Cureus
Part of SPRINGER NATURE

Open Access Case Report

# An Extra-Adrenal Paraganglioma in the Lumbar Spine: A Case Report

Nikhil Reddy P [1], Balasubramanian Kannan [1], Jishnu Nair [2], Venkata Shashank Davuluri [1], Visvanathan Krishnaswamy [1]

1. Neurosurgery, Sri Ramachandra Institute of Higher Education and Research, Chennai, IND 2. Neurosurgery, Salford Royal Hospital, Northern Care Alliance NHS Foundation Trust, Salford, GBR

**Corresponding author:** Visvanathan Krishnaswamy, visvanathan.k@sriramachandra.edu.in

**Review began** 10/04/2026
**Review ended** 10/26/2026
**Published** 10/31/2026

**© Copyright** 2026
Reddy P et al. This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

**DOI:** 10.7759/cureus.118885

## Abstract

Spinal paragangliomas are rare, benign, slow-growing, highly vascular neuroendocrine tumours. We report a 51-year-old male with six months of progressive low back pain radiating to both lower limbs and no neurological deficit. Magnetic resonance imaging of the lumbosacral spine showed a well-circumscribed, ovoid intradural extramedullary lesion at the T12-L1 level measuring 8.1 × 9.2 × 35.0 mm that was hypointense on T1-weighted images, heterogeneously hyperintense on T2-weighted images, and enhanced avidly after gadolinium. The working diagnosis was schwannoma or ependymoma. The tumour, which arose from a single rootlet, was excised en bloc through a T12-L1 laminectomy. Histopathology showed nests of uniform cells in a Zellballen pattern within a rich capillary network with focal perivascular pseudorosettes, fewer than two mitoses per 10 high-power fields and no necrosis. The tumour cells were diffusely positive for synaptophysin and chromogranin and negative for glial fibrillary acidic protein and epithelial membrane antigen, with focal S-100 positivity confined to sustentacular cells and a Ki-67 index of 5%, establishing the diagnosis of a paraganglioma. The patient's pain resolved completely, and he remained free of deficit at six months. This case illustrates that ependymoma-like pseudorosettes do not exclude paragangliomas at this site, that immunohistochemistry is decisive, and that complete excision is curative without adjuvant therapy.

**Categories:** Neurosurgery, Endocrinology/Diabetes/Metabolism
**Keywords:** cauda equina neuroendocrine tumours (ce-nets), extra-adrenal paraganglioma, filum terminale, immunohistochemistry (ihc), intradural extramedullary tumour, synaptophysin

## Introduction

Paragangliomas are neuroendocrine neoplasms of neural crest origin that arise from the paraganglia of the autonomic nervous system. Tumours of the sympathetic paraganglia, including adrenal pheochromocytoma, usually secrete catecholamines, whereas those of parasympathetic paraganglia are typically non-functioning [1]. More than 90% of the latter occur in the head and neck as carotid body, vagal and jugulotympanic tumours [2]. The cauda equina and filum terminale are an uncommon site. The first such tumour was reported by Miller and Torack in 1970 as a "secretory ependymoma" [3], and its paraganglionic nature was recognised by Lerman et al. in 1972 [4]. Roughly 300 cases have since been described, and they are estimated to account for 3.4-3.8% of tumours of the cauda equina region [5,6]. Genome-wide methylation and sequencing of 57 cases have shown that cauda equina paragangliomas are epigenetically distinct from paragangliomas elsewhere, lack the succinate dehydrogenase (SDH) mutations that confer metastatic risk at other sites, frequently express cytokeratins, and arise sporadically [7]. The 2021 WHO classification of central nervous system tumours therefore lists them as a separate tumour type, cauda equina neuroendocrine tumour (previously cauda equina paraganglioma), corresponding to CNS WHO grade 1 [8], and the 2022 WHO classification of endocrine tumours has adopted the same term [1].

