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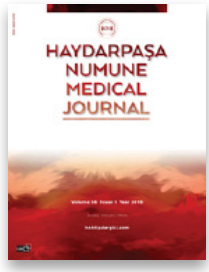

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CASE REPORT

Spinal Myxopapillary Ependymoma: A 12-Year-Old Pediatric Case with Hydrocephalus Presenting with Gait Abnormality

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Abstract

Primary malignant central nervous system tumors are the second most common group of childhood tumors. These tumors can increase the intracranial pressure with compression on the adjacent structures, as well as cause hydrocephalus by obstructing the cerebrospinal fluid (CSF) flow. Ependymomas, which constitute 10% of intracranial tumors, are responsible for 40-60% of spinal tumors in children and young adults. Its incidence has been reported as 0.3/100,000. Myxopapillary ependymomas are usually slow growing and benign lesions. Patients may present with spinal deformities such as scoliosis, torticollis or gait abnormalities.

Keywords: Hydrocephalus; gait apraxia; spinal cord neoplasm.

Primary malignant central nervous system tumors are the most common childhood tumors after hematological malignancies[1]. These tumors can increase intracranial pressure with local invasion, compression of adjacent anatomical structures and mass effect, as well as cause obstruction in CSF flow and cause hydrocephalus[2,3]. Ependymomas, which constitute 10% of intracranial tumors, are responsible for 40-60% of spinal tumors in children and young adults. Its incidence has been reported as 0.3/100,000. Approximately 90% of ependymomas in children are intracranial, 90% of which originate from posterior fossa, and the remainder originates from the spinal cord[4].

Case Report

A 12 years and 10 months-old girl with no known history of chronic disease applied to our clinic with complaints of medially shifting in the right eye for the last 1.5 months, difficulty in walking for 3 months, difficulty bending over, posture disorder, and bilateral thigh and knee pain. In her systemic examination, Glasgow Coma Scale (GCS) score was 15, she had a normal state of consciousness and was oriented; bilateral lower extremity muscle strength was 5/5, sensory examination was normal, had passive medially shifting in the right eye, and had limited outward gaze in the left eye. In the fundus examination and visual field test, bilateral grade 4 optic disc swelling on the right and grade 3 on the left was observed. Total depression in the visual field on the right eye, and nasal step on the left eye was detected. The case was hospitalized considering the differential diagnosis of a brain tumor and pseudotumor cerebri. In blood tests, leukocyte was 6850/ml, ANA was negative, anti-dsDNA was <10, erythrocyte sedimentation rate (ESR) was 4 mm/h, CRP was negative and rheumatoid factor (RF) was 2 IU/ml. Diffuse ventricular dilatation was

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observed in non-contrast cranial computed tomography (NCCT). Cranial magnetic resonance imaging (MRI) showed an increase in CSF distances in both lateral ventricles and the third ventricle, basal cisterns and sella. Due to findings such as enlargement of the optic nerve sheath, kinking of the optic nerve, and flattening on the scleral surface of the optic nerve, more prominently on the right, observed in orbital MRI was evaluated radiologically as compatible with the findings of intracranial hypertension due to hydrocephalus (Fig. 1).

No abnormal findings were detected in the magnetic resonance venography (MRV) which was performed based on the pediatric neurology consultation request.

In the spinal MRI on the 3rd day of the patient's hospitalization; at the level of L4, L5, S1 vertebral bodies, a 7.5 x 2 cm intradural-extramedullary localized lesion was detected between the cauda equina fibers, which was hypointense in T1-weighted (T1W) series and hyperintense in T2W compared to the spinal cord. Hypointense areas in the lesion were observed in T2A series, the lesion showed homogeneous contrast enhancement, and caused peripheral removal of the cauda equina fibers, and was not associated with adjacent vertebral bone structures and subcutaneous soft tissues (Fig. 2). When the patient's age and radiological findings were evaluated together, spinal ependymoma and schwannoma were considered in the differential diagnosis of the lesion. Surgery was planned for the patient by the neurosurgery clinic. The pathology result of the patient was primary myxopapillary ependymoma of the spinal cord (World Health Organization [WHO] 2016 - Grade 1).

