Pediatric spinal schwannomas/neurofibromas constitute only 2.5%–4% of all pediatric spinal tumors. However, subarachnoid hemorrhage (SAH) because of spinal pathologies is very rare, representing 1.5% of all cases of SAH. Spinal nerve sheath tumors such as schwannomas rarely present with SAH, especially before the appearance of overt signs of spinal cord or root compression. We report a case of dorsolumbar schwannoma in an 11-year-old girl presenting clinically with signs and symptoms mimicking meningitis, but meningeal signs later proved to be due to SAH associated with spinal (D12-L1) schwannoma and hydrocephalus. Mass was excised and ventriculoperitoneal shunt was inserted. In our clinical practice, we may sometimes come across some uncommon diseases with even more uncommon presentations as happened with us at our institute. We must always consider that there is a possibility of SAH owing to silent spinal lesion in patients with angiographic negative intracranial SAH as in this case.

**Keywords:** Spinal schwannoma, subarachnoid hemorrhage, hydrocephalus

**INTRODUCTION**

Among pediatric population, primary spinal cord tumors are rare central nervous system neoplasms compared to intracranial tumors, which account for 1%–10% of all pediatric central nervous system tumors.[1-3] Most of these tumors are intramedullary.[4] Pediatric spinal schwannomas/neurofibromas constitute only 2.5%–4% of all pediatric spinal tumors.[1,2] Schwannomas are well-circumscribed intradural or extradural or combined intra-extradural tumors located on peripheral nerves or spinal nerve roots.[5] However, subarachnoid hemorrhage (SAH) because of spinal pathologies is very rare, representing 1.5% of all cases of SAH.[6] The more common causes include spinal trauma, arteriovenous malformations, and saccular aneurysms of spinal arteries.[7,8] Rarely, spinal cord tumors, either primary or metastatic, may cause intracranial SAH, with ependymoma of the conus medullaris accounting for most of these cases.[7,9] Spinal nerve sheath tumors such as schwannomas rarely present with SAH,[6,10] especially before the appearance of overt signs of spinal cord or root compression. Going through literature, we found that very few cases of nerve sheath tumors that developed SAH had been reported in the literature.[8]

We report a case of dorsolumbar schwannoma in an 11-year-old girl presenting clinically with signs and symptoms mimicking meningitis, but meningeal signs later proved to be due to SAH associated with spinal (D12-L1) schwannoma and hydrocephalus.

**CASE DESCRIPTION**

An 11-year-old female child was admitted to this institute with a history of sudden-onset headache and vomiting. On admission, the child was obtunded, afebrile with noticeable neck rigidity, and with positive Kernig’s and Brudzinski’s signs. The computed tomography (CT) scan of brain showed normal parenchyma [Figure 1]. Cerebrospinal fluid (CSF) analysis showed full field of red blood cells. Following lumbar puncture, the patient developed paraparesis with urinary retention.
Examination of lower limbs showed equal bulk, decreased tone, grade II in both the lower limbs, which decreased to grade I, diminished knee and ankle jerks, upgoing plantars, and no sensory loss. General examination also revealed café au lait spots, two on abdomen (3 × 1 and 1 × 1 cm) and three on chest (1 × 1 cm) but no periaxillary or inguinal freckling and no subcutaneous neurofibroma. Both the upper limbs were normal on neurological evaluation. The magnetic resonance imaging (MRI) of brain and spinal cord with contrast was also carried out. MRI brain with angiogram was normal. Spinal MRI showed an intradural-extradural mass at D12-L1 level, which was isointense to hypointense on T1-weighted images, mildly enhancing mass on contrast administration with leptomeningeal enhancement [Figure 2], and hyperintense homogenous on T2-weighted images. D12 laminectomy and micro-decompression with near-total excision of tumor were carried out. Operative findings on opening dura revealed that CSF was hemorrhagic and pressure was elevated. A well-defined capsular intradural-extradural mass engulfing the nerve roots at D12-L1 level anteriorly on the left side of cord was observed. Tumor was soft, suckable, and crushable with intra-tumoral bleed. Histopathologic evaluation showed that tumor was composed of spindle cells with wavy nuclei having hypocellular and hypercellular areas with focal nuclear palisading, the cells stained positive for S100 protein, and the features were suggestive of schwannoma. Postoperatively, the power improved to grade III in both the lower limbs. But 2 weeks after surgery, the child developed sudden-onset frontal headache and deterioration of vision in both the eyes, left more than right. On examination, she had bilateral lateral rectus palsy, only light perception in left eye, and finger counting up to 3 feet in right eye. Light reflex was present in right eye but in left eye, direct reflex was absent but consensual was present. Fundoscopy showed features of papilledema grade II bilaterally. A check on CT brain [Figure 3] showed development of hydrocephalus. Postoperative spinal MRI [Figure 4] revealed no evidence of any residual lesion at operative site with insignificant postoperative changes. The child was then subjected to ventriculoperitoneal shunt. CSF came under high pressure as a jet but was clear watery in appearance. The child also had occasional hypertensive spikes in postoperative period. She was evaluated and showed normal urinary vanillylmandelic acid (VMA) levels. Also, abdominal ultrasonography did not reveal any adrenal mass. With time, significant neurological improvement was observed in both the lower limbs and vision also improved.
D I S C U S S I O N

