Turkish Medical Journal, 2025; 10(3),124-128.

Olgu Sunumu / Case Report
https://doi.org/10.70852/tmj.1642164

A RARE DIAGNOSIS IN A CHILD PRESENTING WITH URINARY INCONTINENCE: SPINAL EPENDYMOMA

Üriner inkontinans ile başvuran çocukta nadir bir tanı: Spinal ependimom

Hatice Zeynep TERZİ[1], Muhammed Mustafa GÜNEYLİOĞLU[2], İnci Pınar BİLEN[1], Tayfun PEHLİVAN[1], Duygu BAYKAL[3], Murat TUTANÇ[1]

Afiliasyon / Affiliation:
[1]Department of Pediatrics University of Health Sciences, Bursa Faculty of Medicine, City Training and Research Hospital, Department of Bursa, Türkiye.

[2]Department of Pediatric Emergency, University of Health Sciences, Bursa Faculty of Medicine, City Training and Research Hospital Bursa, Türkiye.

[3]Department of Neurosurgery, University of Health Sciences, City Training and Research Hospital, Bursa, Türkiye.

Sorumlu Yazar / Correspondence:
Hatice Zeynep Terzi, MD
Department of Pediatrics, University of Health Sciences Bursa Faculty of Medicine, City Training and Research Hospital Doğanköy Neighb., Gümüş Ave #10, 16110, Nilüfer, Bursa, Türkiye
E-mail: zeyneepasann@gmail.com

Geliş / Received: 18.02.2025
Kabul / Accepted: 25.04.2025

Cite as:
Terzi, H. Z., Güneylioğlu, M. M., Bilen, İ. P., Pehlivan, T., Baykal, D., Tutanç, M. (2025). A rare diagnosis in a child presenting with urinary incontinence: spinal ependymoma Turkish Medical Journal, 10(3),124-128. https://doi.org/10.70852/tmj.1642164

ABSTRACT

Patients presenting with various symptoms to the pediatric emergency department may be diagnosed with a spinal mass. Spinal ependymoma is a central nervous system tumor that is quite rare in childhood. Early diagnosis is important in terms of reducing morbidity and mortality. The presenting symptoms should be well understood, and necessary tests for differential diagnosis should be conducted. In this case, a 9-year-old male patient who presented to our emergency department with complaints of urinary incontinence and walking difficulties and was diagnosed with spinal ependymoma will be discussed.

Keywords: Ataxia, Ependymoma, Spinal mass, Urinary incontinence

ÖZET

Çocuk acil servisine çeşitli semptomlarla başvuran hastalar spinal kitle tanısı alabilmektedir. Spinal ependimom, çocukluk çağında oldukça nadir görülen merkezi sinir sistemi tümörüdür. Erken tanı konulması morbidite ve mortalitenin azaltılması açısından önemlidir. Başvuru semptomları iyi bilinmeli ve ayırıcı tanıya yönelik gerekli tetkikler yapılmalıdır. Bu vakada, acil servisimize idrar kaçırma ve yürüme bozukluğu şikayetleri ile başvuran ve spinal ependimom tanısı alan 9 yaşındaki erkek hastadan bahsedilecektir.

Anahtar Kelimeler: Ataksi, Ependimom, Spinal kitle, Üriner inkontinans

INTRODUCTION

In children, spinal cord tumors constitute less than 10% of all pediatric central nervous system tumors, and approximately 30% of these are ependymomas (Hsu & Jallo, 2013). Patients with spinal masses most commonly present with symptoms such as weakness in the lower extremities, numbness, ataxia, and sphincter dysfunction (Bach et al., 1990). In cases where spinal cord tumor is suspected, neuroimaging methods should be performed for diagnosis. Here, we will discuss a 9-year-old male patient who presented to the emergency department with complaints of urinary incontinence, had a detailed history of gait disturbance, constipation, and pain in the sacral region, and was diagnosed with spinal ependymoma by neuroimaging.