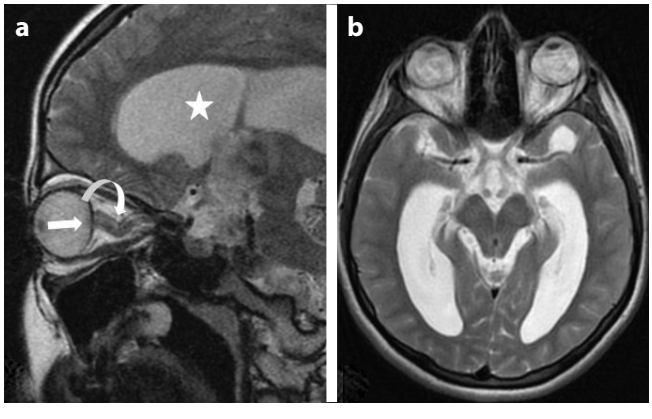

Figure 1. (a) shown that, enlargement of the optic nerve sheath, kinking of the optic nerve (curved arrow), flattening on the scleral surface of the optic nerve (straight arrow) and dilated lateral ventricles (star) on T2-weighted sagittal orbit MRI. **(b)** shown the dilated lateral ventricular trunk and temporal horns on T2-weighted axial cranial MRI of the 12-year-old patient.

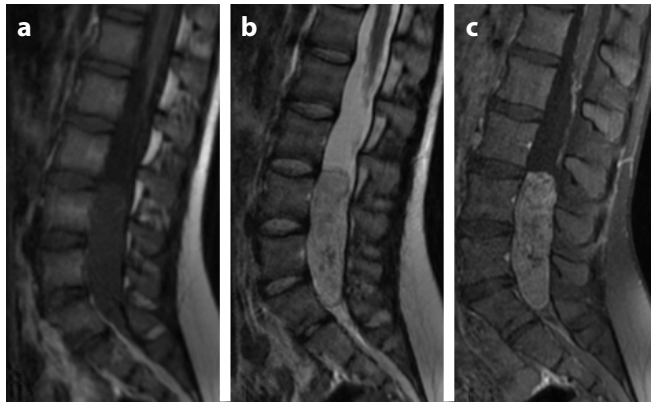

Figure 2. Spinal mass located at the lumbar level was: **(a)** Isointense at T1A sagittal series, **(b)** T2A-weighted sagittal series showed hyperintensity with millimetric hypointense areas, and **(c)** homogeneously enhanced in the post-contrast series.

Discussion and Conclusion

Spinal cord tumors in children show a very low incidence rate with an annual incidence of one in a million[5]. These tumors cause diagnostic delays, especially in young children, due to their high symptom diversity but low specificity. Another reason for diagnostic delay is that secondary spinal deformities such as kyphosis and scoliosis caused by the tumor are already common in this age group[6].

In the study published by Duong et al.[7] in 2012 on spinal tumors, in which 11,712 primary spinal tumors were evaluated between 1999 and 2007, 22% of the cases were reported as malignant and 78% as non-malignant. The incidence of non-malignant spinal tumors was 0.76/100000, which was significantly higher than the incidence of malignant tumors of 0.22/100000. The incidence of malignant spinal tumors was reported as the lowest (0.23/100000) in children aged 0-9 years, and the highest as 2.53/100000 in those aged 70-79 years. The incidence between the ages of 0-19 was stated as 0.27/100000.

In a study published by Schellingen et al.[8] in 2008, they defined the spinal tumor incidence as the lowest between 0-19 years and the highest between 75-84 years.

In the study of Patil et al.[9] in which they analyzed 19,017 spinal cord tumor patients who underwent spinal cord resection, 19% of the patients were under the age of 18. In this study, they found an incidence of 0.24/100000 for all ages, and a lower incidence of 0.014/100000 under 18 years of age. However, this study reported an increased incidence of malignant spinal tumors in patients over 60 years of age.

