---|
| **Pain** | 36/44 | 81.8% | 68.0–90.5 |
| **Muscle weakness** | 30/44 | 68.2% | 53.4–80.0 |
| **Sensory disturbance** | 24/44 | 54.5% | 40.1–68.3 |
| **Gait difficulties** | 13/44 | 29.5% | 18.2–44.2 |
| **Sphincter dysfunction** | 9/44 | 20.5% | 11.2–34.5 |
| **Abnormal reflexes** | 19/44 | 43.2 | 29.7–57.8 |
| **Improvement of symptoms following surgery** | 45/47 | 95.7% | 85.8–98.8 |
| **Adverse events** | 4/19 | 21.1% | 8.5–43.3 |

Symptoms and neurological status were extracted from all articles in Table 1. When the patient's symptoms and neurological status were mentioned, but certain aspects were omitted (e.g. motor weakness), it was considered that the patient did not have such symptoms. When the patient's preoperative/postoperative clinical condition was not mentioned, the patient was excluded from the corresponding analysis. When the presence/absence of postoperative complications was not mentioned, the patient was not included in the corresponding analysis, which may explain the complication rate of 21%.

with hypoesthesia in both lower legs and gait ataxia. Two days prior to the consultation, MRI of the brain and the lumbar spine were conducted, which showed a protrusion of the L4/5 disc with suspicion of bilateral recess stenosis and compression of the descending L5 nerve roots. The lumbar X-ray showed no abnormalities. During the consultation, the patient described back pain for several years, subjective leg weakness over the past two months, and more recently, difficulties walking with a notable deterioration over the past week. Moreover, she reported voiding difficulties over the past day (subjective feeling of urinary retention).

On clinical examination, the patient had local tenderness around the thoracolumbar junction, muscle weakness in the lower extremities (thigh flexion on the right 4/5 and on the left 4-/5, knee extension bilaterally 4/5), hypoesthesia below T12, and decreased voluntary contraction of the anal sphincter in the presence of a preserved tone. The sensation of urinary retention could not be objectified as there was no retention on sonography. In addition, there was a pronounced gait ataxia with hyperreflexia of the lower extremities. The patient had no other relevant symptoms, history, medication, or laboratory changes.

On the MRI studies of the lumbar spine, an intradural cystic lesion of the lower thoracic spine was merely identified (Fig. 4, panel A) and the patient therefore underwent urgent MR imaging of the thoracic spine. The MRI of the thoracic spine, which was conducted without contrast agent at the patient's explicit request, showed a cystic, septated, intradural-extramedullary mass extending from T7 to T11 (1x1.3 × 10cm), with protrusion into the left T10 neuroforamen (Fig. 4). There was no diffusion restriction and no evidence of an infiltration into surrounding tissues. There was evidence of minimal intralesional hemorrhage and slight scalloping of the T9 vertebral body. The spinal cord was clearly displaced antero-laterally, with edema distal to the mass a T12-L1.

An urgent laminoplasty T7-T11 with complete microsurgical resection of the tumor was performed the same day (Fig. 5). The fascicles on the affected segments (T7-11) were adherent to the lesion and thinned out, but they could be partially preserved. There were no adverse events during or following surgery. The histopathological diagnosis of schwannoma was established. An MRI of the thoracic spine was conducted 3 days postoperatively and there was no evidence of tumor remnants. The intra- and postoperative course was uncomplicated and there was a gradual improvement of the neurological state. The patient was discharged to rehabilitation 10 days following surgery. At the follow-up consultation 6 weeks after surgery, the patient's symptoms had greatly improved. She showed slight back (visual analogue scale (VAS) 1/10) and leg (VAS 2/10) pain as well as a slight residual ataxia in the heel-to-toe walking test, but her sensorimotor and bladder symptoms had completely resolved. The Oswestry Disability Index (ODI) was 6/100 and the Core Outcome Measures Index (COMI) for the Back was 2.9/10. There was further, almost complete recovery with persisting minimal ataxia at the 6-months follow-up.

### *4.2. Case 2*

A 35-year-old male with a three-month history of back and bilateral leg pain presented with an exacerbation of his pain symptoms along with new onset of neurologic symptoms over the past 3 weeks. On clinical examination, the patient had difficulties ambulating, diffuse muscle weakness in the left lower extremity (thigh flexion 3+/5, other key muscle groups 4/5) with 5/5 muscle strength in his right lower extremety, hypoesthesia in the distribution of the left L3 and L4 dermatomes, along with normal reflex status and preserved sphincter function. The patient had no other relevant symptoms, past medical history, medication, or laboratory changes.

