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An Unusual Giant Neuroendocrine Tumor of the Cauda Equina: A Case Report and Review of the Literature

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Abstract

Cauda equina neuroendocrine tumors (CENETs), historically termed cauda equina paragangliomas, are exceptionally rare neuroendocrine neoplasms arising in the cauda equina or filum terminale region. Although their cellular origin remains uncertain, molecular and epigenetic studies demonstrate that CENETs are biologically distinct from conventional paragangliomas. Histologically, they typically exhibit a characteristic zellballen architecture with neuroendocrine differentiation, including expression of synaptophysin, chromogranin A, INSM1, and CD56. We report a giant CENET with extensive calcification and an unusual immunohistochemical finding. This case highlights the diagnostic and surgical challenges posed by giant CENETs. Its extensive calcification, vascularity, and unusual calcitonin expression expand the recognized clinical and immunophenotypic spectrum of this rare entity.

Categories: Pathology, Oncology, Neurosurgery
Keywords: calcitonin, cauda equina, neuroendocrine tumor, paraganglioma, spine

Introduction

The cauda equina consists of a bundle of numerous nerve roots that project distally and innervate both sensory and motor targets at the lumbar, sacral, and coccygeal spinal cord levels. Each nerve exits at its respective vertebral level toward targets supplied by the L2-S5 spinal cord level [1].

Similar to the rest of the spinal cord, the cauda equina is enveloped in the dura, arachnoid, and pia mater [1]. Therefore, tumors in this zone can produce severe Numeric Rating Scale (NRS) (10/10) low back pain [2], sciatica (often bilateral), saddle and/or genital sensory disturbance, and bladder, bowel, and sexual dysfunction [3].

Among intradural neoplasms of the cauda equina, ependymoma and schwannoma are among the most common. However, other neoplasms can be located at this specific location of the spinal cord, some of which are exceptionally rare, such as cauda equina neuroendocrine tumor (CENET).

Historically termed cauda equina paraganglioma, it is currently recognized as cauda equina neuroendocrine tumor [4]. The 2021 World Health Organization (WHO) classification recognizes that these tumors exhibit immunohistochemical and DNA methylation characteristics that distinguish them from paragangliomas in other locations. CENET is a neuroendocrine neoplasm arising from specialized neural crest cells in the cauda equina/filum terminale region [4-6].

Neuroendocrine tumors (NETs) constitute a heterogeneous group of neoplasms originating from neuroendocrine enterochromaffin cells of the gut and bronchi, which produce monoamines, peptides, and other biologically active substances widely distributed throughout the body [6].

The incidence of NETs has progressively increased over recent decades, rising from 1.09 cases per 100,000 inhabitants in 1975 to 6.98 cases per 100,000 in 2012 in the United States. Advances in diagnostic techniques and demographic changes in the population are believed to contribute [7].

Cauda equina neuroendocrine tumors (CENETs) were initially described as cauda equina paragangliomas due to their histomorphological and immunohistochemical characteristics, specifically their nested architecture (or zellballen pattern) and the expression of neuroendocrine markers such as chromogranin and synaptophysin [5,8].

Cauda equina neuroendocrine tumors (CENETs) are rare neoplasms; consequently, data on their epidemiology are derived primarily from small case series and case reviews. Published literature indicates a predominance in adults, most frequently during the fourth and fifth decades of life, and a slight male predominance. These tumors are located mainly in the lumbar and lumbosacral regions and account for a small proportion of intradural spinal neoplasms [9]. These can cause motor deficits due to spinal cord compression, as assessed using the Medical Research Council (MRC) scale [10].

A systematic review of 143 studies involving 334 patients with spinal paragangliomas found that 84.1% of the cases involved cauda equina lesions; among these, the lumbar and lumbosacral regions were the most common sites (49.1% and 29%, respectively). The median age of patients was 46 years (range: 6-85 years), and these tumors have been classified as WHO grade 1 [5,7-9,11].

Histologically, CENETs are characterized by a zellballen architecture, consisting of nests of chief cells surrounded by a fibrovascular stroma and sustentacular cells. The tumor cells are typically polygonal, with round or oval nuclei and finely granular "salt-and-pepper" chromatin. Immunohistochemically, they stain positive for synaptophysin, chromogranin A, INSM1, and CD56, while S100 and SOX10 can highlight the sustentacular cells. Variable cytokeratin expression and their immunophenotypic profile allow for their differentiation from other cauda equina neoplasms, particularly ependymomas and schwannomas [8,11].

