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Rare spinal tumor: a case of peripheral intradural primitive neuroectodermal tumor

Tumor espinal poco frecuente: un caso de tumor neuroectodérmico primitivo intradural periférico

Antuan Lacayo-Simón,* Ana Luisa Galicia-Zamalloa,‡ Diego Ivan Díaz-Haaz,§ David Campos-Flores¶

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Palabras clave:
tumor espinal, sarcoma de Ewing, tumores neuroectodérmicos, dolor lumbar, resección quirúrgica.

ABSTRACT

Peripheral primitive neuroectodermal tumors (pPNETs) are rare and aggressive neoplasms that originate from neural crest cells. Their presentation in the spine, especially intradural and extramedullary, is extremely rare and poses a diagnostic challenge due to their clinical and radiological similarity to other spinal lesions. This report presents the case of a 19-year-old male, who presented with progressive low back pain and weakness in the lower extremities, as well as bilateral paresthesias from L2 to S1, bilateral hyporeflexia at L4 and complete areflexia at S1, and positive signs of Lasègue, Bragard and Babinski. Simple and contrasted magnetic resonance imaging (MRI) of the lumbar spine revealed an abnormal mass in the cauda equina. Surgical treatment consisted of total resection of the mass by posterior approach, decompression and fixation from L2 to S1, guided with intraoperative neurological monitoring. The round tumor lesion had irregular borders, a blackish and gritty appearance, and measured approximately 3 × 4 cm. The lesion was intradural and centrally located, compressing the cauda equina between L2 and L5. Histopathological analysis confirmed a peripheral primitive neuroectodermal tumor with immunohistochemical positivity for CD99 and Ki-67 at 40%, and negativity for epithelial membrane antigen (EMA) and glial fibrillary acidic protein (GFAP). The patient received complementary treatment with three-dimensional conformal radiotherapy. In the immediate postoperative period he showed clinical improvement, with partial recovery of muscle strength. However, these tumors have a poor prognosis, with a median survival of 12 to 18 months, and require medium and long-term follow-up to evaluate response and sequelae. This case underlines the importance of pPNET in the differential diagnosis of spinal tumors and the need for immediate multimodal treatment.

RESUMEN

Los tumores neuroectodérmicos primitivos periféricos (pPNETs) son neoplasias poco frecuentes y agresivas que se originan a partir de células de la cresta neural. Su presentación en la columna vertebral, en especial en forma intradural y extramedular, es extremadamente rara y plantea un desafío diagnóstico debido a su similitud clínica y radiológica con otras lesiones espinales. En este informe se presenta el caso de un varón de 19 años, que acudió con dolor lumbar progresivo y debilidad en las extremidades inferiores, además, presentaba parestesias bilaterales de L2 a S1, hiporreflexia bilateral en L4 y arreflexia completa en S1, así como signos positivos de Lasègue, Bragard y Babinski. La resonancia magnética (RM) simple y contrastada de la columna lumbar reveló una masa anormal

* Orthopedist and Spine Surgeon. Hospital Ángeles Puebla, Puebla, Mexico. ORCID: 0009-0002- 2169-9183
‡ Department of Orthopedic and Traumatology, Hospital de Especialidades Instituto de Seguridad y Servicios Sociales de los Trabajadores al Servicio de los Poderes del Estado de Puebla (ISSSTEP). Puebla, Mexico. ORCID: 0000-0002-8120-6232
§ Faculty of Human Medicine "Dr. Manuel Velasco Suárez" Campus II, Universidad Autónoma de Chiapas. Chiapas, Mexico. ORCID: 0009-0003-0597-4795
¶ Department of Orthopedic and Traumatology, Hospital Regional ISSSTE Puebla. Puebla, Mexico. ORCID: 0000-0002-4328-9217

Correspondence:
David Campos Flores
E-mail: campos.tyo@gmail.com

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en la cauda equina. El tratamiento quirúrgico consistió en una resección total de la masa mediante abordaje posterior, descompresión y fijación de L2 a S1, guiado con monitorización neurológica intraoperatoria. La lesión tumoral redonda era de bordes irregulares, aspecto negruzco y arenoso, tenía un diámetro aproximado de 3 × 4 cm. La lesión era intradural y estaba situada en posición central, comprimiendo la cauda equina entre L2 y L5. El análisis histopatológico confirmó un tumor neuroectodérmico primitivo periférico con positividad inmunohistoquímica para CD99 y Ki-67 al 40%, y negatividad para el antígeno de membrana epitelial (EMA) y la proteína ácida fibrilar glial (GFAP). El paciente recibió tratamiento complementario con radioterapia conformacional tridimensional. En el postoperatorio inmediato mostró mejoría clínica, con recuperación parcial de la fuerza muscular. No obstante, estos tumores tienen un mal pronóstico, con una media de supervivencia de 12 a 18 meses, y requieren seguimiento a mediano y largo plazo para evaluar respuesta y secuelas. Este caso subraya la importancia del pPNET en el diagnóstico diferencial de los tumores espinales y la necesidad de un tratamiento multimodal inmediata.