Tumors of the central nervous system are common in the pediatric population. However, spinal tumors constitute only 1%–10% of all pediatric central nervous system tumors.[11,12]

Among these spinal neoplasms, schwannomas are benign tumors originating from Schwann cells, comprising approximately 30% of primary intraspinal neoplasms in the general population.[13] Schwannomas are typically seen in adults between 40 and 60 years of age and are rare in children. In a large referral center, only 0.7% of all schwannomas occurred in children during a 10-year period.[14] Age of our patient was 11 year.[3] But in our case, the patient was an 11-year-old female child.

The frequency of motor involvement and localized pain in the available literature ranged from 40% to 60%.[11] The occurrence of autonomic (bowel and bladder) involvement is found in 10%–20% cases in the literature.[11] In our case, although the patient initially presented with cranial symptoms, later, she developed spinal signs of motor weakness, paraplegia followed by retention of urine, which are in accordance with the findings in the available literature. However, our patient did not give any history of backache or spinal tenderness.

As per literature, the majority of the tumors are located in the cervical and lumbar regions.[3,15,16] In our patient, the tumor was localized at D12-L1 level and was intradural-extradural with the bulk of tumor lying anteriorly toward left side. According to the available literature, 70%–80% of spinal schwannomas are intradural in location and those extending through the dural aperture as the dumbbell mass involving both intradural as well as extradural space account for another 15%.[17] Intramedullary schwannomas are extremely rare.[18,19]

Most schwannomas are solid or mixed cystic–solid tumors and can rarely undergo cystic degeneration, xanthomatous change, or hemorrhage.[5,20] Nerve sheath tumors may also cause SAH, but it is exceedingly rare for these intradural lesions to come to clinical attention with only intracranial SAH without spinal symptoms.[6] Spontaneous SAH of spinal origin is uncommon and accounts for less than 1% of all cases of nontraumatic SAH.[21,22] Hence, our case is also among the rare cases of spinal schwannoma presenting with subarachnoid bleed. Some series on SAH have reported even lower incidence akin to Halpern et al.[23] who reported 0.6%

Figure 3: Postoperative CT brain (axial view): dilated ventricles suggestive of hydrocephalus