The objective of this study is to review the existing literature and present a rare case of a giant CENET, which, in addition to its size, exhibited specific and unusual immunohistochemical markers, such as calcitonin, not previously described in this type of tumor; this makes the case noteworthy, justifying its presentation and publication.

Case Presentation

Case report

A 53-year-old woman presented with a five-year history of progressively worsening radicular pain in the sacral region and lower extremities, with a maximum intensity of NRS 5/10, controlled with nonsteroidal anti-inflammatory drugs (NSAIDs), associated with bilateral paresthesias in the legs and feet.

The symptoms were initially sporadic but increased over time. Until in April 2026, she experienced sudden, electric shock-like pain with an intensity of NRS 10/10, radiating to the thighs, gastrocnemius muscles, and tips of the toes; paraparesis, saddle anesthesia, urinary incontinence, and constipation were also present, resulting in multiple falls from a standing height.

Analgesic management with opioids was initiated, and complementary studies were performed, revealing an intraspinal, intradural tumor lesion involving the cauda equina and conus medullaris, corresponding to the T12-L2 vertebral levels, with an increase in the diameter of the affected vertebral canal (Figure *1*).

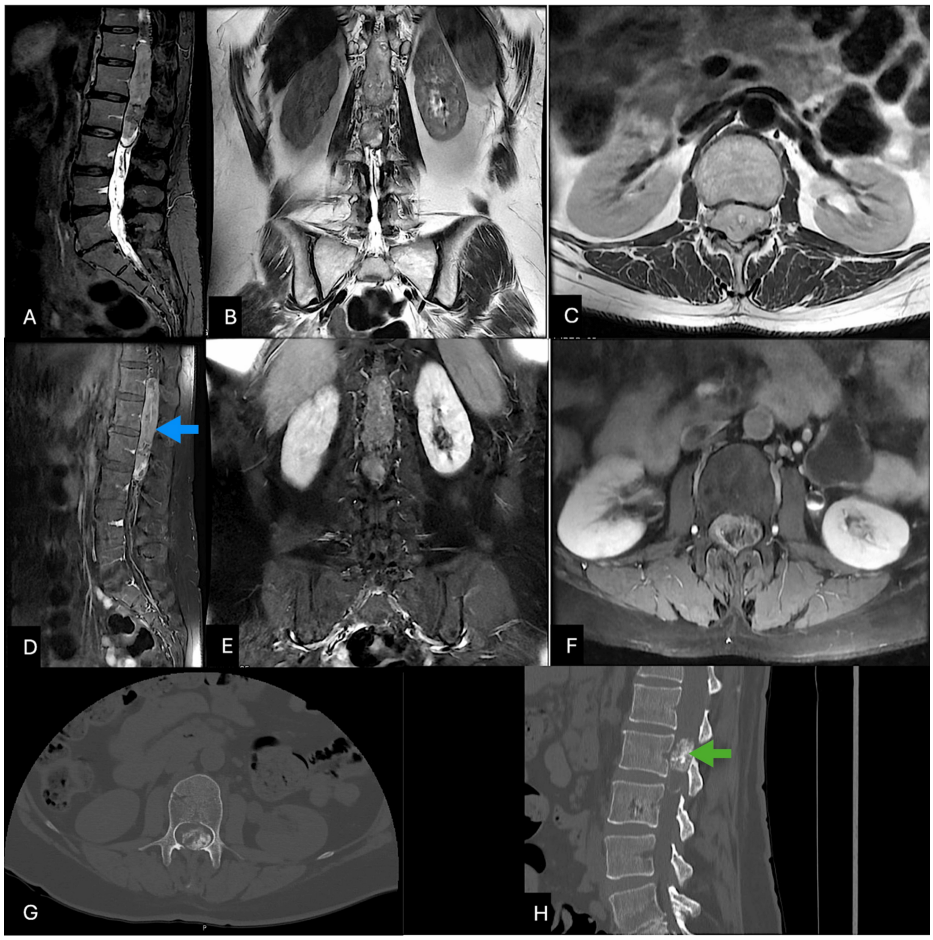


FIGURE 1: Preoperative MRI (sagittal, coronal, and axial views) showing an intradural extramedullary intraspinal lesion spanning the T12-L2 thoracolumbar segment