Patients present in the fourth to sixth decades, with a modest male predominance, with low back pain and often radicular pain; neurological deficit and catecholamine-related symptoms are uncommon [9,10]. Contrast-enhanced magnetic resonance imaging (MRI) is the investigation of choice but rarely allows a specific diagnosis, so most tumours are operated on as presumed ependymomas or schwannomas and diagnosed only on histopathology and immunohistochemistry [9,11,12]. On MRI, the tumour is a well-circumscribed, intensely enhancing intradural extramedullary mass, usually iso- or hypointense to the cord on T1-weighted and hyperintense on T2-weighted images; serpiginous flow voids, a haemosiderin 'cap sign' or a stalk-like attachment to the filum terminale, suggest the diagnosis but are inconstant, and in their absence, the lesion is indistinguishable from the far commoner myxopapillary ependymoma and schwannoma [9,12]. Histologically, it shows the nested 'Zellballen' architecture of paragangliomas elsewhere, but ependymoma-like perivascular pseudorosettes are frequent and can mislead, so the diagnosis rests on diffuse synaptophysin and chromogranin positivity, absence of glial fibrillary acidic protein (GFAP) and epithelial membrane antigen (EMA), and S-100 confined to sustentacular cells [5,11]. Complete excision is curative in the great majority of patients [6,9,10]. We report a tumour at the thoracolumbar junction that was radiologically indistinguishable from ependymoma and whose histology contained ependymoma-like perivascular pseudorosettes, and we discuss the features that secured the diagnosis.

## Case Presentation

A 51-year-old male presented with progressively worsening low back pain of six months' duration radiating to both lower limbs, which had not responded to analgesics. He denied limb weakness, sensory disturbance, or bowel or bladder dysfunction, and there was no history of palpitations, headache, sweating, or hypertension. There was no family history of endocrine or nervous system tumours. On examination, the blood pressure was 140/80 mmHg and the pulse 80/min; there was no facial flushing. Tone and power were normal in all four limbs, sensation was intact, deep tendon reflexes were symmetrical, plantar responses were flexor, and straight-leg raising was negative bilaterally.

MRI of the lumbosacral spine with whole-spine screening demonstrated a longitudinally oriented, ovoid, intradural extramedullary lesion at the T12-L1 level in the region of the conus medullaris, measuring 8.1 × 9.2 × 35 mm (anteroposterior × transverse × craniocaudal). It was hypointense on T1-weighted images, heterogeneously hyperintense on T2-weighted images (Figure *1A*). MR myelography showed an intradural filling defect (Figure *1B*), which enhanced avidly and homogeneously after gadolinium (Figure *1C*). There were no intratumoral flow voids, no peripheral hypointense rim, no associated cyst or syrinx, and no other lesion was seen in the spinal axis. The working diagnosis was schwannoma, with myxopapillary ependymoma as the principal alternative; meningioma, neurofibroma and metastasis were considered less likely. Plasma and urinary metanephrines were not measured, as the clinical picture gave no suspicion of a secretory tumour.