The origin of primary spinal tumors was found most frequently in the spinal cord composition (60.5%), followed by the spinal meninges (36%) and in the cauda equina (3.5%)[7].

Ependymomas can occur anywhere along the spinal cord, but the cervical cord is the most commonly affected (44%). In addition, 23% occurs within the cervical cord and extends into the upper thoracic cord; 26% occur only in the thoracic cord. Myxopapillary ependymomas constitute 13% of all spinal ependymomas and are the most common tumors of the cauda equina. The majority are intradural and extramedullary spinal tumors that occur at the level of the lumbosacral vertebrae including the filum terminale and/or the conus medullaris. Less commonly, they extend from the lumbar level to the level of the thoracic vertebrae[10]. In our case, the lesion was observed in the filum terminale fibers, where it is most frequently seen.

Among the studies on symptoms in the literature, in the review where Merlot et al.[11] evaluated 21 cases, the main symptom was spinal pain in 12 cases. As sensory symptoms, hypoesthesia and anesthesia were present in 7 of 14 cases, and paresthesia was present in 4 cases. Scoliosis was present in 11 cases at the time of diagnosis. Three cases presented with torticollis. Thirteen cases showed motor deficit; 8 cases had mild weakness; and 3 cases were paraplegic; 9 cases showed unilateral weakness. Urinary incontinence requiring urinary catheterization was reported in 6 cases. Tumors were found at the dorsal level in 9 cases, at the cervical level in 4 cases, and at the dorsolumbar level in 4 cases. Again in this study, diagnostic delay was attributed to low specificity of existing symptoms. In the presence of findings such as scoliosis, constipation and back pain, the possibility of spinal tumor is not considered in the first place, since the incidence is low in the childhood age group. In addition, it was emphasized in this study that idiopathic scoliosis was painless and showed right convexity, and especially scoliosis caused by spinal tumors were painful and showed left convexity. In the presence of painful and levoconvex scoliosis, evaluation with pineal MRI is recommended to exclude a spinal mass. In our case, complaints of pain with movement and posture disorder were described, and a mild levoconvex scoliosis in the thoracic axis was detected.

In the study of Auguste et al.,[12] in accordance with the literature, back pain is expressed as the most common symptom in 2/3 of cases with spinal tumors.

In the literature, motor deficits in spinal tumor symptomatology are findings that occur in the early period in children and are seen in 70% of the cases. Most commonly, an increase in falls, gait disturbances, weakness, limping and difficulty walking, even progressing to crawling in young children have been shown.[13] In our case, difficulty in walking, poor posture and need for support were the main symptoms.

Weakness in the upper extremity and dropping objects are other motor deficits that can be seen. These findings are initially unilateral. Sphincter dysfunctions occur late. Sometimes, symptoms such as constipation, increased urine volume, or inability to have full bladder control at normal age may be present at the beginning. Spinal cord tumors may cause 10% of intracranial hypertension syndrome cases. In our case, headache, detection of papilledema in physical examination, and presence of dilatation in the lateral ventricles in cranial MRI indicated an increased CSF pressure. Although sensory deficits may be difficult to diagnose in young children, they can be observed in 20% of cases. Pain can be observed in infants as irritability and behavioral changes[14].

Myxopapillary ependymomas are usually slow growing and benign lesions. Sometimes they spread with CSF and intracranial secondary lesions in the central nervous system can be seen with a frequency of 14-43%[15]. Some sacral lesions are locally aggressive and metastasize to lymph nodes, lung, and bone. Aggressive behavior is more common in children. In our case, no finding compatible with metastasis was found in the screening examination performed for other systems; the lesion was observed as isolated only in the region where it was located.

MRIs of myxopapillary ependymomas, which are the most prone to bleeding subtype of ependymomas, may show a "cap sign" finding due to bleeding[16]. However, this finding was not found in our case.

