



Case Report

Online ISSN (3219-2789)

Multilevel Spinal Schwannomas with Sacrococcygeal Dural Ectasia in a Young Female: A Case Report and Literature Review

Wassan Nori^{1*}, Muna Abdul Ghani Zghair^{2,3}, Nour Fawzi Shaheed¹, Shaymaa Khalid Abdulqader⁴

¹Department of Obstetrics and Gynecology, College of Medicine, Mustansiriyah University, Baghdad 10052, Iraq; ²Department of Radiology, College of Medicine, Mustansiriyah University, Baghdad, Iraq; ³Department of Radiology, Al Yarmouk Teaching Hospital, Baghdad, Iraq; ⁴Department of Radiology, Al-Kindy College of Medicine, University of Baghdad, Baghdad, Iraq

Received: 30 June 2026; Revised: 25 August 2026; Accepted: 30 August 2026

Abstract

Renal cell carcinoma (RCC) is a rare urological malignancy that may present during multilevel spinal schwannoma, a rare type of spinal cord tumor that is commonly observed in the cervical and thoracic spine as a solitary lesion. In this report, we present a case of a 24-year-old woman who experienced a gradual onset of neck and back pain accompanied by upper limb numbness. No focal motor deficits or urinary dysfunctions were reported. The results of the magnetic resonance imaging of the neuroaxis revealed a multilevel thoracic, lumbar, and sacrococcygeal involvement, characterized by a massive extramedullary tumor burden, a highly suspected intramedullary extension, and prominent sacrococcygeal dural ectasia. Only a handful of cases in the literature have described lumbar region involvement by multilevel spinal Schwannoma. No earlier case has documented concurrent lower sacral dural ectasia. Furthermore, we present a comprehensive review regarding the current knowledge, diagnostic challenges, and therapeutic options for multilevel spinal schwannomas of the spinal cord.

Keywords: Multi-level spinal schwannomas; MRI; Sacral dural ectasia; Spinal cord tumors.

الأورام الشفانية متعددة المستويات في العمود الفقري

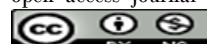
الخلاصة

تُعد الأورام الشفانية متعددة المستويات في العمود الفقري من الأورام النادرة، وغالبًا ما تظهر كافة منفردة في الفقرات العنقية أو الصدرية. نعرض في هذه الدراسة حالة امرأة تبلغ من العمر 24 عامًا، عانت من ألم تدريجي في الرقبة والظهر مصحوب بخدر في الطرفين العلويين، دون وجود ضعف حركي بؤري أو اضطرابات بولية. أظهر التصوير بالرنين المغناطيسي لمحور الجهاز العصبي وجود إصابات متعددة المستويات شملت الفقرات الصدرية والقطنية والمنطقة العجزية العصبية، مع كتلة ورمية كبيرة خارج النخاع، وامتداد محتمل داخل النخاع، إضافة إلى توسع واضح في الأم الجافية بالمنطقة العجزية العصبية. تُعد إصابة المنطقة القطنية في الأورام الشفانية متعددة المستويات نادرة للغاية، ولم يُوثق سابقًا وجودها متزامنة مع توسع الأم الجافية في الجزء السفلي من المنطقة العجزية. كما تتضمن هذه الدراسة مراجعة موجزة للأدبيات الحالية، مع مناقشة التحديات التشخيصية والخيارات العلاجية المتاحة لهذه الحالة النادرة.

* **Corresponding author:** Wassan Nori. Department of Obstetrics and Gynecology, College of Medicine, Mustansiriyah University, Baghdad 10052, Iraq; Email: dr.wassan76@uomustansiriyah.edu.iq

Article citation: Nori W, Zghair MA, Shaheed NF, Abdulqader SK. Multi-level Spinal Schwannomas with Sacrococcygeal Dural Ectasia in a Young Female: A Case Report and Literature Review. *Al-Rafidain J Med Sci.* 2026;11(2):86-90. doi: <https://doi.org/10.54133/ajms.v11i2.3226>

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INTRODUCTION

Schwannomas (SCs) are benign tumors of the central nervous system that were initially documented in 1910 [1]. They arise from Schwann cells located in the nerve sheath, typically in the dorsal sensory roots of the spinal nerves and constitute around 10% of all tumors affecting the spinal region. SCs can occur sporadically or in association with neurofibromatosis-spectrum disorder (NF); they commonly present as solitary cervical and thoracic spine lesions in the intradural-extramedullary compartment [2,3]. While the discovery of multiple SCs strongly indicates an underlying genetic tumor: schwannomatosis [2,4]. Whereas 'schwannoma' describes a histological diagnosis of a single benign nerve sheath tumor, the term 'schwannomatosis' represents a clinical syndrome that presents with multiple schwannomas in one patient, classically in the lack of bilateral vestibular schwannomas that would otherwise indicate NF2. The historical classification system for NF was updated in 2022 by the International Consensus Group on Neurofibromatosis Diagnostic Criteria, which

retired legacy terms like "NF Type 2" and repositioned terminology as described in Figure 1.

