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Cauda Equina Neuroendocrine Tumor: Report of Two Cases and Review of Literature

Cui XD[1], Song LL[2], Zhang W[3], Xie WG[4*] and Gao YJ[5*]

[1]Department of Spinal Surgery, North Medical District of Linyi People's Hospital, Linyi, Shandong 276005, P.R. China

[2]Department of Spinal Surgery, North Medical District of v, Linyi, Shandong 276005, P.R. China

[3]Department of Spinal Surgery, North Medical District of Linyi People's Hospital, Linyi, Shandong 276005, P.R. China

[4]Department of Spinal Surgery, North Medical District of Linyi People's Hospital, Linyi, Shandong 276005, P.R. China

[5]Department of Spinal Surgery, North Medical District of Linyi People's Hospital, Linyi, Shandong 276005, P.R. China

[*]Corresponding author:

Wen-Gui Xie,
Department of Spinal Surgery, North Medical District of Linyi People's Hospital, Linyi, Shandong 276005, P.R. China,
E-mail: wenguixiely@163.com.

Ying-Ji Gao,
Department of Spinal Surgery, North Medical District of Linyi People's Hospital, Linyi, Shandong 276005, P.R. China,
E-mail: yingjigaoly@163.com.

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1. Abstract

Cauda equina Neuroendocrine Tumor (NET), previous known as cauda equina paragangliomas, is uncommon neoplasm in adults. In this study, we retrospectively reviewed two cases of cauda equina NET patients who had underwent surgical treatment in our department. We reviewed patient age, gender, presenting symptom, tumor radiological feature, surgical procedure, pathologic data and postoperative complications. Two cases were all clinically non-functional cauda equina NETs without sympathetic symptom. While image-based differential diagnosis of cauda equina tumors is still difficult and definite diagnosis depends on pathology and immunohistochemical staining, resecting the tumor with minimally invasive technique has good curative effect.

2. Introduction

Paragangliomas are rare Neuroendocrine Tumors (NETs), derived from the adrenal medulla (specifically named pheochromocytomas) or extra-adrenal paraganglia [1, 2]. They have close association with components of the sympathetic and parasympathetic nervous systems, therefore, they can arise in any location where paraganglia normally reside in adult tissues or during embryonic development [3]. About 90% of paragangliomas arise within the carotid and jugular bodies, while only 10% are associated with the spinal conus medullaris or cauda equina [4, 5]. Cauda equina neuroendocrine tumor, previous known as cauda equina paragangliomas, are frequently occurs in the middle age 40-50 years old, male is predominance [6]. However, the non-specific clinical features and radiological findings make the diagnosis difficult and misdiagnose frequent and the final diagnosis require pathological examination. There is very little research on cauda equina NET and thus limited experience in the diagnosis and treatment. Here, we present two cases of cauda equina paragangliomas and described the diagnostic and treatment strategy, and correlate our findings with a brief review of literature.

3. Case Report

3.1. Case 1

A 56-year-old man was admitted to the hospital with lower back pain radiating to the lower extremity of 3 years' duration. The pain had progressed to both posterior region of the thighs, as well as the posterolateral region of lower legs and foot for 3 months. Physical examination revealed no motor weakness but sensation was decreased over the both L5 and S1 dermatomes. Magnetic Reso-

nance Imaging (MRI) was performed and revealed a well circumscribed 1.5 cm diameter intradural mass-like lesion at the L3 vertebral body level and splaying the cauda equina. The mass showed iso-intensity signal in T1WI and uneven slight-high-intensity signal in T2WI, with heterogeneous enhancement in T1-weighted enhancement imaging (Figure 1).

A posterior approach L2, L3 and L4 laminectomies were performed, followed by total tumor resection using microsurgical technique. Postoperative pathology was consistent with paraganglioma, and immunohistochemical examination revealed Ki67-M1B1 was below 1%. The patient improved after surgery and post-operative MRI showed no tumor residual.