We presented this case to draw attention to the diagnosis of spinal tumors, which are rare in childhood, and the presence of non-specific symptoms that may delay the diagnosis. We wanted to emphasize that in the presence of symptoms related to the central nervous system; demyelinating diseases, hydrocephalus or tumors, especially in the presence of symptoms such as lower extremity or upper extremity weakness, gait disturbance, back and low back pain, spinal MRI along with cranial MRI imaging will be very helpful in making the diagnosis. We also point out that spinal tumors should be excluded with spinal MRI in the differential diagnosis of spinal deformities such as scoliosis or torticollis.

Informed Consent: Written, informed consent was obtained from the patient's family for the publication of thís case report and the accompanying images.

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References

1. Linabery AM, Ross JA. Trends in childhood cancer incidence in the U.S. (1992-2004). Cancer 2008;112:416–32.
2. Wilne SH, Dineen RA, Dommett RM, Chu TP, Walker DA. Identifying brain tumours in children and young adults. BMJ 2013 Oct 9;347:f5844.
3. Wilne S, Collier J, Kennedy C, Koller K, Grundy R, Walker D. Presentation of childhood CNS tumours: A systematic review and meta-analysis. Lancet Oncol 2007;8:685–95.
4. Ostrom QT, Gittleman H, Truitt G, Boscia A, Kruchko C, Barnholtz-Sloan JS. CBTRUS statistical report: Primary brain and other central nervous system tumors diagnosed in the United States in 2011-2015. Neuro Oncol 2018;20(Suppl 4):iv1–iv86.
5. Constantini S, Houten J, Miller DC, Freed D, Ozek MM, Rorke LB, et al. Intramedullary spinal cord tumors in children under the age of 3 years. J Neurosurg 1996;85:1036–43.
6. Benes V 3rd, Barsa P, Benes V Jr, Suchomel P. Prognostic factors in intramedullary astrocytomas: A literature review. Eur Spine J 2009;18:1397–422.
7. Duong LM, McCarthy BJ, McLendon RE, Dolecek TA, Kruchko C, Douglas LL, et al. Descriptive epidemiology of malignant and nonmalignant primary spinal cord, spinal meninges, and cauda equina tumors, United States, 2004-2007. Cancer 2012;118:4220–7.
8. Schellinger KA, Propp JM, Villano JL, McCarthy BJ. Descriptive epidemiology of primary spinal cord tumors. J Neurooncol 2008;87:173–9.
9. Patil CG, Patil TS, Lad SP, Boakye M. Complications and outcomes after spinal cord tumor resection in the United States from 1993 to 2002. Spinal Cord 2008;46:375–9.
10. Weber DC, Wang Y, Miller R, Villà S, Zaucha R, Pica A, et al. Long-term outcome of patients with spinal myxopapillary ependymoma: Treatment results from the MD Anderson cancer center and institutions from the rare cancer network. Neuro Oncol 2015;17:588–95.
11. Merlot I, Francois J, Marchal JC, Joud A, Guerbouz R, Chastagner P, et al. Spinal cord tumors in children: A review of 21 cases treated at the same institution. Neurochirurgie 2017;63:291–6.
12. Auguste KI, Gupta N. Pediatric intramedullary spinal cord tumors. Neurosurg Clin N Am 2006;17:51–61.
13. McGirt MJ, Chaichana KL, Atiba A, Attenello F, Woodworth GF, Jallo GI. Neurological outcome after resection of intramedullary spinal cord tumors in children. Childs Nerv Syst 2008;24:93–7.
14. Epstein FJ, Farmer JP. Pediatric spinal cord tumor surgery. Neurosurg Clin N Am 1990;1:569–90.
15. Koeller KK, Rosenblum RS, Morrison AL. Neoplasms of the spinal cord and filum terminale: Radiologic-pathologic correlation. Radiographics 2000;20:1721–49.
16. Kahan H, Sklar EM, Post MJ, Bruce JH. MR characteristics of histopathologic subtypes of spinal ependymoma. AJNR Am J Neuroradiol 1996;17:143–50.