Figure 4: Postoperative MRI spine (T1-weighted sagittal view): no evidence of any residual lesion at operative site with insignificant postoperative changes
of their cases of SAH to have spinal origin, whereas Sahs et al.\cite{24} reported an even lower incidence of 0.05%. Going through literature, we could find first written account of spinal SAH by Michon\cite{25} which he likened to being stabbed in the spine (le coup de poignard rachidien). Very few cases presenting with predominant motor and sphincter deficits in the absence of pain have been reported.\cite{26} The more frequent causes of spinal SAH include trauma and vascular malformations.\cite{7,8,27,28} Among the spinal tumors, conus ependymoma was found in most of the cases of SAH.\cite{7,9} whereas for the nerve sheath tumors, it was exceedingly rare for them to come to clinical attention with only intracranial SAH without spinal symptoms. A thorough search of literature led us to only 22 cases of spinal schwannomas with SAH till now.\cite{6,29,30} In literature review that has the largest series on spinal schwannomas with SAH, Parmar et al.\cite{31} identified 20 reports of spinal nerve sheath tumors that caused SAH, of which only 8 cases (28%) presented exclusively with intracranial symptoms as in our patient.\cite{6,8,10,13,25} As hemorrhage is unusual in this tumor, various theories, including tumor location and histologic features, have been proposed to explain this phenomenon.\cite{5,8,23,27} First one is vascular theory according to which ectatic and hyalinized vessels of the tumor may undergo spontaneous thrombosis, followed by distal tumor necrosis and hemorrhage.\cite{23} Second is mechanical theory\cite{5,8,13,23} which suggests that hemorrhage into subarachnoid space occurs when there is trauma at the interface between the tumor and the normal neural tissue, particularly at the conus medullaris and the cauda equine regions.\cite{13,23} Moreover, traction on microvascular attachments to nerve roots may also occur, causing disruption of the blood vessels on the surface of the tumor.\cite{28} These tractional forces act mostly on the areas of high mobility and transition zones such as cervicothoracic and thoracolumbar junctions. This may explain why in many cases, bleed is related to exertion.\cite{23,25} Other causes of hemorrhage because of central ischemic necrosis associated with tumor growth\cite{26,27} or malignant transformation and neovascularization can further increase the susceptibility of such tumors to hemorrhage by accelerating the aforementioned process, especially within large schwannomas.\cite{19} In our case, no clear history of trauma or exertion before the onset of symptoms or pathologic evidence of malignant transformation was observed. We believe that the degenerative changes, along with the mechanical stress at the conus (D12-L1 level in our patient), may have caused rupture of the fragile, engorged tumor vessels and subsequent dissection of the blood into the subarachnoid space.

Schwannoma presenting in pediatric age group is often associated with neurofibromatosis (NF). Spinal nerve sheath tumors carry excellent prognosis in patients with NF1 and their recurrence rate is very low. On the contrary, symptomatic neurofibromas occurring in NF2 have more severe neurological deficit, poor postoperative recovery, and high recurrence rate of 10.7% at 5 years and 28.2% at 10 years, respectively.\cite{7} Our patient also had some features suggesting NF1-like presence of schwannoma, café au lait spots, and elevated urinary VMA levels, suggestive of pheochromocytoma, but no history suggestive of NF was observed in the family. Gene evaluation could not be performed as this facility was not yet available in our center. With recent technology, genetic proof is helpful in differentiating between NF1 (chromosome 17q) 8 and NF2 (chromosome 22q) 9, which is valuable for prognostication of disease.

**Conclusion**

Spinal schwannoma is rare among the pediatric population. Though histologically benign, they can produce profound neurological deficits. Microsurgical excision for spinal schwannoma usually results in good postoperative functional outcomes. In our clinical practice, we may sometimes come across some uncommon diseases with even more uncommon presentations as happened with us at our institute. We must always consider that there is a possibility of SAH owing to silent spinal lesion in patients with angiographic negative intracranial SAH as observed in this case.

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**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Hardison HH, Packer RJ, Rorke LB, Schut L, Sutton LN, Bruce DA. Outcome of children with primary intramedullary spinal cord tumors. Childs Nerv Syst 1987;3:89-92.
2. Stiller CA, Nectoux J. International incidence of childhood brain and spinal tumours. Int J Epidemiol 1994;23:458-64.
3. Nadkarni TD, Rekate HL. Pediatric intramedullary spinal cord tumors. Critical review of the literature. Childs Nerv Syst 1999;15:17-28.
4. Constantini S, Epstein F. Pediatric intraspinal tumors. In: Choux M, Di Rocco E, Hockley A, Walker M, editors. Pediatric neurosurgery. London, UK: Churchill Livingstone; 1999. pp. 601-2.
5. Parmar H, Patkar D, Gadani S, Shah J. Cystic lumbar nerve sheath tumours: MRI features in five patients. Australas Radiol 2001;45:123-7.
6. Parmar H, Pang BC, Lim CC, Chng SM, Tan KK. Spinal schwannoma with acute subarachnoid hemorrhage: a diagnostic challenge. AJNR Am J Neuroradiol 2004;25:846-50.

7. Cummings TM, Johnson MH. Neurofibroma manifested by spinal subarachnoid hemorrhage. AJR Am J Roentgenol 1994;162:959-60.

8. Saunders FW, Birchard D, Willmer J. Spinal artery aneurysm. Surg Neurol 1987;27:269-72.

9. Scotti G, Filizzolo F, Scialfa G, Tampieri D, Versari P. Repeated subarachnoid hemorrhages from a cervical meningioma. Case report. J Neurosurg 1987;66:779-81.