A-C (sagittal, coronal, and axial STIR-weighted views): The lesion is heterogeneous, diffuse, and ill-defined, occupying the entire spinal canal and causing widening of the canal and neuroforamina at these levels, with approximate dimensions of 103 × 18 × 28 mm along its craniocaudal, dorsoventral, and laterolateral axes; no arachnoid interface is visible. D-F (sagittal, coronal, and axial T1-weighted views with gadolinium (blue arrow)): The lesion exhibits heterogeneous internal enhancement following gadolinium administration. G and H: Axial and sagittal simple CT scans depicting a hyperdense image that indicates a solid calcified tumor (green arrow).

CT: computed tomography, MRI: magnetic resonance imaging, STIR: short tau inversion recovery

Surgical procedure

A posterior midline thoracolumbar approach was performed, along with a T12-L3 laminectomy and resection of the tumor, which enveloped the entirety of the cauda equina, partially infiltrated the tip of the conus medullaris, and displaced it cranially. The filum terminale was not visualized at any point, and the lesion presented abundant tortuous peritumoral and intratumoral vessels, leading to continuous and profuse oozing and bleeding. The surgery was conducted using intraoperative neurophysiological monitoring.

During the resection, motor evoked potentials dropped to an isoelectric line; therefore, the decision was made to halt the procedure at that moment, achieve hemostasis, and close in a single plane, as the dura mater was so thinned that it sustained multiple tears. Closure was achieved with primary closure using a synthetic dural substitute, and full-thickness sutures were applied with a drain directed toward the surgical bed.

Postoperative evolution

The patient progressed favorably, maintaining 3/5 bilateral MRC strength and controlling neuropathic pain of NRS 3/10. Postoperative and follow-up studies showed a residual lesion with a small, contained cerebrospinal fluid (CSF) fistula (Figure *2*).

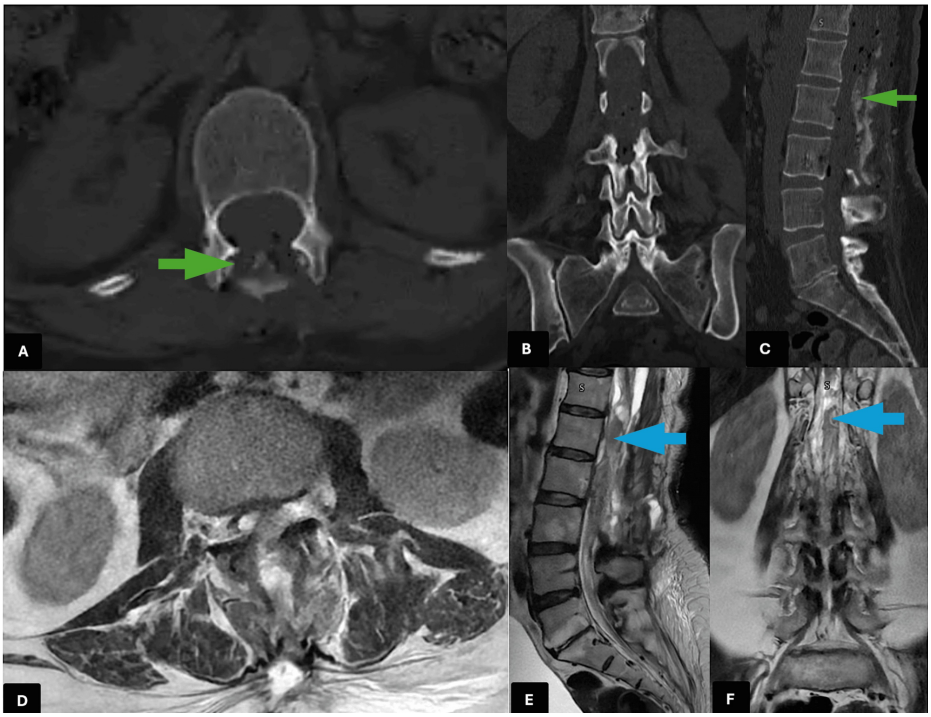


FIGURE 2: Postoperative CT and MRI scans (sagittal, coronal, and axial views) showing an intraspinal, extramedullary lesion spanning the T12-L2 thoracolumbar segments

A-C: Axial, coronal, and sagittal CT postoperative scans showing T12-L3 laminectomy (green arrow) and postoperative changes in intra- and extraspinal regions after removal of a thoracolumbar spinal lesion. D-F: Axial, coronal, and sagittal STIR-weighted postoperative scan depicting postoperative changes with an estimated 40% residual lesion (blue arrow) at the very apex of the original tumor, secondary to a firmly attached portion to the cauda equina roots.