On the MRI studies of the lumbar spine, a rim-enhancing intradural cystic lesion at the L3 level was identified (Fig. 6, panel A). A L3 laminectomy with a laminotomy of the L2 and L4 laminae was performed. Intraoperative ultrasound enabled to precisely delineate the margins of the tumor (Fig. 6, panel B). Under neurophysiological monitoring (MEP, SSEP, direct nerve stimulation with evoked EMG monitoring), the adherent nerve roots were microsurgically dissected from the capsule and an ultrasonic aspirator was used decompress the cystic contents in order to collapse the tumor while keeping the capsule intact. One single non-eloquent sensory fascicle entering and exiting the tumor was divided. A complete microsurgical resection was achieved. There was no intra- and postoperative adverse event. The histopathologic

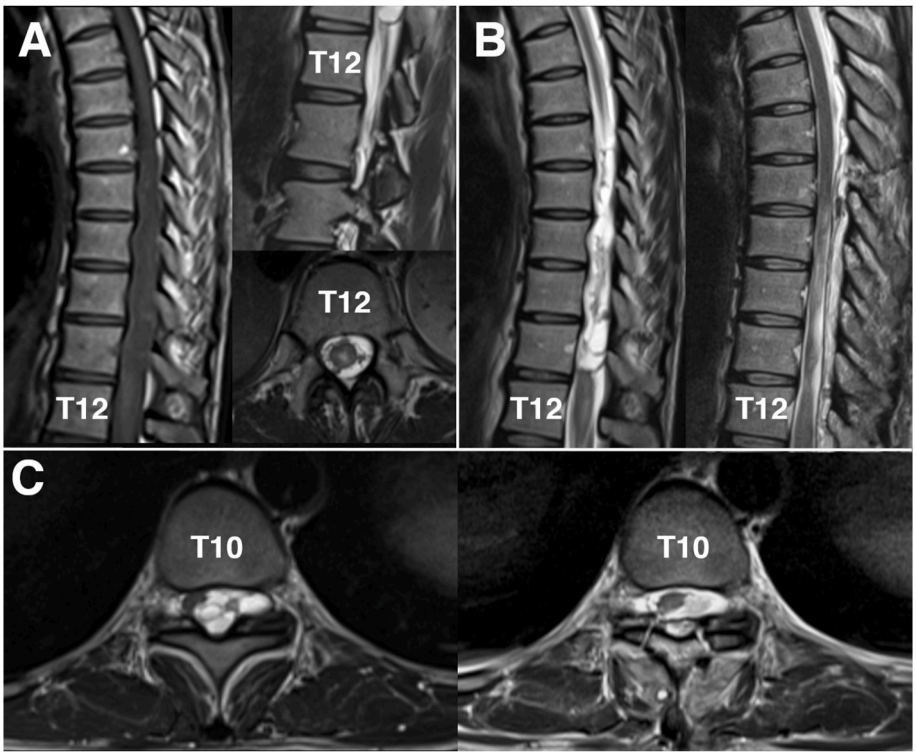


**Fig. 4.** Pre- and postoperative MRI findings of case 1
**A:** left: preoperative sagittal T1-weighted MRI showing an (hypo-)iso-intense lesion upper right: preoperative lumbar sagittal T2-weighted MRI showing the cystic lesion at the edge of the picture lower right: preoperative axial T2-weighted MRI (T12 level) showing intramedullary edema distal to the mass. **B:** left: preoperative sagittal T2-weighted MRI showing the cystic lesion right: postoperative sagittal T2-weighted MRI showing no tumor remnant (3 days after surgery). **C:** left preoperative axial T2-weighted MRI showing the septated, cystic lesion with displacement of the spinal cord antero-laterally (T10 level) right: postoperative axial T2-weighted MRI at the same level showing regressive mass effect (3 days after surgery).

examination showed typical Antoni A and B patterns, with a strong nuclear and cytoplasmic S100 immunoreactivity, typical for a schwannoma. The patient was discharged home 4 days after surgery, with a total resolution of pain and neurologic deficits. At the follow-up consultations 3 and 6 months after surgery, the patient was still asymptomatic.