Abbreviation:

AP = Anterior posterior
CNS = Central nervous system
cPNETs = Central primitive neuroectodermal tumors
EMA = Epithelial membrane antigen
EMG = Electromyography
GFAP = Glial fibrillary acidic protein
MRI = Magnetic resonance imaging
pPNETs = Peripheral primitive neuroectodermal tumors
SSEPs = somatosensory evoked potentials
VAS = Visual analogue scale

INTRODUCTION

Primitive neuroectodermal tumors (PNETs) are grouped into two categories based on their anatomical location: central primitive neuroectodermal tumors (cPNETs) and peripheral primitive neuroectodermal tumors (pPNETs).[1] Although they share a similar histology, cPNETs develop in the central nervous system (CNS), while pPNETs occur outside the CNS. pPNETs are remarkably malignant tumors that can develop in either the bone or soft tissue. Also referred to as Ewing sarcomas, pPNETs are commonly diagnosed in children and young adults, with the median age being around 15 years. These tumors are believed to originate from neuroectodermal cells, the cells contributing to the formation of the nervous system and other tissues.[2]

PNETs of the spine are extremely rare, with only a few cases reported in the literature. PNETs represent less than 1% of spinal tumors according to international literature, the annual incidence of this condition is estimated to range from 0.2-0.4 cases per 100,000.[3,4] Epidemiological evidence in Mexico is similar to that documented in international reports.[5] Symptoms can vary from the initial onset of pain to established radiculopathy, and it is even a diagnosis to consider in cases of cauda equina syndrome.[2] A final diagnosis requires immunohistochemical analysis and cytogenetic studies.[6]

The treatment for pPNETs of the spine is multimodal, involving surgery, chemotherapy, and radiation therapy. Unfortunately, the prognosis for patients with spinal pPNETs is disheartening, with a median survival rate spanning just 18 months.[6] This article presents a case involving a 19-year-old male who was diagnosed with a primary spinal extradural PNET. We will outline the patient's clinical presentation, detail the radiological findings, discuss the pathological diagnosis, and ultimately describe the outcome post-surgery.

CASE PRESENTATION

A 19-year-old male patient with no significant past medical history came to the clinic, reporting four days of lower back pain visual analogue scale (VAS) 7/10 and weakness in his lower extremities. Physical examination revealed an antalgic gait, with dominance in the support phase outweighing initial contact and mid-support phases which caused decreased final support. He displayed bilateral increases in initial and mid-sway, decreased shoulder girdle rotation, increased pelvic girdle rotation and tilt, knee and ankle flexion, and compromised toe-heel gait variants. He also reported severe pain (VAS 9/10) during deep palpation of the lumbar region and displayed bilateral paravertebral contracture. The patient's strength levels were measured as 4/5 for bilateral L2, L3, and L4, whereas they were only 3/5 for L5 and S1. Moreover, he experienced bilateral paresthesia from L2 to S1, hyporeflexia in L4 bilaterally, and complete areflexia in S1. Lasègue, Bragard, and Patrick's signs were positive on the

