

Pediatric Intramedullary Schwannoma without Neurofibromatosis: Case Report

Brahim Eljebbouri, Miloudi Gazzaz, Ali Akhaddar,
Brahim Elmostarchid, and Mohamed Boucetta

Department of Neurosurgery, Med V University, Mohamed V Teaching Hospital, Rabat, Morocco

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Abstract- Pediatric intramedullary schwannoma without neurofibromatosis is extremely rare with only five cases reported so far. We present this rare finding in an 10-year-old boy who presented with a sudden onset of weakness in the lower limbs. An intraoperative diagnosis of schwannoma enabled us to carry out a total excision of the tumor, which resulted in near complete recovery at 10 months follow-up. Although rare, this diagnosis should be considered when a child presents with a solitary intramedullary tumor since its total resection can be achieved improving surgical outcome.

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Introduction

Schwannomas arise from nerve sheath Schwann cells in the spinal canal; thus, their location is usually intradural extramedullary and/or extradural (1). Intramedullary (IM) schwannomas are rare tumors, accounting for 0.3-1% of intraspinal neoplasms and 1.1% of spinal Schwannomas (1,2). Their rarity may be explained by the lack of Schwann cells in the spinal cord (1-5). It has been postulated that the small perivascular bundles of peripheral nerves which normally occur within the spinal cord are the logical source of these tumors (1,2). Other sources suggested are dorsal or ventral nerve roots with an intraspinal extension; from neural crest cells that may be displaced into the parenchyma of the central nervous system during embryonic development; or from pial cells that may have undergone metaplasia into Schwann cells (2,4).

Preoperative diagnosis of IM schwannoma can be suspected in a patient with associated neurofibromatosis (NF), an entity frequently related to this lesion (3,8). In about one-fifth of the cases of NF, an IM lesion has been documented, prevalently schwannomas (8). On review of the literature, only five cases of pediatric IM schwannoma in the absence of signs of NF have been reported (3,8).

Case Report

A 10-year-old boy presented with a sudden onset of weakness of the lower limbs associated with bladder and bowel incontinence. Preceding this episode there was a six-month history of frequent falls, which was disregarded as clumsiness by parents. General examination was normal, with no signs of neurofibromatosis. Neurological examination revealed the strength of Grade 1-2 in the lower limbs. Deep tendon reflexes were accentuated in both lower limbs with bilateral up-going plantar reflex. All modalities of sensation below D8 were decreased bilaterally with associated saddle anesthesia.

MRI examination demonstrated an IM lesion from the upper border of D7 to the lower border of D9 with cord edema (Figures 1 and 2). A presumptive diagnosis of IM astrocytoma (commonest IM lesion in this age group) was considered.

The patient underwent a D7-9 laminectomy. Opening the dura mater revealed a well-demarcated, soft, greyish-pink tumor. Surprisingly, the imprint cytology from the tumor was suggestive of schwannoma (at that time, facilities of frozen section were not available at our center). Total excision of the lesion was performed with the help of microsurgical techniques taking care to avoid damage to the adjoining normal

Corresponding Author: Brahim Eljebbouri

Department of Neurosurgery, Mohammed V Military Teaching Hospital, University of Mohammed V Souissi, Bloc V2, Appt 5, Avenue Kamal Zebdi, Secteur 21, Hay Riyad 10100, Rabat, Morocco

Tel: +21 261 9310303, E-mail: brahimjebbouri@gmail.com

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neural tissue. This diagnosis was later confirmed on histopathological examination which showed proliferation of nerve fibers with a mixture of dense cellular Antoni A and hypocellular Antoni B areas, consistent with the diagnosis of schwannoma (Figure 3). The patient made an uneventful post-op recovery. Postoperative MR confirmed complete tumor excision.