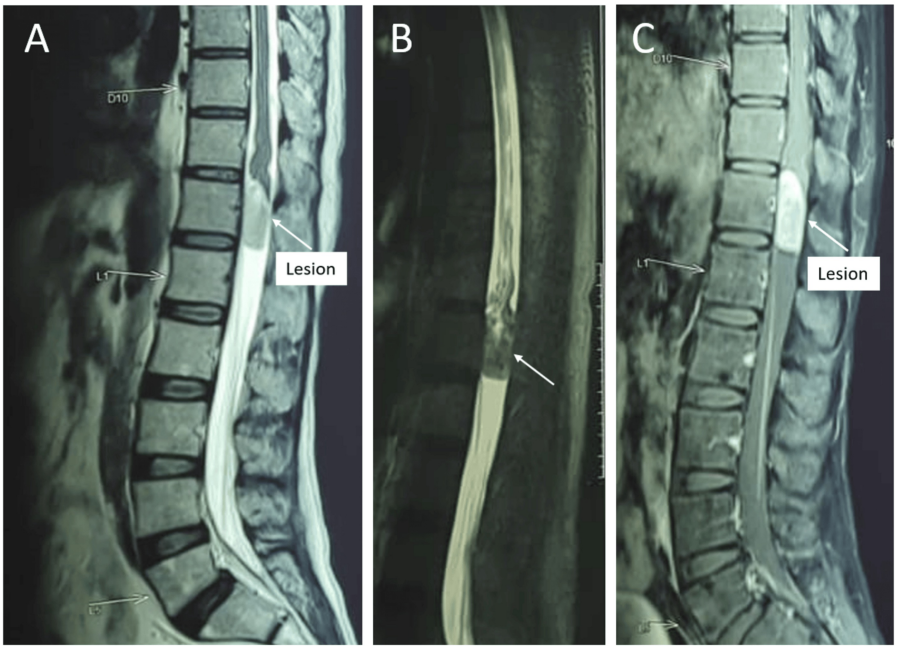



**FIGURE 1: Preoperative MRI. (A) Sagittal T2-weighted image showing a well-circumscribed intradural extramedullary lesion at T12–L1 (arrow), immediately below the conus medullaris, of a heterogeneous signal that is hyperintense to the cord and hypointense to cerebrospinal fluid. (B) Sagittal heavily T2-weighted MR myelographic image showing the corresponding intradural filling defect (arrow) at the conus level, with the cauda equina roots outlined below it. (C) Sagittal contrast-enhanced T1-weighted image showing intense, homogeneous enhancement of the lesion.**

Under general anaesthesia, the patient was positioned prone, and the level was confirmed with fluoroscopy. Through a midline incision and subperiosteal muscle dissection, a T12-L1 laminectomy was performed, and the dura was opened in the midline. A well-defined, reddish-brown, encapsulated and moderately vascular intradural extramedullary tumour lay immediately below the conus medullaris, arising from a single rootlet; the adjacent roots were displaced but not infiltrated (Figure *2*). The tumour was removed en bloc with its capsule intact, together with the short segment of the rootlet of origin, and all other roots were preserved. Intraoperative neuromonitoring was not used. Blood pressure remained stable throughout. After haemostasis, the dura was closed watertight, and the wound was closed in layers. Postoperatively, there was

no new neurological deficit.