Modern Schwannomatosis Classification	NF1	NF2-related Schwannomatosis	SMARCB1-related Schwannomatosis	LZTR1-related Schwannomatosis	Schwannomatosis-NOS
Primary Gene	NF1	NF2	SMARCB1	LZTR1	Unknown
Key Differentiators	Neurofibromas, café-au-lait macules	Vestibular schwannomas, meningiomas	Multiple schwannomas, chronic pain	Multiple schwannomas	Meets clinical criteria

Figure 1: The new classification system was developed per the global International Consensus Group on Neurofibromatosis Diagnostic Criteria (I-NF-DC). The classification system was officially updated to prioritise genetic variants and use Schwannomatosis as the modern overarching umbrella term [3].

Cases that meet clinical criteria without confirmed predisposing genetic testing (NF2-, LZTR1-, or SMARCB1-related) are now designated as Schwannomatosis–Not Otherwise Specified (NOS) [3]. Many hypotheses were proposed to explain why SCs

develop. Notably, the observation of a tumor connection to the nerve root in magnetic resonance imaging (MRI) of the spinal cord aligns with the hypothesis of origin from the nerve root entry zone. However, this anatomical mechanism is insufficient in providing a complete explanation for the formation of multi-level spinal SCs. Current knowledge is also insufficient to draw a firm conclusion regarding the distinctions in the pathogenesis of solitary SCs versus multiple Schwannomatosis, which calls for further research [5]. Patients with SCs typically manifest during their 4th to 5th decade of life, with a higher prevalence in men as compared to women (3:2). The lack of symptoms in earlier stages is often the reason for delayed presentation [6]. Signs and symptoms are usually of neurological origin, and the resulting deficit varies according to the SC's site. Patients suffer from pain, motor, and sensory changes. The loss of sphincter control tends to be a late symptom resulting from severe cord compression. Patients typically exhibit radicular and axial symptoms. The definitive confirmation of SCs based solely on MRI presents a diagnostic challenge without the gold standard of histopathological or genomic correlation [7,8]. A solitary mass located in the cervical and thoracic intradural-extramedullary compartment is a remarkably frequent presentation for these tumors. Conversely, the presentation of multiple concurrent tumors is exceptionally rare; to date, only six cases have been documented in the literature, with our case representing the 7th [1]. Herein, we present a rare case of multiple Schwannomatosis-NOS presented with massive multilevel intradural extramedullary tumors, a highly suspected intramedullary extension, and concurrent sacrococcygeal dural ectasia alongside a review of published literature.

Case Presentation

A neurologist specialist at Al-Yarmouk Hospital in Baghdad, Iraq, referred a 24-year-old female to the MRI unit with a 2-year history of a gradual, insidious onset of pain in the neck, middle, and lower back. The pain was aggravated by activity during the daytime and radiated to both arms and the shoulder region as well as the lateral chest wall and was accompanied by bilateral upper limb numbness. For the last three months, the pain has become more progressive, which has led her to counselling. The medical, surgical, and family history was negative regarding chronic illness and epilepsy. She denied a history of falling, trauma, visual disturbances, and hearing loss. No complaints regarding bladder dysfunction. No history of current drug allergies or intake. There were no prior records of similar cases in her family, nor a history of genetic counselling. Clinical assessment showed the patient was alert and oriented, with normal speech and no mental deficit. Crucially, she had no NF1 stigma (e.g., skin pigmentation). Local examination showed normal muscle bulk, yet there were elements of sensory loss, and no motor or autonomic involvement was seen.

Radiological examination

A magnetic resonance imaging (MRI) of the brain was normal, which ruled out bilateral vestibular tumors. Lumbosacral and lower dorsal spine MRI reveals

multi-level intradural extramedullary and intramedullary rounded masses of variable size distributed along the lumbosacral and lower dorsal region (Figure 2A-B), extending through exit nerve roots (Figure 2C), but no obvious extradural progression.

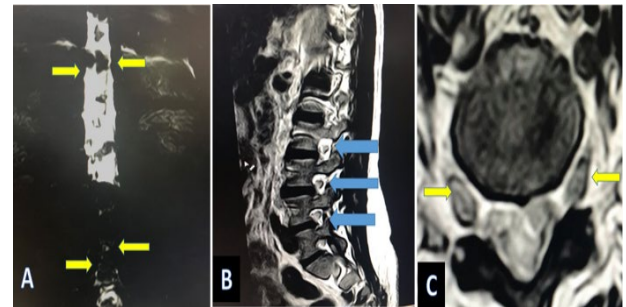


Figure 2: (A) Coronal T2 MRI myelogram showing multiple intradural masses and bilateral defects involving the exit roots of the lower lumbar and sacral spine as well as the lower dorsal region. (B) Sagittal MRI T2 image through the neural foramina showing multi-level nerve root thickening L5, L4, L3, and L2 at the exit nerve root. (C) Axial T2-weighted MRI image at the level of L2 showing bilateral thickened nerve roots extending through the exit foramen as an isointense signal in relation to the spinal canal.