CASE

A 9-year-old male patient presented to our pediatric emergency department with complaints of urinary incontinence and sacral region pain that started 3 days ago. The patient, unable to urinate voluntarily, was experiencing dribbling incontinence that was continuous and had inability to pass flatus or stool for the past three days. In his history, there was no trauma, poisoning, dysuria, discoloration of urine, headache, fever, recent infection, or vaccination history. Upon detailed questioning, it was learned that he had been limping for 3 months but had not consulted any center for this complaint. At the time of admission, the patient's general condition was good, vital signs were stable, and the Glasgow Coma Scale was 15. In the neurological examination, cranial nerves were intact, deep tendon reflexes were normal, and pathological reflexes were not detected. There were no signs of meningeal irritation. In the upper extremities, muscle strength was 5/5, while in the lower extremities, it was 3/5 bilaterally in the distal regions. In the sensory examination, loss of sensation was detected in the L3 - L5 dermatomes. In the examination of other systems, a palpable distended bladder was detected, and due to the presence of dribbling-type urinary incontinence, overflow urinary incontinence was considered. No pathology was detected in the examination of the sacral region. During the rectal examination, there was fecal contamination, but the anal tone was weak. There were no significant features in the personal and family history. In the preliminary evaluation, differential diagnoses included myelitis, Guillain-Barré syndrome, and a spinal mass. To assess for urinary retention possibly related to neurogenic bladder dysfunction, bladder catheterization was performed, yielding 600 cc of urine output. This finding supported a neurological etiology. Laboratory investigations, including blood and urine tests, were unremarkable, reducing the likelihood of infectious or systemic inflammatory causes such as myelitis. Additionally, abdominal ultrasound was performed to rule out urological or structural abdominal pathologies and revealed no abnormalities. Given the persistence of neurological symptoms and suspicion of a spinal lesion, contrast-enhanced lumbosacral magnetic resonance imaging (MRI) was obtained. The MRI revealed an intradural, extra-axial mass extending from the L4 to S1 vertebral levels, occupying the spinal canal and demonstrating heterogeneous hypointense signals (Figure 1). Based on these imaging findings, the patient was diagnosed with a spinal cord tumor.

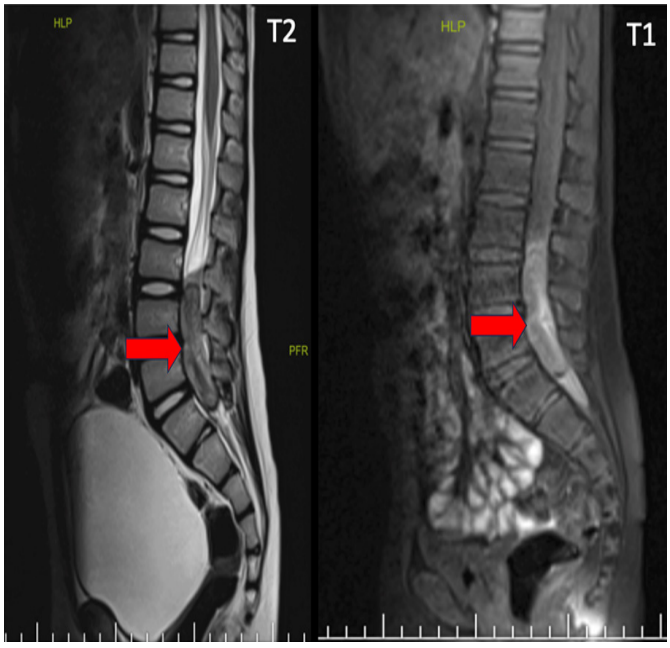

Figure 1. Lumbar magnetic resonance imaging a) T2 sequence showing an intradural extra-axial mass with heterogeneous hypointense signals in the spinal canal at the L4-L5-S1 level and a globular bladder appearance b) T1 sequence showing heterogeneous enhancement of the mass at the L4-L5-S1 level.

Dexamethasone at a dose of 0.5 mg/kg was administered to the patient with spinal cord compression in the emergency department. During follow-up, there was no hypotension or bradycardia. The patient was consulted with neurosurgery. A contrast-enhanced cranial MRI was performed for metastasis; no lesions compatible with metastasis were observed. The patient was operated on by neurosurgery, and the mass was totally resected. The patient discharged on the 7th day of follow-up with recommendations for physical therapy and during the first week, the patient continued to experience urinary incontinence and constipation. By the end of the first month, these symptoms had significantly improved. The pathology result was grade 2 papillary ependymoma. The pediatric oncology department did not recommend sequential treatment and decided to follow up with intermittent imaging. The patient's follow-up is being continued at our center. Informed consent has been obtained from the family for this case presentation.