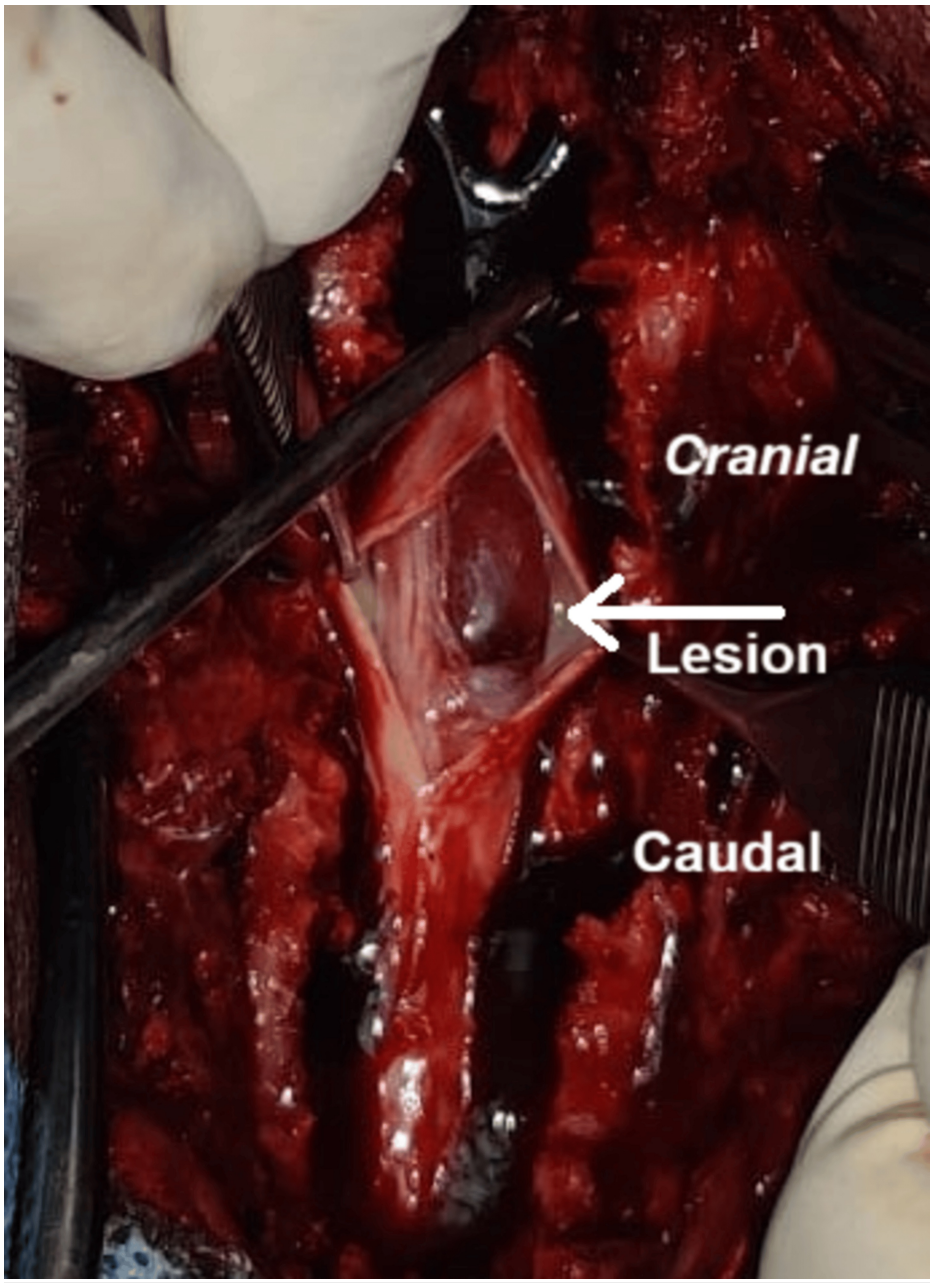



**FIGURE 2: Intraoperative photograph after T12–L1 laminectomy and midline durotomy showing the encapsulated, reddish-brown tumour (arrow) lying below the conus medullaris and displacing the adjacent cauda equina roots.**

The fixed specimen was an ovoid, smooth, encapsulated, grey-brown mass measuring 3 × 1.5 × 1.2 cm with a grey-white to grey-brown cut surface. Microscopy showed a well-circumscribed tumour composed of nests of uniform round-to-oval cells with finely stippled chromatin, inconspicuous nucleoli and finely granular eosinophilic cytoplasm, arranged in a Zellballen pattern within a delicate capillary network (Figure *3A*); mild nuclear pleomorphism and focal perivascular pseudorosettes were present. Mitoses numbered fewer than two per 10 high-power fields, and there was no necrosis or vascular invasion. On immunohistochemistry, the tumour cells were diffusely and strongly positive for synaptophysin and chromogranin (Figures *3B*, *3C*) and negative for GFAP and EMA; S-100 protein was focally positive, confined to the peripheral sustentacular cells (Figure *3D*). The Ki-67 labelling index was 5%. The final histopathological diagnosis was cauda equina neuroendocrine tumour (paraganglioma of the cauda equina), CNS WHO grade 1.