## 5. Discussion

### 5.1. Key findings and implications

We present the most comprehensive review of the literature about spinal cystic schwannomas. To date, 56 patients with non-syndromal (mostly) cystic intradural-extramedullary schwannomas were reported (Table 1). The most prevalent subpopulation of patients were 50–55 year-old males (Fig. 2), and the vast majority of lesions were located or extended to the lumbar spine (Table 2), most commonly spanning over two spinal segments (Fig. 3); only six lesions from four patients were monosegmental while the largest lesion extending over eight spinal segments from L1 to S3. In all reported patients, a resection of the lesion was performed; one patient refused surgery but was not followed-up (Corley et al., 2014). Most patients presented with pain and neurological abnormalities (Table 3); symptoms which often resolved and almost always improved following surgery. Intra- and/or postoperative complications were rare but they may have been underreported since articles often did not explicitely address this topic. Thus, this review confirms that complete surgical resection of spinal cystic schwannomas is safe and has good to excellent outcomes.

### 5.2. Histopathological features

Macroscopically, schwannomas present as smooth nodular tumors with a tan or yellow cut surface (Hilton and Hanemann, 2014). Microscopically, they usually consist of a well delineated capsule surrounding highly cellular zones with wavy nuclei palisades known as Verocay bodies (Antoni A pattern) and less cellular regions with tinny and wispy cells (Antoni B pattern). Within Antoni B patterns, degenerative changes such as thrombosis, hemorrhage, vascular wall ectasia with hyaline thickening, calcification, lipidization, necrosis, and micro-to macrocystic areas may be present (Hilton and Hanemann, 2014; Wippold et al., 2007). Known variants of schwannomas include cellular schwannoma, plexiform schwannoma, epithelioid cell schwannoma, reticular/microcystic schwannoma, and ancient schwannoma (Magro et al., 2022). The latter has been particularly associated with degenerative changes such as cysts or hemorrhages. A current hypothesis for the pathogenesis of schwannomas is that they result from uncontrolled proliferation of Schwann cells in response to nerve injury, which is thought to occur due to a loss-of-function mutation of the Nf2 tumor-suppressor gene and interactions with the cellular microenvironment (Helbing et al., 2020). Other genes such as CHD4, FAT1, KMT2D, MED12 and SUFU are related to sporadic spinal schwannomas, as familial spinal schwannomas are often associated with SMARCB1, SMARCE1 or LZTR1 (Gao et al., 2020).

### 5.3. Radiological features and differential diagnosis

Typical MRI features of schwannomas include T1 iso-to hypointensity (compared to the spinal cord), T2 hyperintensity, and post-contrast enhancement (Crist et al., 2017; Samman et al., 2021). However, T2 signal patterns can greatly vary from hypointense to hyperintense signal