CT: computed tomography, MRI: magnetic resonance imaging, STIR: short tau inversion recovery

The tissue obtained from the 60% cytoreduction was sent to pathology, where immunohistochemical studies were performed, yielding a final diagnosis of neuroendocrine tumor of the cauda equina (previously known as paraganglioma of the cauda equina) (Figure *3*).

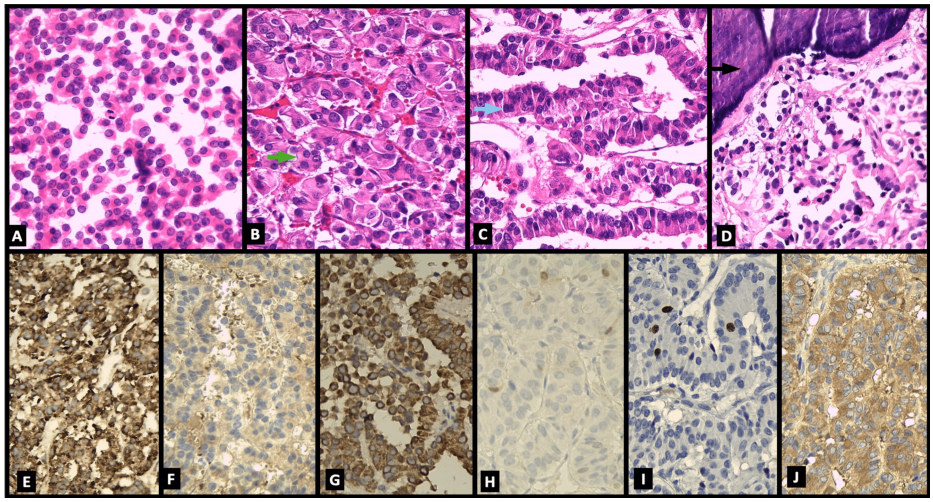


FIGURE 3: Immunohistochemical and histopathological findings

A: Intraoperative smear showed plasmacytoid cells with eosinophilic cytoplasm and eccentric round nuclei (H&E, 40×). B-D: H&E (40×) sections demonstrated the same cells disposed in nests (zellballen) (green arrow) and ribbon patterns (blue arrow) focally, with some mature bone spicules (black arrow), as well as the presence of sparse mitoses and necrosis. E-H: The tumor cells were positive for chromogranin A, neuron-specific enolase, cytokeratin cocktail, and S100 protein staining observed in the nucleus and cytoplasm of some of the sustentacular cells (40×), consistent with a neuroendocrine tumor. I and J: The proliferative index is low, around 3%, as assessed by visual estimation; calcitonin was also very strongly positive (40×).

H&E: hematoxylin and eosin

Following a positive result for calcitonin on immunohistochemistry, a protocol was implemented to rule out metastases; this included a hormonal profile, an ultrasound of the thyroid and parathyroid glands, and a thoraco-abdominopelvic computed tomography (CT) scan. These studies showed no evidence of secondary lesions (Figure *4*).