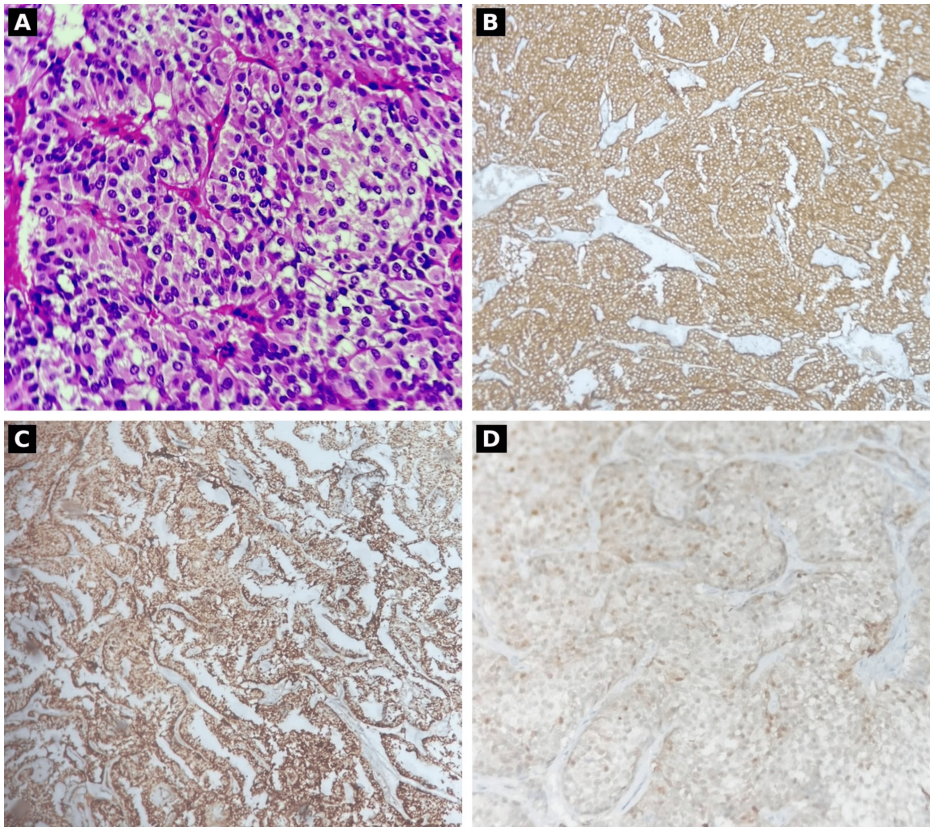



**FIGURE 3: Histopathology and immunohistochemistry. (A) Haematoxylin and eosin, ×400: nests of uniform round-to-oval cells with finely stippled chromatin (Zellballen) separated by a delicate capillary network. (B) Synaptophysin: diffuse strong cytoplasmic positivity. (C) Chromogranin: diffuse strong cytoplasmic positivity. (D) S-100: scattered positivity confined to sustentacular cells.**

He was mobilised on the first postoperative day and discharged on the third postoperative day. At one-month and six-month follow-up, the neurological examination was normal, and he had returned to full daily activity.

## Discussion

The clinical profile of this patient is typical of the entity. In a systematic review of 334 primary spinal paragangliomas, 84% lay in the cauda equina region, the median age was 46 years, and low back pain (64%) and radicular pain (54%) were the commonest complaints, while sympathetic symptoms occurred in only 5% [10]. A scoping review of 121 patients found a mean age of 49 years, a male-to-female ratio of 71:50, and back pain in 94% [9]. Symptom duration tends to be long and correlates with tumour size [13]; six months of pain before diagnosis, as here, is usual. The T12-L1 site is at the upper limit of the usual distribution: cauda equina tumours are mostly lumbar (49%) or lumbosacral (29%) and only 5% are thoracolumbar [10], which is why a conus-level lesion tends to be assumed to be a myxopapillary ependymoma.

Imaging did not point to the diagnosis, and it rarely does. Spinal paragangliomas are classically iso- or hypointense to the cord on T1-weighted images, hyperintense on T2-weighted images, and enhance intensely [12,14,15]; our lesion was T1-hypointense with heterogeneous T2 signal, a pattern that has been reported and probably reflects the tumour's vascularity and small foci of haemorrhage [12]. Features that should raise suspicion of paraganglioma are serpiginous flow voids giving a "salt-and-pepper" appearance, a peripheral hypointense haemosiderin rim ("cap sign"), an associated cyst or syrinx, and a stalk-like attachment to the filum terminale or a rootlet [9,12,14,15]. None of these was present in our patient, and because myxopapillary ependymoma is the commonest tumour at this level and shares every non-specific feature, the preoperative label in most reported series, as in ours, is ependymoma or schwannoma [9,11]. Preoperative recognition matters for two reasons: the tumour is vascular, and a small proportion of cauda equina tumours are functional and have caused intraoperative hypertensive crises [9]. The Endocrine Society recommends plasma free or urinary fractionated metanephrines whenever a paraganglioma is suspected

[16]. Our patient had no clinical features of catecholamine excess, the diagnosis was not suspected preoperatively, and metanephrines were not measured.

The histological finding that most complicated the diagnosis was the presence of perivascular pseudorosettes, traditionally regarded as a hallmark of ependymoma, together with mild nuclear pleomorphism and scattered mitoses. However, the two largest clinicopathological series of cauda equina paragangliomas, of 31 and 30 cases, respectively, both describe perivascular pseudorosettes and ependymoma-like areas within otherwise typical Zellballen-patterned tumours [5,17], and cauda equina tumours frequently show gangliocytic differentiation and cytokeratin expression that would be unusual for paragangliomas elsewhere [7]. Pseudorosettes therefore cannot be used to favour ependymoma, and the discrimination rests on the immunohistochemical panel: paraganglioma is diffusely positive for chromogranin and synaptophysin, negative for GFAP and EMA, and shows S-100 only in the peripheral sustentacular cells, as in our patient; ependymoma is GFAP-positive with dot-like EMA positivity; schwannoma is diffusely S-100-positive; and meningioma is EMA-positive [5,11,17]. In the series of Yang et al., synaptophysin and chromogranin were positive in every case, GFAP and EMA were negative in every case, S-100 was positive in 84%, and the Ki-67 index was below 10% in all cases [11]. Fiorini et al. reported a mean Ki-67 of 5.7% (range 1-10%) [14]. The Ki-67 index of 5% and the mitotic count of fewer than two per 10 high-power fields, with no necrosis or vascular invasion, in our patient are in keeping with the indolent behaviour expected of this tumour.