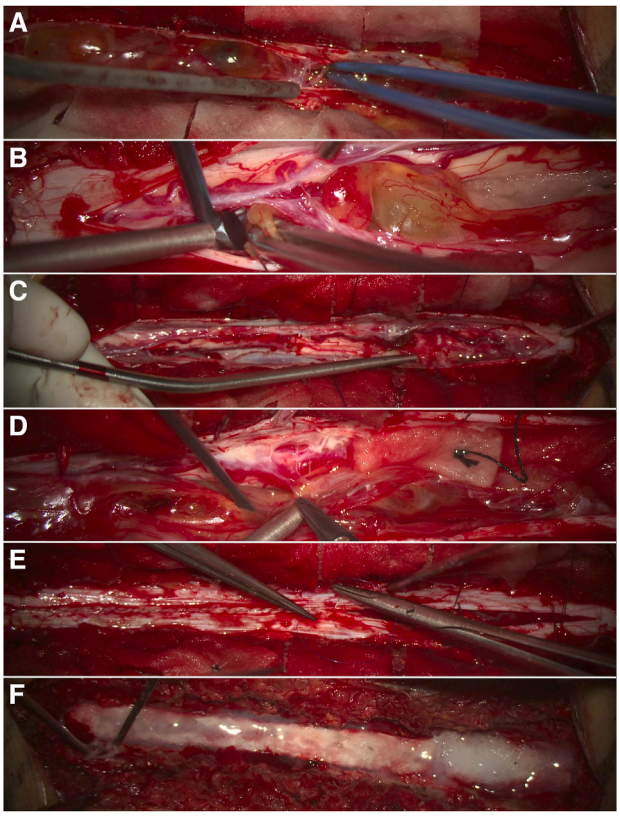


**Fig. 5.** Intraoperative findings for case 1
**A:** After T7-11 laminoplasty, the cystic, yellowish gelatinous lesion was revealed. The spinal cord was displaced ventrally and laterally to the right. Dorsal, sensitive nerve branches ran across the lesion, which were somewhat adherent. These nerves were carefully dissected from the lesion in microsurgical fashion. **B:** After some internal debulking, the tumor was dissected and separated from the spinal cord. **C:** After complete resection, the spinal cord remained adherent to the dura and therefore continued to remain displaced laterally. **D:** Adhesiolysis was conducted to release the tension of the spinal cord and allow for its physiological repositioning. **E:** Dural closure was performed using a 6-0 non-absorbable running suture. **F:** Reinforcement of the suture using a fibrin sealant patch and fibrin glue.

changes depending on the cellularity of the lesion and microstructural abnormalities such as the presence of hemorrhage, necrosis, or cysts (Ito et al., 2018). Signal heterogeneity is therefore most commonly seen within Antony B areas (Koeller and Shih, 2019). T1 hyperintensity is seen in melanotic schwannomas due to paramagnetic free radicals found within melanin (Koeller and Shih, 2019). Differential diagnosis of intradural extramedullary tumors include meningioma, schwannoma, neurofibroma, malignant peripheral nerve sheath tumor, myxopapillary ependymoma, paraganglioma, solitary fibrous tumors, and metastases (Koeller and Shih, 2019). The differential diagnosis of cystic lesions may be more difficult to establish since differences in signal intensities are usually confined to the solid portions of tumors. Ultimately, the differential diagnosis of intraspinal cystic lesions includes hydatid, neurenteric, epidermoid, dermoid, and arachnoid cysts, but those can usually easily be recognized because of distinct imaging features such as a non-enhancing rim (Samman et al., 2021; Netra et al., 2014). This was more challenging in our first case where no contrast agent was used (due to the patients' explicit request) and an arachnoid cyst could not be excluded. Although the presence of cystic changes is more common in schwannomas, predominantly cystic meningiomas and extramedullary ependymomas have also been described previously (Gkasdaris et al., 2022; Graça et al., 2006). The anatomic location of the lesion (the vast majority of arachnoid cysts are thoracic), (Kalsi et al., 2022) its

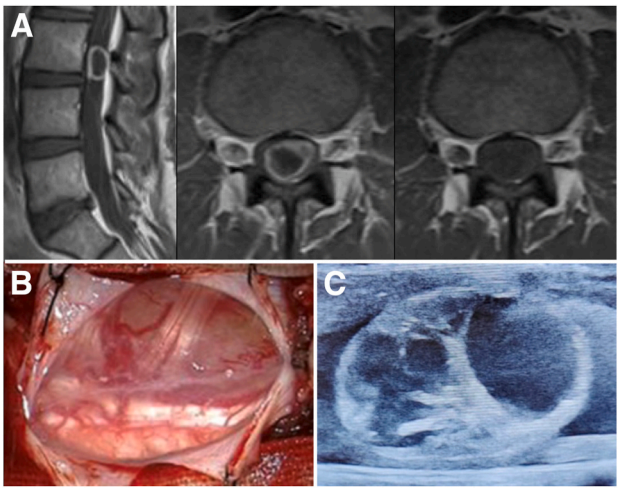


**Fig. 6.** Preoperative MRI findings and intraoperative findings for case 2
**A:** left: preoperative sagittal T1-weighted post-contrast MRI showing a cystic rim-enhancing lesion middle: preoperative axial T1-weighted post-contrast MRI showing displacement of the cauda equina right: preoperative axial T1-weighteed MRI showing the isointense lesion. **B:** intraoperative findings after L3 laminectomy with undercutting of the L2 and L4 laminae and durotomy. **C:** intraoperative ultrasound.

immediate vicinity to the dura (as seen with meningioma) or with the conus medullaris or filum terminale (as seen with ependymoma) may further narrow down the potential differential diagnosis.

### 5.4. Clinical presentation

Patients with intraspinal cystic schwannomas most commonly present with pain and a variable degree of neurologic deficits, which arise from spinal cord compression. Most patients have subtle, long-standing symptoms over the course of months to years prior to diagnosis. Although neurologic abnormalities may be mild and only include abnormal reflexes, sensory disturbances or mild muscle weakness, a considerable number of patients present with disabling symptoms such as gait difficulties (29%) and sphincter dysfunction (20%). Although chronic or subacute symptoms are more frequent because of the benign and slowly growing nature of the lesion, intratumoral hemorrhages can cause a rapid deterioration with acute, and sometimes severe, neurologic deficits (Gandhoke et al., 2018; Jenkins et al., 2015; Hisaoka et al., 1991).