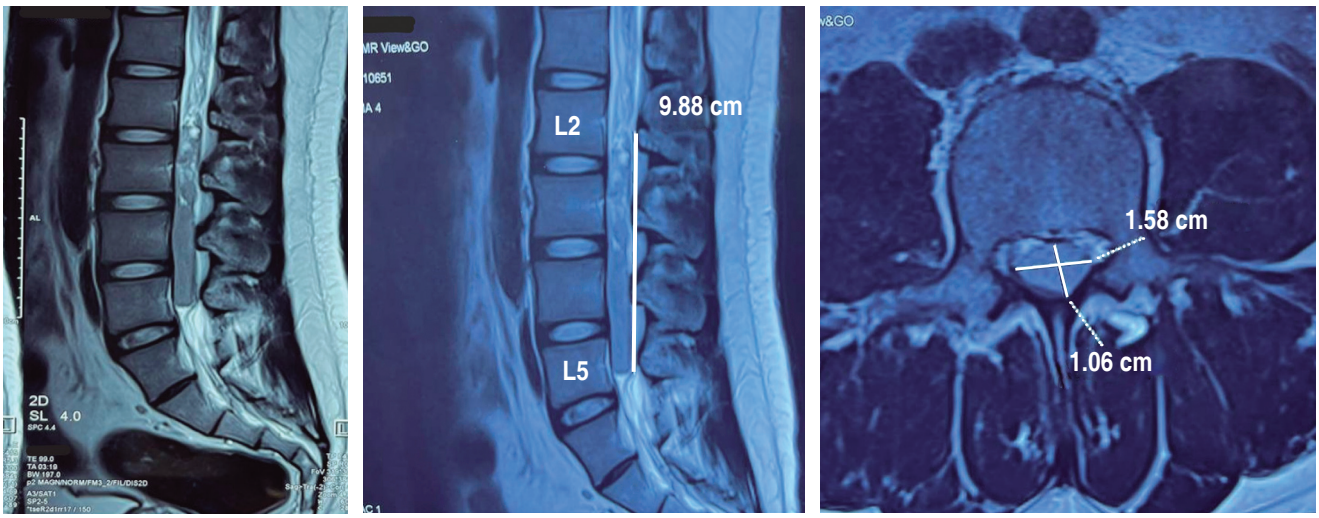


Figure 1: Magnetic resonance imaging of the lumbar spine. The cauda equina exhibited clear irregularity, associated with a heterogeneous lesion that was primarily hyperintense on the T1 sequence and slightly hyperintense on the T2 sequence. This lesion was approximately 9.3 cm in length.

right while Clonus and Babinski's signs were positive bilaterally. The normal functioning of the patient's urinary and anorectal sphincters was observed.

Anterior posterior (AP), lateral, oblique, and dynamic lumbar spine X-rays were normal. The simple and contrasted magnetic resonance imaging (MRI) of the lumbar spine revealed an abnormal mass in the cauda equina *(Figure 1)*.

The electromyography (EMG) and somatosensory evoked potentials (SSEPs) of the lower extremities revealed a quality decrease in the amplitude of myotomes from L2 to L4 bilaterally, which were absent in L5 and S1.

The posterior lumbar approach was utilized for decompression and posterolateral fixation of L2 to S1, along with total excision of the intradural tumor, all of which were guided by intraoperative neurological monitoring *(Figure 2)*.

The round tumor lesion had irregular borders, appearing blackish and sandy. It was roughly 3 × 4 cm in diameter. The lesion was intradural and centrally positioned, compressing the cauda equina between L2 and L5 *(Figure 3)*.

The histological examination of the tumor revealed a primary neuroectodermal tumor with a positive report for malignant neoplasm. An immunohistochemical analysis was conducted, and the neoplastic cells exhibited positive and widespread expression for CD99 and Ki-67 at 40%. However, epithelial membrane antigen (EMA) and glial fibrillary acidic protein GFAP were negative *(Figure 4)*.

The medical treatment was established with three-dimensional conformal radiotherapy, 25 sessions in five weeks follow-up. In the immediate postoperative period, the patient was alert and neurologically sound. He experienced pain in the lumbar region (VAS 2/10) and bilaterally and had a strength of 5/5 for L2, L3, and L4, and 4/5 for L5 and S1. His sensitivity remained normal on both sides, with no signs of neural tension. Neither Clonus nor Babinski's signs were present, the cremasteric reflex was active, whereas the bulbocavernosus reflex was diminished. Unfortunately, the patient did lose control over his urinary and anorectal sphincters. Accordingly, a pelvic floor examination by urology and physiotherapy was requested.

Three weeks later, after undergoing urological treatment with a urinary catheter and completing 12 sessions of pelvic floor physiotherapy, the patient regained control of his anorectal sphincter and experienced a reduction in lower back pain. He was able to walk plantigrade bipedally, with only minor claudication in the right pelvic limb.

DISCUSSION

PNETs originate in cells derived from the neural crest and can develop in either the central or

peripheral nervous system.[6] About 85% of tumors diagnosed as PNETs have t(11;22) (q24;q12) chromosomal translocations. At the molecular genetic level, the chromosome 22q12 breakpoint is involved within a single gene called EWS, whereas the chromosome 11q24 breakpoint is located in the FL1 gene. The resulting EWS-FLI1 fusion is consistently found in these tumors, highlighting its pivotal role in the biological processes underlying PNET development and progression.[7] PNETs are rare, particularly in adolescents, and literature on the matter is limited.