Cauda equina paragangliomas differ biologically from paragangliomas at other sites. Schweizer et al. showed that they have a unique DNA methylation profile, lack the chromosomal imbalances and SDH mutations found in abdominothoracic paragangliomas, occur without a familial syndrome, recur in about 9% of cases and do not metastasise outside the central nervous system [7]. This underlies their reclassification as a distinct tumour type, cauda equina neuroendocrine tumour, in the 2021 WHO classification of CNS tumours and the 2022 WHO classification of endocrine tumours [1,8]. Consequently, the SDHB-associated risk of metastatic disease that governs management of sympathetic paragangliomas [18] does not apply to this site, and germline testing is not routinely indicated in the absence of multifocal disease, secretory activity, or a suggestive family history, none of which was present here. Consistent with this, cauda equina tumours are almost always non-secretory [9,19], and our patient's blood pressure remained stable throughout tumour manipulation.

Gross total resection is the treatment of choice and is usually achievable because the tumour is encapsulated and attached by a stalk to the filum or, as in this case, to a single rootlet, which can be sacrificed with little or no deficit [6,9,14]. Reported recurrence rates are low, 3.9% in the pooled systematic review [10], 7% in the scoping review [9] and 9% in the molecular series [7], and recurrence is associated with subtotal removal or capsular breach; adjuvant radiotherapy is reserved for incompletely resected or histologically aggressive tumours [10]. Late recurrence and, exceptionally, cerebrospinal fluid dissemination with intracranial and intraspinal metastases have been reported [20], so periodic clinical and MRI surveillance is advisable even after complete excision, with the interval and duration left to the treating surgeon [9]. In our patient, the tumour was removed en bloc with the capsule intact, no adjuvant therapy was given, and he remains under follow-up.

This report has the limitations of a single case: follow-up is limited to six months, so late recurrence cannot be excluded; plasma and urinary metanephrines were not measured preoperatively; and intraoperative neuromonitoring was not used.

## Conclusions

Paraganglioma of the cauda equina, now termed a cauda equina neuroendocrine tumour, should be included in the differential diagnosis of any enhancing intradural extramedullary lesion at the conus or filum level, because MRI cannot reliably separate it from myxopapillary ependymoma or schwannoma and because its vascularity and rare secretory potential have operative implications. Perivascular pseudorosettes on histology do not exclude the diagnosis; a neuroendocrine immunohistochemical panel comprising synaptophysin, chromogranin, GFAP, EMA and S-100, with a Ki-67 index, is decisive. These are sporadic CNS WHO grade 1 tumours that are cured by complete en bloc excision in the great majority of patients without adjuvant therapy, although long-term surveillance remains prudent.

## Additional Information

### Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

**Concept and design:** Nikhil Reddy P, Balasubramanian Kannan, Venkata Shashank Davuluri, Visvanathan Krishnaswamy

**Acquisition, analysis, or interpretation of data:** Nikhil Reddy P, Balasubramanian Kannan, Venkata

Shashank Davuluri, Jishnu Nair, Visvanathan Krishnaswamy

**Drafting of the manuscript:** Nikhil Reddy P, Balasubramanian Kannan, Venkata Shashank Davuluri, Jishnu Nair

**Critical review of the manuscript for important intellectual content:** Nikhil Reddy P, Venkata Shashank Davuluri, Visvanathan Krishnaswamy