### 5.5. Treatment and outcome

All but one of the patients reported in the literature underwent surgery. This is a self-fulfilling prophecy because definitive diagnosis requires surgery or biopsy, the latter being inadequate in most cases, considering risks associated with close proximity to the spinal cord and nerve structures, the uncertainty of obtaining reasonable specimen in predominantly cystic lesions and the potential cure after complete resection. Within the reported literature, only one patient refused surgery. In this particular patient, the diagnosis was known because the lesion was a recurrent schwannoma years after sub-total resection. The vast majority of patients underwent laminectomy rather than laminoplasty, however, the rationale for this decision was generally not provided. Especially in junctional regions of the spine and in multi-level approaches, we prefer restoring the anatomy by laminoplasty - an approach that appears to be supported by recent literature (Sun et al., 2019; Byvaltsev et al., 2023). In one of our cases, we also chose laminoplasty over laminectomy in order to reduce the risk of post-laminectomy kyphosis and to potentially facilitate the approach in case of a potential future revision surgery.

Regardless of the approach chosen, good exposure is crucial in order to allow adequate microsurgical management of cystic lesions. In order to achieve complete resection while avoiding neurological complications, microdissection of nerve roots away from the capsule of the tumor has to be conducted with great care. This may be facilitated by decompressing the lesion by evacuating the cyst's content while maintaining control of the tumor capsule. In order to better understand the intricate relationship between the lesion and neural structures as well as to preserve functional outcome, we strongly recommend the use of intraoperative ultrasound and neurophysiological recording when available.

Although most patients presented with long-lasting symptoms and neurologic deficits, more than two-third of patients showed complete symptom relief after surgery. However, these results must be interpreted in a context where most studies provided limited post-operative data, using imprecise terms such as "improvement" or "resolution" of symptoms instead of using standardized outcome measures or precise descriptions of the neurological status. Only four surgeries resulted in complications, yet only in 19 patients the authors explicitly specified whether a complication had occurred or not. As a result, the reported complication rate is 21.1% which is consistent with the existing neurosurgical literature (Sarnthein et al., 2022). These findings must be interpreted with caution since both the number of cases with (n = 4) and without complications (n = 15) were likely underestimated. Furthermore, non-neurologic complications may have been underreported since all reported complications were neurologic.

### 5.6. Limitations

The aim of this article was to compile and discuss all documented cases of intraspinal extramedullary schwannomas with predominantly cystic or with very large cystic changes. This definition is subjective, which inevitably introduces a selection bias. We chose not to include only patients with schwannomas described as purely cystic, as it has already been demonstrated that lesions described as such actually have a solid component (De La Peña et al., 2022). We also decided not to define a limit for the cystic/solid ratio, as the required data was missing in most cases. Moreover, we did not restrict the review to predominantly cystic lesions, as this would have been equivalent to setting a cystic/solid ratio of >0.5 (an information that was missing in most cases) and we would have had to exclude some lesions with a very large cystic component. Unarguably, there are reports of schwannomas with large cystic components that were not described as such in the title/abstract of the article and may therefore have been omitted in this review. Furthermore, there is certainly a strong publication bias since patients presenting with small lesions and/or few symptoms are not usually the subjects of case reports. Finally, as only histopathologically confirmed cases of schwannoma were included, some lesions that were not resected and where diagnosis remains unclear were inevitably excluded. We therefore do not know whether and how cystic schwannomas spontaneously progress in conservatively-treated patients.

## 6. Conclusion

Schwannomas are the second most common entity of spinal intradural extramedullary tumors. Although often associated with microcystic changes, they rarely present as predominantly or complete cystic lesions. As a result, such lesions may be misdiagnosed, which can delay or prevent optimal treatment. Complete surgical resection of the cystic lesions is the therapeutic modality of choice. Careful attention to microsurgical principles along with the use of ancillary tools such as intraoperative ultrasound and intraoperative neurophysiologic monitoring may further add to the surgical safety. As seen in our systematic review, complete surgical resection offers good to excellent outcomes for the majority of patients.

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## Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

## Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.bas.2024.102843.

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