The cauda equina region was compressed by the tumor masses with mild sacrococcygeal dural ectasia (Figure 3A-B). Vertebral bodies, neural arch, vertebral height, and shape showed normal signal intensity. We advised the patient to extend the MRI study to the cervical spines; however, the patient declined. The differential diagnoses of extramedullary intradural masses included meningioma, schwannomas, neurofibromas, and metastasis. The most probable diagnosis in this case, based on the heterogenous T2-weighted MRI signal, normal brain imaging, the lack of NF1 stigma, and the lack of genetic testing under updated consensus naming conventions, is Schwannomatosis-Not Otherwise Specified (NOS). A definitive diagnosis should require a histopathology specimen to rule out concurrent ependymomas.

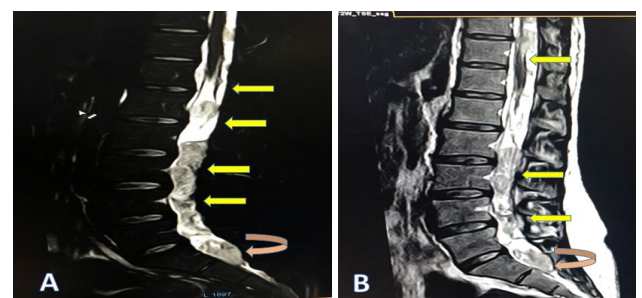


Figure 3: (A) Sagittal MRI T2 SPIR image sequence demonstrates multiple intradural rounded masses (straight arrows) involving lumbar, L1, L3-5, and lower dorsal T11 and 12, as well as sacral spine with moderate sacrococcygeal dural ectasia (curved yellow arrow). (B) Sagittal MRI T2 weighted image TSE showing multiple variable sizes intra-dural extra medullary rounded mass (yellow arrows) involving the lower dorsal, a lumbar and sacrococcygeal region with sacral dural ectasia (curved yellow arrow).

The current case shows multiple spinal Schwannomas, a normal MRI of the brain, and no molecular genomic analysis. Consequently, it is officially classified under these updated consensus naming conventions as Schwannomatosis – Not Otherwise Specified (NOS).

DISCUSSION

Swiatek *et al.* conducted a systematic review in 2021 that included 165 reported cases of the entire neuraxis reaching spinal schwannomas [1]. Of those cases, 6 had extremely rare presentations of intramedullary

schwannomas; the current case highlights an equally extraordinary presentation of a massive, multi-level intradural-extramedullary tumor burden with a highly suspected intramedullary extension. The current case is unique in multiple ways. To begin with, multilevel spinal schwannomas were reported in 6/165 cases only; females predominated in 4/7 cases (Table 1).

Table 1: The previously reported multilevel spinal Schwannomas cases reported in the literature, including our case

First Author & Year	Age (Year)	Gender	Spinal Levels & Compartments Involved
Slooff <i>et al.</i> (1964) [9]	62	Female	Cervical
Kumar & Gulati (1977) [10]	24	Female	Cervical + (T7–9)
Pardatscher <i>et al.</i> (1979) [11]	41	Male	Thoracic (T2–T8)
Bhayani <i>et al.</i> (1996) [12]	15	Male	Cervical (C4–C5) and Lumbar (L5)
Bernal-García <i>et al.</i> (2010) [13]	35	Female	Thoracic (T1–T5)
Swiatek <i>et al.</i> (2021) [1]	53	Female	Cervical (C2–C3) and Lumbar (L2–L3)
Present Case (2026)	24	Female	Cervical, Thoracic, and Lumbar (Multilevel Intradural-Extramedullary with suspected Intramedullary component and Sacrococcygeal Dural Ectasia)

Second, the involvement of the lumbar spine was scarce, reported in only 10/165 cases (Table 2), and none of the reported cases showed sacrococcygeal region involvement with sacral dural ectasia, which is a complication of a massive concurrent extramedullary tumor burden. Males predominated in 7/10 of those specific historical periods. Crucially, none of the previously recorded cases showed involvement of the sacrococcygeal region, which was characterized as

sacrococcygeal dural ectasia. The latter is characterized by ballooning or widening of the dural sac, which is often seen in connective tissue diseases or in neurofibroma type 1. In the current case, there was a combined massive, multi-level intradural-extramedullary tumor burden that created a high-grade mechanical block within the spinal canal. The severely disrupting tumor burden may have blocked normal cerebrospinal fluid pathways.