## Disclosures

**Human subjects:** Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

# References

1. Mete O, Asa SL, Gill AJ, Kimura N, de Krijger RR, Tischler A: Overview of the 2022 WHO classification of paragangliomas and pheochromocytomas. Endocr Pathol. 2022, 33:90-114. 10.1007/s12022-022-09704-6
2. Corinaldesi R, Novegno F, Giovenali P, Lunardi T, Floris R, Lunardi P: Paraganglioma of the cauda equina region. Spine J. 2015, 15:e1-8. 10.1016/j.spinee.2014.10.021
3. Miller CA, Torack RM: Secretory ependymoma of the filum terminale . Acta Neuropathol. 1970, 15:240-50. 10.1007/BF00686770
4. Lerman RI, Kaplan ES, Daman L: Ganglioneuroma-paraganglioma of the intradural filum terminale. Case report. J Neurosurg. 1972, 36:652-8. 10.3171/jns.1972.36.5.0652
5. Sonneland PR, Scheithauer BW, LeChago J, Crawford BG, Onofrio BM: Paraganglioma of the cauda equina region: clinicopathologic study of 31 cases with special reference to immunocytology and infrastructure. Cancer. 1986, 58:1720-35. 10.1002/1097-0142(19861015)58:8%3C1720::AID-CNCR2820580824%3E3.0.CO;2-U
6. Gelabert-González M: Paragangliomas of the lumbar region. Report of two cases and review of the literature. J Neurosurg Spine. 2005, 2:354-65. 10.3171/spi.2005.2.3.0354
7. Schweizer L, Thierfelder F, Thomas C, et al.: Molecular characterization of CNS paragangliomas identifies cauda equina paragangliomas as a distinct tumor entity. Acta Neuropathol. 2020, 140:893-906. 10.1007/s00401-020-02218-7
8. Louis DN, Perry A, Wesseling P, et al.: The 2021 WHO Classification of Tumors of the Central Nervous System: a summary. Neuro Oncol. 2021, 23:1231-51. 10.1093/neuonc/noab106
9. Shtaya A, Iorga R, Hettige S, Bridges LR, Stapleton S, Johnston FG: Paraganglioma of the cauda equina: a tertiary centre experience and scoping review of the current literature. Neurosurg Rev. 2022, 45:103-18. 10.1007/s10143-021-01565-7
10. Palmisciano P, Sagoo NS, Haider AS, et al.: Primary paraganglioma of the spine: a systematic review of clinical features and surgical management in cauda equina versus non-cauda equina lesions. World Neurosurg. 2022, 161:190-197.e20. 10.1016/j.wneu.2022.01.110
11. Yang C, Li G, Fang J, Wu L, Yang T, Deng X, Xu Y: Clinical characteristics and surgical outcomes of primary spinal paragangliomas. J Neurooncol. 2015, 122:539-47. 10.1007/s11060-015-1742-0
12. Sundgren P, Annertz M, Englund E, Strömblad LG, Holtås S: Paragangliomas of the spinal canal . Neuroradiology. 1999, 41:788-94. 10.1007/s002340050843
13. Aggarwal S, Deck JH, Kucharczyk W: Neuroendocrine tumor (paraganglioma) of the cauda equina: MR and pathologic findings. AJNR Am J Neuroradiol. 1993, 14:1003-7.
14. Fiorini F, Lavrador JP, Vergani F, et al.: Primary lumbar paraganglioma: clinical, radiologic, surgical, and histopathologic characteristics from a case series of 13 patients. World Neurosurg. 2020, 142:e66-72. 10.1016/j.wneu.2020.05.144
15. Abe H, Maeda M, Koshimoto Y, et al.: Paraganglioma of the cauda equina: MR findings . Radiat Med. 1999, 17:235-7.
16. Lenders JW, Duh QY, Eisenhofer G, et al.: Pheochromocytoma and paraganglioma: an Endocrine Society clinical practice guideline. J Clin Endocrinol Metab. 2014, 99:1915-42. 10.1210/jc.2014-1498
17. Moran CA, Rush W, Mena H: Primary spinal paragangliomas: a clinicopathological and immunohistochemical study of 30 cases. Histopathology. 1997, 31:167-73. 10.1046/j.1365-2559.1997.2300841.x
18. Dahia PL: Pheochromocytoma and paraganglioma pathogenesis: learning from genetic heterogeneity . Nat Rev Cancer. 2014, 14:108-19. 10.1038/nrc3648
19. Aghakhani N, George B, Parker F: Paraganglioma of the cauda equina region--report of two cases and review of the literature. Acta Neurochir (Wien). 1999, 141:81-7. 10.1007/s007010050269
20. Roche PH, Figarella-Branger D, Regis J, Peragut JC: Cauda equina paraganglioma with subsequent intracranial and intraspinal metastases. Acta Neurochir (Wien). 1996, 138:475-9. 10.1007/BF01420312