DISCUSSION

This study presents the pediatric case with complaints of urinary incontinence, gait disturbance, constipation, and pain in the sacral region, and was diagnosed with spinal ependymoma. Although central nervous system tumors are frequently seen in pediatric age, spinal cord tumors are quite rare (~10%) (Farzan et al., 2022). Their frequency is higher in men compared to women (McGuire, Sainani, & Fisher, 2009). They are histologically benign character and slow-growing tumors (Martinez-Perez et al., 2012). Ependymal tumors are neuroepithelial malignancies of the central nervous system that can be seen in both children and adults (Korshunov et al., 2010). Spinal ependymomas are most common in adults between the ages of 20 and 40 (Pajtler et al., 2015). They stated that in children with spinal tumors, the most common symptoms are impairment of motor functions (74.2%), pain (40%), sensory findings (paresthesia, hypoesthesia, anesthesia) (16.6%), and urinary symptoms (11.4%) (inability to urinate/ incontinence). Due to the non-specific early-stage symptoms, it is difficult to diagnose without high clinical suspicion (Farzan et al., 2022). Our case also presented to our emergency department with the urinary incontinence. Early diagnosis of pediatric spinal tumors is important for preventing morbidity and ensuring effective treatment. Due to the spinal canal being narrower in children compared to adults, symptoms related to spinal masses appear at an earlier stage (Ekuma et al., 2017). In cases of spinal ependymoma, the most common symptoms are localized pain, spasticity in the lower extremities, sensory loss, and gait disturbances (Handbook of Clinical Neurology, 1997). Sofuoğlu et al., in their study of 46 pediatric spinal cord tumor cases, stated that the most commonly observed presenting symptoms were weakness and numbness in the lower extremities (Sofuoğlu & Abdallah, 2018). There is also a risk of spontaneous bleeding in spinal ependymomas. In cases with bleeding, sudden onset back pain may occur, and this can happen before neurological deficits appear (Ekuma et al., 2017). Our patient presented with motor weakness, sensory deficits, ataxia, and sphincter dysfunction. Spinal cord tumors can be intradural (45%) or extradural (55%), and those that are intradural are divided into intramedullary or extramedullary (Parsa et al., 2004). The mass in our case was situated intradural extramedullary. Ependymomas are graded histologically based on their characteristics, such as cell structure, mitotic activity, pleomorphism, necrosis, and vascular proliferation (Louis et al., 2016), and are classified into three grades. Myxopapillary ependymoma and subependymoma are benign and grade I lesions. Grade II lesions have weak anaplastic features; they have classic, cellular, papillary, clear cell, and tanycytic subtypes. Our patient was classified histopathologically as a grade 2 papillary ependymoma. Anaplastic ependymomas, on the other hand, are grade III and are the most aggressive type (Godfraind et al., 2012; Kresbach, Neyazi, & Schüller, 2022). In pediatric patients with central nervous system tumors, the 10-year survival rate is approximately 64% (Tihan et al., 2008). In addition to surgical resection and radiotherapy, corticosteroids such as dexamethasone are frequently used to reduce peritumoral edema and alleviate symptoms. Several studies have reported its short-term benefits in controlling neurological symptoms (Dixit & Kumthekar, 2020). In our case, dexamethasone was used and a response was obtained to the treatment. Gross total resection

forms the basis of spinal ependymoma treatment; in cases with a residual mass, radiotherapy can be added to the treatment. Benesch et al. reported that the overall survival rate was 100% with gross total resection treatment in 29 pediatric primary spinal ependymoma cases (Benesch et al., 2010). The extent of surgical resection in ependymomas has long been considered the only prognostic factor associated with survival (Merchant et al., 2009). Khalid et al. retrospectively evaluated survival in histologically confirmed spinal ependymomas in patients aged 17 and under; they found that tumor size and the extent of total resection were associated with survival (Khalid et al., 2018). Tihan et al., in their study of 96 pediatric central nervous system ependymomas, associated the patient's age, tumor location, extent of resection, and radiation therapy status with survival (Tihan et al., 2008). Rudà et al. stated that histopathological criteria alone cannot predict prognosis; more studies are needed for this (Rudà et al., 2018). Our patient was 9 years old and the intradural and extraaxial location of the tumor may have caused the clinical course to be milder.