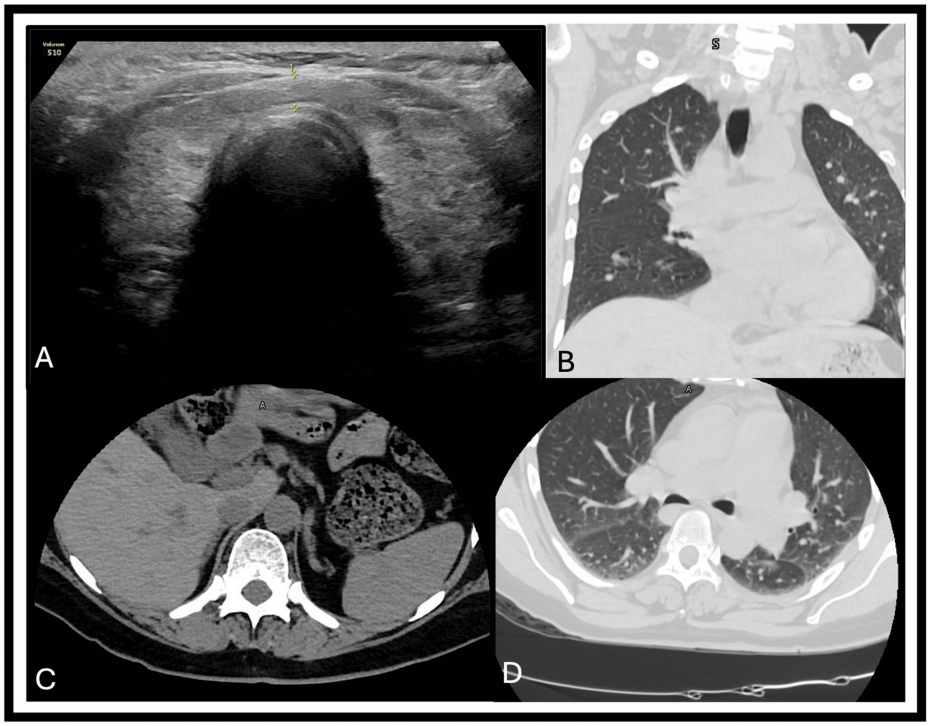


FIGURE 4: Systemic workup to exclude an extraspinal neuroendocrine primary tumor

Imaging studies of the (A) thyroid, (B and D) lungs, and (C) pancreas showed no evidence of an identifiable metastatic disease from an extraspinal primary site.

Taken together with the histopathological and immunohistochemical findings, these results support a primary spinal origin of the neoplasm despite the elevated calcitonin expression.

At the six-month follow-up, the patient continues to show favorable progress and is undergoing physical rehabilitation therapy; imaging studies show no changes compared to the previous examination.

Discussion

Cauda equina neuroendocrine tumors (CENETs) are exceptionally rare neoplasms, and their presentation as a giant, extensively calcified lesion is a particularly unusual characteristic [12].

In the present case, this tumor showed all expected immunohistochemical markers, in addition to presenting an unexpected positive calcitonin expression, which represents a distinctive histopathological and immunohistochemical finding [5,11].

Given the unusual calcitonin expression observed in the tumor, a systematic evaluation for a metastatic disease from an extraspinal primary site was performed. Consistent with the literature, the use of new tools is recommended to identify genetic predisposition syndromes; furthermore, molecular immunohistochemistry can be useful for identifying associations with other pathologies and detecting precursor lesions [13]. Thyroid, lung, and pancreatic imaging revealed no evidence of an identifiable primary lesion or metastatic disease, supporting the diagnosis of a primary neuroendocrine tumor of the cauda equina.

Due to the rarity of CENETs worldwide, with only a limited number of cases reported, and the combination of its particular characteristics, this makes this case particularly noteworthy and supports its inclusion in the international literature [4,7,11].

Conclusions

Cauda equina neuroendocrine tumors (CENETs) are extremely rare and are predominantly of a non-established cellular origin. The specific neuroendocrine, histopathological, and immunohistochemical markers of CENETs distinguish them from the classification established by the World Health Organization in 2021.

CENETs usually tend to have various immunohistochemical markers. Although calcitonin is not the most common of these, its detection should be considered to rule out other associated conditions. Through this clinical case, we highlight the importance of identifying such markers to ensure an accurate diagnosis. Furthermore, given that some of these markers may also be present in metastatic disease, additional diagnostic studies, if not already performed, are necessary; these may lead to adjustments in treatment, follow-up, and prognosis.

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: Zulma Cuevas Beltran, Jorge Luis Olivares Peña

Acquisition, analysis, or interpretation of data: Zulma Cuevas Beltran, Hector Lara, Jorge Luis Olivares Peña, Jesus Cazadero, Jorge Olivares Camacho

Drafting of the manuscript: Zulma Cuevas Beltran, Hector Lara, Jorge Luis Olivares Peña, Jesus Cazadero

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Supervision: Zulma Cuevas Beltran, Hector Lara, Jorge Luis Olivares Peña, Jesus Cazadero

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.

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