The incidence of pPNETs is more common in the thoracic and cervical regions, and it is more frequent in males. Clinical findings are nonspecific and depend on the location and extent of tumor invasion.[1]

The literature provides little information about the imaging characteristics due to the rarity and lack of awareness of this specific presentation.[8] Generally, a combination of imaging methods, such as magnetic resonance imaging and computed tomography, is required. Furthermore, tissue studies through a biopsy or radical resection of the tumor are needed to confirm the diagnosis and determine appropriate treatment, given its rarity and limited evidence on therapeutic aspects.[9-13] Microscopically, these tumors are characterized by small, uniform round cells with round nuclei, fine chromatin, and scanty eosinophilic cytoplasm. These tumors are categorized as either Ewing's sarcoma or PNET based on their level of differentiation. Ewing's sarcoma is assigned to tumors that show no evidence of neuroectodermal differentiation, while PNET is attributed to tumors that display Homer Wright rosettes, which are structures formed by tumor cells arranged in a circular pattern around a central neurite.[14] In our patient, the final diagnosis was immunohistochemical; tumor cells were characterized by the presence of CD99 and Ki-67 and immunoreactivity for synaptophysin, indicating neuroectodermal differentiation. These findings led the pathologist to classify the tumor as a PNET.

PNET develops from early nerve cells. These cells are typically present in the body during embryonic development, but they usually mature into neurons and disappear. However, in rare cases, these cells can develop into a tumor.[11,12] Spinal cord and cauda equina tumors can be of many different types.

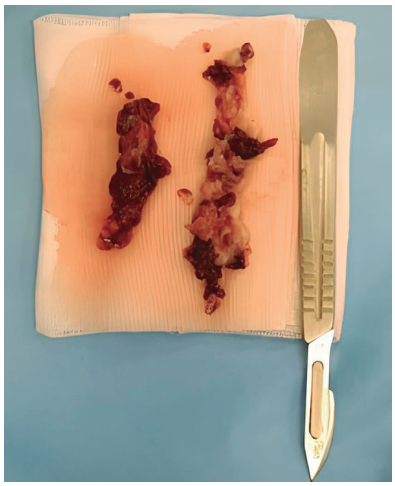
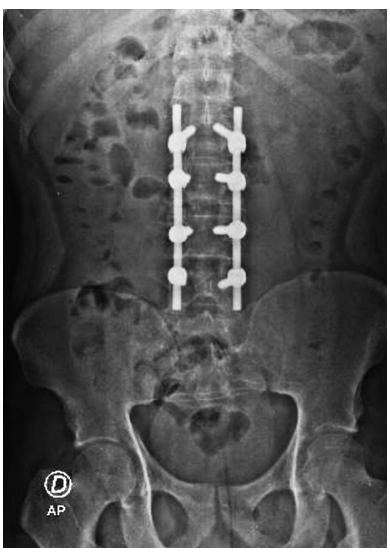


Figure 3:

Intraoperative tumor. A well-delineated, round tumor approximately 3 × 4 cm in diameter is observed. The lesion displays irregular borders and a blackish, granular appearance.



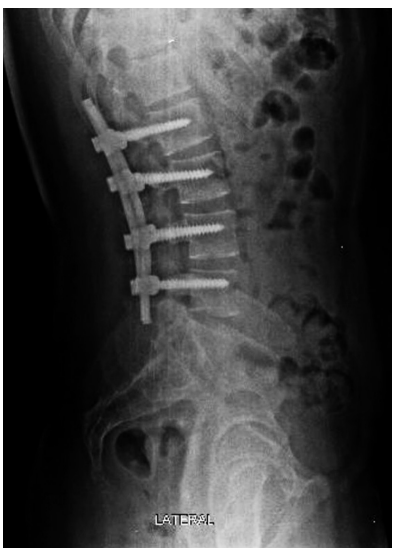


Figure 2:

Postquirurgic anterior posterior and lateral X-rays. Posterior lumbar approach, decompression, posterolateral fixation of L2 to S1.