3.2. Case2

A 43-year-old female presented with back pain and right lower leg radiating pain from right buttock to posterior thigh for 20 days. There was no neurological deficit in physical examination. Her right straight-leg raising test was positive. Both her lower limbs sensation and muscle strength were normally. CT examination showed the soft tissue without calcification in the spinal canal at the level of L3. MRI was performed and revealed a round shaped intradural lesion with clear boundary extend along the whole L4 vertebral body level measuring 1.1*1.3*1.6 cm. The mass showed isointensity signal in T1WI and slight-high-intensity signal in T2WI, and markedly inhomogeneous enhancement after gadolinium infusion (Figure 2).

Complete L4 and partial L3 and L5 laminectomy was performed. Openning the dura by a midline incision revealed a lobulated dark red lesion, well circumscribed, both cranial and caudal tumor were supplied with abundant blood vessels. We separated the tumor, coagulated the vessels, and complete resect the tumor. The histopathology of the tumor was diagnosed as cauda equina paragangliomas, and immunohistochemical examination revealed CK (-) , EMA (-), Vimentin (+), S-100 (+), GFAP (-), CgA (+), Syn (+), CD56 (+), Ki67-MIB1 (<5%_. Postoperatively, symptoms were well relieved at 2 years' follow-up.

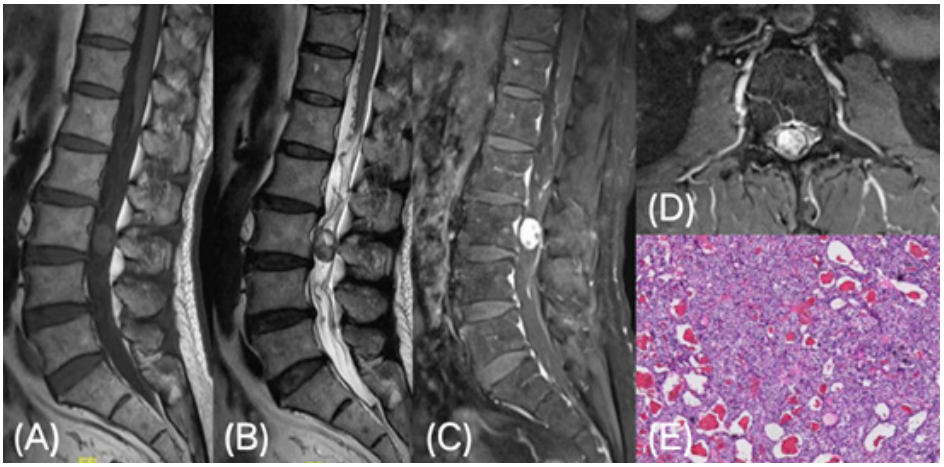

Figure 1: Case 1 (A) Sagittal T1-weighted image of the lumbar spine demonstrating the round-shaped, isointensity mass at the L3 vertebral body level; (B) Sagittal T2-weighted image demonstrating an intradural extramedullary tumor, with uneven slight-high-intensity signal and intratumoral cyst; (C, D) Sagittal and axial contrast-enhancement image demonstrating inhomogeneous enhancement of the tumor and "salt-and-pepper" appearance; (E) Pathologic diagnosis was consistant with paraganglioma, with well-formed perivascular pseudorosettes.

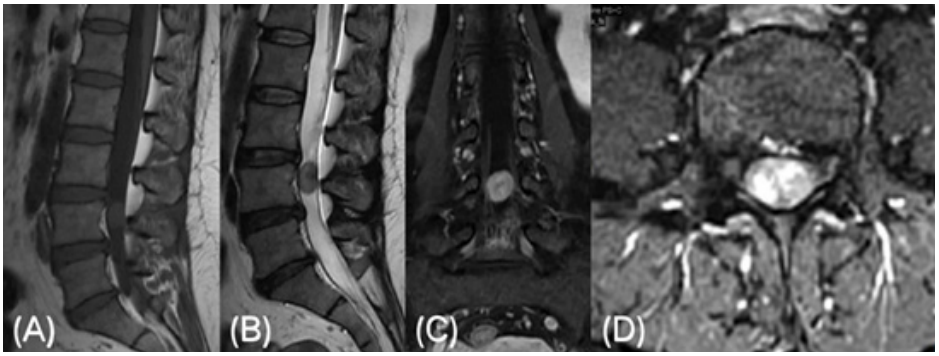

Figure 2: Case 2 (A) Sagittal T1-weighted image revealed a round shaped intradural lesion with clear boundary extend along the whole L4 vertebral body level measuring 1.1*1.3*1.6 cm; (B) Sagittal T2-weighted image showed slight-high-intensity signal; (C, D) Sagittal and axial contrast-enhancement image demonstrating inhomogeneous enhancement after gadolinium infusion.