Table 2: Describes demographic criteria of previously reported cases of spinal Schwannomas that involved Lumbar spines alongside their neurological assessment, including our case

Author (year)	Age (year)	Gender	Vertebrae Involved	Neurological Examination Findings
McCormick <i>et al.</i> (1964) [14]	62	Male	Lumbar (L2)	Normal
Schmitt (1975) [15]	68	Male	Lumbar (L1)	Sensory and motor deficits
Cantore <i>et al.</i> (1982) [16]	57	Male	Thoracic & Lumbar (T12–L1)	Normal
Jacquet <i>et al.</i> (1992) [17]	49	Male	Thoracic & Lumbar (T12–L1)	Normal
Hejazi & Hassler (1998) [18]	65	Male	Thoracic & Lumbar (T12–L1)	Sensory and motor deficits
O'Brien <i>et al.</i> (2003) [19]	48	Male	Thoracic & Lumbar (T11–L1)	Sensory and motor deficits
Kahilogullari <i>et al.</i> (2005) [20]	38	Female	Thoracic & Lumbar (T12–L2)	Sensory deficit
Hayashi <i>et al.</i> (2009) [21]	78	Male	Thoracic & Lumbar (T11–L1)	Motor and sensory deficits
Ohtonari <i>et al.</i> (2009) [22]	29	Female	Thoracic & Lumbar (T12–L1)	Motor deficit only
Singh <i>et al.</i> (2018) [2]	27	Female	Thoracic & Lumbar (T12–L2)	Sensory and motor deficits
Present Case (2026)	24	Female	Cervical, Thoracic, Lumbar, and Sacral regions	Axial spinal pain, sensory disturbances, radicular and axial symptoms (without rapid motor deterioration)

Over time, the chronic elevation of hydrostatic pressure associated with mechanical compression of the cauda equina caused a gradual expansion of the lower dural borders. The diagnosis of multilevel spinal schwannomas presents major obstacles. The associated symptoms include weakness, numbness, tingling, and loss of bladder or bowel control. All these manifestations are non-specific in nature and may share symptoms related to other spinal cord conditions such as transverse myelitis or multiple sclerosis [23]. The diagnostic modality of choice for spinal cord tumor detection is MRI. Nonetheless, there are certain limitations, such as the inability to definitively

distinguish between intradural extramedullary schwannomas and other nerve sheath tumors or alternative spinal canal neoplasms on imaging alone [24]. Additionally, neurofibromas and schwannomas may exhibit overlapping or hybrid histological characteristics, complicating definitive diagnosis based solely on biopsy findings [25]. To achieve an accurate diagnosis and effective management of multi-level spinal Schwannomatosis presentation, a multidisciplinary approach involving neurologists, neurosurgeons, radiologists, and pathologists is essential. While surgery remains the definitive treatment for symptomatic extra-

medullary masses [26], managing multi-level tumors represents a surgical challenge. Subtotal resection is often performed when severe adhesion to surrounding nerve roots exists to preserve baseline neurological function. Fortunately, Schwannomas carry a low rate of recurrence [27]. Adjuvant therapy is ineffective for benign intraspinal extramedullary schwannomas and therefore has no viable role in treatment [28]. Presently, there is ongoing research into advanced treatment modalities such as targeted drug delivery and nanomedicine methodologies [29]. Currently, our patient is kept under conservative management and monitoring, coupled with pain relief medication, since she declined surgery. For multi-level Schwannomatosis-NOS, this conservative approach offers a highly viable alternative by avoiding cumulative neurological morbidity, surgical risk, and progressive spinal instabilities linked with multi-stage spinal surgeries in the lack of rapid motor deterioration. Our case had declined advice for invasive medical investigation and intervention, which highlights a current clinical dilemma of how to foster patient autonomy and still deliver the best possible diagnostic and therapeutic services. One of the study limitations is the lack of histopathological confirmation, as the patient refused surgery and the diagnosis was made solely on highly suggestive radiological and clinical features. Second, the lack of cervical spine MRI limited the full extent of disease mapping since she already reported symptoms of radiating pain and numbness in both upper limbs; thus, the disease burden was not comprehensively evaluated.

Conclusion

The current case was a unique multilevel spinal SC with sacrococcygeal dural ectasia; it emphasizes the necessity of imaging the entire neuraxis to reach the precise diagnosis. For patients who decline surgery, personalized conservative treatment with clinical and radiological monitoring can be a feasible approach. Conservative management offers a more balanced way without infringing on the patient's right to choose and adds to an already scarce body of literature on the conservative management of spinal schwannomas.

Conflict of interests

The authors declared no conflict of interest.

Funding source

The authors did not receive any source of funds.

Data sharing statement

The data that supports the findings of this study are available from the corresponding author upon a reasonable request.

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