CONCLUSION

Patients presenting to the pediatric emergency department with overflow urinary incontinence should be evaluated for signs of spinal cord compression. The rarity of spinal ependymoma cases in children makes it difficult to diagnose cases and establish a standard treatment protocol. Our case is significant not only for contributing to the pediatric spinal ependymoma literature but also for demonstrating that cases can be diagnosed in the emergency department with a detailed history and physical examination. Spinal tumors, although rare in children, should be considered in the differential diagnosis of persistent or progressive neurological symptoms, back pain, gait disturbances, or urinary incontinence. Early spinal MRI is crucial for timely diagnosis and intervention.

Ethical approval: A written consent form was obtained from the families for this publication.
Conflict of Interest: The authors declared no conflicts of interest with respect to authorship and/or publication of the article.
Financial Disclosure: The authors received no financial support for the research and/or publication of this article.
Author Contributions: Hatice Zeynep Terzi: Having an idea/opinion or contributing to the emergence and maintenance of the article/study,Muhammed Mustafa Güneylioğlu: Plan, design or pattern. İnci Pinar Bilen: Revision, audit, review. Tayfun Pehlivan: Fund supply. Duygu Baykal: Revision, audit, review,Having an idea/opinion or contributing to the emergence and maintenance of the article/study. Murat Tutanç: Literature review.

REFERENCES

Bach, F., Larsen, B. H., Rohde, K., Børgesen, S. E., Gjerris, F., Bøge-Rasmussen, T., Agerlin, N., Rasmusson, B., Stjernholm, P., & Sørensen, P. S. (1990). Metastatic spinal cord compression. Occurrence, symptoms, clinical presentations and prognosis in 398 patients with spinal cord compression. Acta neurochirurgica, 107(1-2), 37–43. https://doi.org/10.1007/BF01402610

Benesch, M., Weber-Mzell, D., Gerber, N. U., von Hoff, K., Deinlein, F., Krauss, J., Warmuth-Metz, M., Kortmann, R. D., Pietsch, T., Driever, P. H., Quehenberger, F., & Urban, C. (2010). Ependymoma of the spinal cord in children and adolescents: A retrospective series from the HIT database. Journal of Neurosurgery: Pediatrics, 6(2), 137–144. https://doi.org/10.3171/2010.5.PEDS09553

Dixit, K. S., & Kumthekar, P. U. (2020). Optimal Management of Corticosteroids in Patients with Intracranial Malignancies. Current Treatment Options in Oncology, 21(9), 77. https://doi.org/10.1007/s11864-020-00771-7

Ekuma, E. M., Ito, K., Chiba, A., Hara, Y., Kanaya, K., Horiuchi, T., Ohaegbulam, S., & Hongo, K. (2017). A Rare Case of Pediatric Lumbar Spinal Ependymoma Mimicking Meningitis. World neurosurgery, 100, 710. e1–e5. https://doi.org/10.1016/j.wneu.2017.02.016

Farzan, A., Ahmadi, P., Tasdighi, E., Zinatzadeh, M. R., & Pourbakhtyaran, E. (2022). Primary spinal tumors and masses in children. Iranian Journal of Child Neurology, 16(2), 129–135. https://doi.org/10.22037/ijcn.v16i2.30614

Godfraind, C., Kaczmarska, J. M., Kocak, M., Dalton, J., Wright, K. D., Sanford, R. A., Boop, F. A., Gajjar, A., Merchant, T. E., & Ellison, D. W. (2012). Distinct disease-risk groups in pediatric supratentorial and posterior fossa ependymomas. Acta neuropathologica, 124(2), 247–257. https://doi.org/10.1007/s00401-012-0981-9

Handbook of Clinical Neurology. (1997). Intrinsic spinal cord tumors (Vol. 24, p. 497). Elsevier.