4. Discussion and Conclusion

Cauda equina neuroendocrine tumors are very rare, account for about 3-4% of cauda equina tumors [7]. Since the morphologically, histogenetically, and molecularly was different from traditional extra-spinal paragangliomas, according to 2022 WHO classification of neuroendocrine neoplasms, previously known as cauda equina paraganglioma has been reclassified as cauda equina Neuroendocrine Tumor (NET). In the present study, we report two cases of cauda equina NETs and emphasize to display the clinical presentation, radiological images, operative findings and pathological results.

The clinical presentation of cauda equina NET is nonspecific. Though paraganglioma has the potential to secrete catecholamines, most of these tumors may appear to be clinically non-functional [3, 8, 9]. conducted a systematic review of primary spine paragangliomas,334 patients from 143 studies were included, cauda equina paragangliomas were account for 81.4%, only 5.4% were functional primary spine paragangliomas [10]. The most common complaint of cauda equina NETs were back pain, sciatica, and seldom accompany with acute-onset paraplegia. In our cases, back pain and lumbar sciatica were the main symptoms, and none symptoms associated with the catecholamine excess. However, even so, catecholamine related symptoms (like unstable blood pressure, cardiac dysfunction, and acute vascular catastrophes) should not take lightly, if functional, the management requires a multidisciplinary approach to optimize perioperative blood pressure and carefully management the tumor masses [3, 11].

Magnetic Resonance (MR) imaging plays a central role in the imaging of cauda equina NETs. They usually present as inhomogeneous, isointense with spinal cord on T1WI and hyperintense on T2WI, with heterogeneous gadolinium contrast enhancement [12]. Myxopapillary ependymomas, schwannomas, meningiomas or hemangioblastomas are the differential diagnosis of cauda equina NET [10]. Although cauda equina NETs have well-encapsulation and hypervascular characteristics, overlap in their imaging findings make the differential diagnosis difficult [12]. The hypervascular nature of cauda equina NET is frequently mentioned distinctive nature, the punctate flow void region in the matrix with increased signal intensity caused by slow flow and tumor cells made the tumors present with “salt-and-pepper” appearance on T2WI [7, 13-15]. However, small tumors tend not to show the flow void and hypervascularity phenomenon, and definition diagnosis is confirmed by surgical operation and pathology [12, 16].

Since the 4th edition of the WHO, paragangliomas have no longer been classified as benign and malignant, as any lesion can have metastatic potential and there are no clear-cut features that can predict metastatic behavior [17]. Factors such as range and dimension of pathology, location, relationship with the neighboring structures of the lesion and mutation status were more important to predict the prognosis of patients [18]. Moreover, the 2022 WHO classification of neuroendocrine neoplasms emphasis Ki67 labeling index as prognostic and predictive tool for classification and an integral part of the diagnosis [1]. Ki67 is a well-known proliferation marker for evaluating cell proliferation. All our two cases showed the Ki67-M1B1 was below 5%, which may point out the tumor with good prognosis and long progression-free survival [1, 19].

Gross-total resection is the first choice of treatment for cauda equina NET and the prognosis is good. However, intraoperative catecholamine release, tumor bleeding, aggressive bone erosion would increase surgical risk and postoperative recurrent. Therefore, some authors recommend presurgical and anesthetic management protocols, preoperative tumor embolization, microsurgical technique and postoperative adjuvant radiotherapy in certain patients (aggressive lesions or unrespectable tumor portions) to increase the security of the operation and reduce complications of the operation [10]. In view of this, dependable diagnosis before surgical operation would increase safety of operation and decrease operative complications. In our cases, preoperative diagnosis was missed as schwannoma or meningioma, although no symptoms and signs related to cathecholamine secretion during and after operation, we also emphasis to include the cauda equina NET as the potential diagnosis list with the aid of the MRI.

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