10. Cervoni L, Franco C, Celli P, Fortuna A. Spinal tumors and subarachnoid hemorrhage: pathogenetic and diagnostic aspects in 5 cases. Neurosurg Rev 1995;18:159-62.

11. Loh JK, Lin CK, Hwang YF, Hwang SL, Kwan AL, Howng SL. Primary spinal tumors in children. J Clin Neurosurg 2005;12:246-8.

12. Wilson PE, Oleszek JL, Clayton GH. Pediatric spinal cord tumors and masses. J Spinal Cord Med 2007;30:15-20.

13. Kim NR, Suh YL, Shin HH. Thoracic pediatric intramedullary schwannoma: report of a case. Pediatr Neurosurg 2009;45:396-401.

14. Tarik T. Schwannoma. In: Adesina AM, Tarik T, Fuller CE, Poussaint TY, editors. Atlas of pediatric brain tumors. New York: Springer; 2010. pp. 145-52.

15. Selosse P, Granieri U. [Spinal intradural meningiomas and neurinomas. Review of the literature and current situation]. Neurochirurgie 1968;14:135-54.

16. Seppälä MT, Haltia MJ, Sankila RJ, Jääskeläinen JE, Heikkanen O. Long-term outcome after removal of spinal schwannoma: a clinicopathological study of 187 cases. J Neurosurg 1995;83:621-6.

17. Jeon JH, Hwang HS, Jeong JH, Park SH, Moon JG, Kim CH. Spinal schwannoma; analysis of 40 cases. J Korean Neurosurg Soc 2008;43:135-8.

18. Conti P, Pansini G, Mouchaty H, Capuano C, Conti R. Spinal neurinomas: retrospective analysis and long-term outcome of 179 consecutively operated cases and review of the literature. Surg Neurol 2004;61:34-43; discussion 44.

19. De Verdelhan O, Haegelen C, Carsin-Nicol B, Riffaud L, Amlashi SF, Brassier G, et al. MR imaging features of spinal schwannomas and meningiomas. J Neuroradiol 2005;32:42-9.

20. Albert AF, Kirkman MA, du Plessis D, Sacho R, Cowie R, Tzerakis NG. Giant solitary cystic schwannoma of the cervical spine: a case report. Clin Neurosurg Neurosci 2012;14:396-8.

21. Grollmus J. Spinal subarachnoid hemorrhage with schwannoma. Acta Neurochir (Wien) 1975;31:253-6.

22. Kormos RL, Tucker WS, Bilbao JM, Gladstone RM, Bass AG. Subarachnoid hemorrhage due to a spinal cord hemangioblastoma: case report. Neurosurgery 1980;6:657-60.

23. Halpern L, Feldman S, Peyser E. Subarachnoid hemorrhage with papilledema due to spinal neurofibroma. AMA Arch Neurol Psychiatry 1958;79:138-41.

24. Sahs AL, Perret GE, Locksley HB, Nishioka H, editors. Intracranial aneurysm and subarachnoid hemorrhage. Philadelphia, PA: JB Lippincott; 1969.

25. Michon P. Le coup de poignard rachidien: symptome initial de certaines hemorragies sousarachnoidiennes. Essai sur les Hemorragies Meningees Spinales Presse Med 1928;36:964-6.

26. Campbell FG. Painless tumors of the cauda equina. A case report. Neurology 1963;13:341-3.

27. Cordan T, Bekar A, Yaman O, Toluay S. Spinal subarachnoid hemorrhage attributable to schwannoma of the cauda equina. Surg Neurol 1999;51:373-5.

28. Bruni P, Esposito S, Oddi G, Hernandez R, Martine A. Subarachnoid hemorrhage from multiple neurofibromas of the cauda equina: case report. Neurosurgery 1991;28:910-3.

29. Kukreja S, Ambekar S, Sharma M, Nanda AJ. Cauda equina schwannoma presenting with intratumoral hemorrhage and intracranial subarachnoid haemorrhage Case report. Neurosurg Spine 2014;13:1-4.

30. Ji C, Ahn JG, Huh HY, Park CK. Cervical Schwannoma presenting with acute intracranial subarachnoid haemorrhage. J Korean Neurosurg Soc 2010;47:137-9.