Hsu, W., & Jallo, G. I. (2013). Pediatric spinal tumors. Handbook of Clinical Neurology, 112, 959–965. https://doi.org/10.1016/B978-0-444-52910-7.00016-7

Khalid, S. I., Kelly, R., Adogwa, O., Carlton, A., Woodward, J., Ahmed, S., Khanna, R., Bagley, C., Cheng, J., Shah, S., & Mehta, A. I. (2018). Pediatric spinal ependymomas: An epidemiologic study. World Neurosurgery, 115, e119–e128. https://doi.org/10.1016/j.wneu.2018.03.206

Korshunov, A., Witt, H., Hielscher, T., Benner, A., Remke, M., Ryzhova, M., Milde, T., Bender, S., Wittmann, A., Schöttler, A., Kulozik, A. E., Witt, O., von Deimling, A., Lichter, P., & Pfister, S. (2010). Molecular staging of intracranial ependymoma in children and adults. Journal of clinical oncology : official journal of the American Society of Clinical Oncology, 28(19), 3182–3190. https://doi.org/10.1200/JCO.2009.27.3359

Kresbach, C., Neyazi, S., & Schüller, U. (2022). Updates in the classification of ependymal neoplasms: The 2021 WHO Classification and beyond. Brain Pathology, 32(4), e13068. https://doi.org/10.1111/bpa.13068

Louis, D. N., Perry, A., Wesseling, P., Brat, D. J., Cree, I. A., Figarella-Branger, D., Hawkins, C., Ng, H. K., Pfister, S. M., Reifenberger, G., Soffietti, R., von Deimling, A., & Ellison, D. W. (2021). The 2021 WHO Classification of Tumors of the Central Nervous System: a summary. Neuro-oncology, 23(8), 1231–1251. https://doi.org/10.1093/neuonc/noab106

Martinez-Perez, R., et al. (2012). Acute neurological deterioration as a result of two synchronous hemorrhagic spinal ependymomas. Surgical Neurology International, 3, 33. https://doi.org/10.4103/2152-7806.93865

McGuire, C. S., Sainani, K. L., & Fisher, P. G. (2009). Incidence patterns for ependymoma: A surveillance, epidemiology, and end results study. Journal of Neurosurgery, 110(4), 725–729. https://doi.org/10.3171/2008.9.JNS08117

Merchant, T. E., Li, C., Xiong, X., Kun, L. E., Boop, F. A., & Sanford, R. A. (2009). Conformal radiotherapy after surgery for paediatric ependymoma: A prospective study. Lancet Oncology, 10, 258–266.

Pajtler, K. W., Witt, H., Sill, M., Jones, D. T., Hovestadt, V., Kratochwil, F., Wani, K., Tatevossian, R., Punchihewa, C., Johann, P., Reimand, J., Warnatz, H. J., Ryzhova, M., Mack, S., Ramaswamy, V., Capper, D., Schweizer, L., Sieber, L., Wittmann, A., Huang, Z., … Pfister, S. M. (2015). Molecular Classification of Ependymal Tumors across All CNS Compartments, Histopathological Grades, and Age Groups. Cancer cell, 27(5), 728–743. https://doi.org/10.1016/j.ccell.2015.04.002

Parsa, A. T., Lee, J., Parney, I. F., Weinstein, P., McCormick, P. C., & Ames, C. (2004). Spinal cord and intradural-extraparenchymal spinal tumors: Current best practices and strategies. Journal of Neuro-oncology, 69, 291–311.

Rudà, R., Reifenberger, G., Frappaz, D., Pfister, S. M., Laprie, A., Santarius, T., Roth, P., Tonn, J. C., Soffietti, R., Weller, M., & Moyal, E. C. (2018). EANO guidelines for the diagnosis and treatment of ependymal tumors. Neuro-Oncology, 20(4), 445–456. https://doi.org/10.1093/neuonc/nox166

Sofuoğlu, Ö. E., & Abdallah, A. (2018). Pediatric spinal ependymomas. Medical Science Monitor, 24, 7072–7089. https://doi.org/10.12659/MSM.910447

Tihan, T., Zhou, T., Holmes, E., Burger, P. C., Ozuysal, S., & Rushing, E. J. (2008). The prognostic value of histological grading of posterior fossa ependymomas in children: a Children's Oncology Group study and a review of prognostic factors. Modern Pathology, 21(2), 165–177. https://doi.org/10.1038/modpathol.3800999