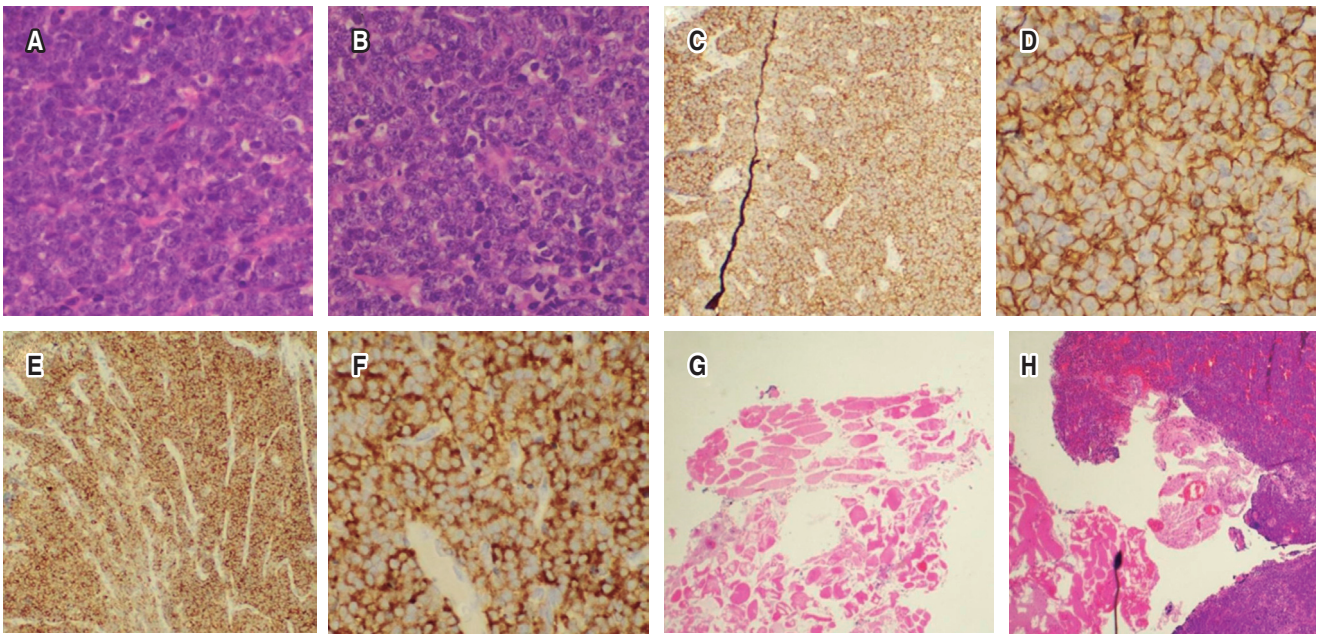


Figure 4: Immunohistochemical results. **A-B)** Histological sections with hematoxylin and eosin identify high-grade neoplasia arranged in a growth pattern of sheets of small, round, blue cells; the features include a nucleus with fine chromatin and scanty cytoplasm, revealing abundant mitosis figures in the neoplasia. **C-D)** The samples express CD99 and Ki-67 at 40%, with a diffuse positive expression, while EMA and GFAP show negative expression. **E-F)** Synaptophysin markers are also presented. **G-H)** Striated muscle and a neoplasm are identified, and arranged in a solid growth pattern.
EMA = Epithelial membrane antigen. GFAP = Glial fibrillary acidic protein.

Potential diagnoses for intradural spinal tumors include meningioma, nerve sheath tumors such as schwannoma or neurofibroma, astrocytoma, ependymoma, and metastatic growth.[12]

Since there are no standard treatment guidelines, treatment typically involves a multidisciplinary approach that may include chemotherapy, surgery, radiation therapy, and bone marrow transplants. Some patients might benefit from targeted therapies that focus on the specific genetic characteristics of cancer cells.[14,15] Radiation therapy, used as part of a multimodal therapeutic approach, has shown satisfactory responses in some cases.[13-15] However, we believe its use should be individually planned, taking into account the aforementioned clinical factors. Numerous factors determine prognosis and guide the choice of appropriate treatment. These include the age of the patient, the size and location of the tumor, the existence of distant metastases.[16] It also necessitates medium to long-term follow-up to assess treatment response and detect potential side effects.

Nonetheless, the long-term outlook remains unfavorable due to the aggressive nature of these tumors. On average, survival is around 18 months after diagnosis.[6]

PNET is rare and challenging to treat. Early detection and specialized care are crucial for favorable outcomes. Our patient underwent radiotherapy and recovered well with only minor complications.

CONCLUSIONS

This case report highlights the rarity of intradural peripheral primitive neuroectodermal tumors (PNETs) within the spinal cord. Given the scarcity of similar cases described in both national and international literature, this article provides valuable clinical information on a highly aggressive and rare tumor entity. It underscores the importance of considering PNETs in the differential diagnosis of adolescents presenting with lower extremity weakness and back pain. Moreover, the detailed description of the clinical presentation, radiological findings, surgical treatment, and immunohistochemical results enriches the available literature, particularly in Latin American populations where such documentation remains

limited. Early detection of this neoplasm may allow timely diagnosis and improve therapeutic decision making and patient outcomes.

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