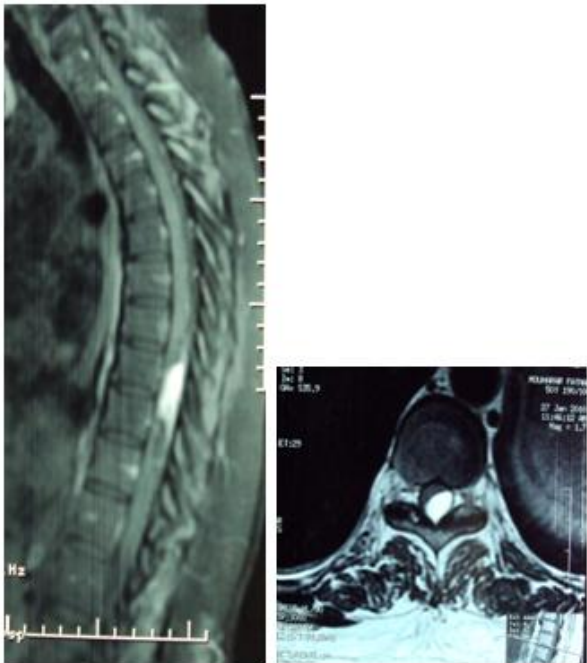


Figure 1. MRI T1 injected weighted into sagittal and axial sections showing intradural heterogeneous intramedullary process facing D8 repressing spinal cord forward and right.



Figure 2. MRI T2-weighted into sagittal and axial sections showing a heterogeneous process in hypersignal and partly cystic intramedullary location.

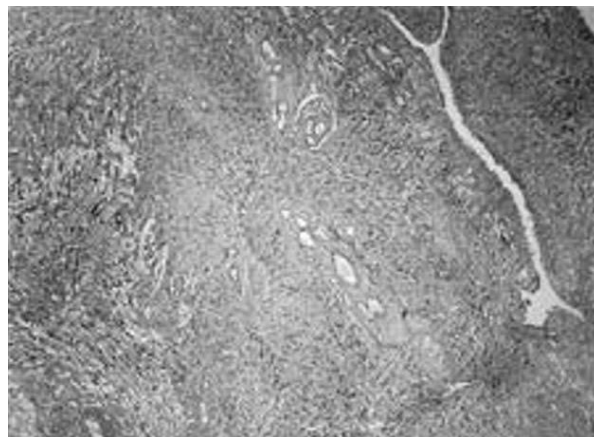


Figure 3. Histological aspect of the intramedullary schwannoma.



Figure 4. Spinal MRI made 15 months after surgery shows no tumor recurrence.

At 15 months follow-up, he has made a remarkable neurological recovery. Sensations and strength in all limbs are normal; however, deep tendon reflexes in the right lower limb are still exaggerated. His bladder and bowel functions are normal. Follow-up MRI at 18 months shows the patient to be free of recurrence of the lesion (Figure 4).

Discussion

Intramedullary spinal cord tumors in children account for 4-10% of central nervous system tumors (6,7). In the differential diagnosis, one typically considers astrocytomas which represent 60% of childhood IM tumors, followed by ependymomas (30%) (7). Intramedullary schwannomas are extremely rare tumors and are usually associated with neurofibromatosis (NF 1

and 2) (2). Neurofibromatosis¹ generally presents with schwannomas along nerves in the skin, brain and other parts of the body, whereas, in NF2, schwannomas develop along nerves in the inner ear and brain.

Intramedullary schwannomas are mostly located in the cervical spinal cord (51%), followed by the thoracic region (22%) (1,2). They may remain asymptomatic for a long time or cause nonspecific complaints, which makes the diagnosis difficult in children (6). The most common symptom is pain (6). Sometimes, they present with motor deficits, which can progress to the clinical syndrome highlighted by Lesoin, which includes motor deficiency, reduced sensation and loss of genitourinary function (5), as seen in our patient. On magnetic resonance imaging (MRI), these tumors are well delineated with moderate peri-lesional edema (8) and no syringomyelia. Administration of Gadolinium (Gd) enhances the thickened spinal nerve root in continuity with the tumor which helps to differentiate these tumors from other intramedullary lesions (1) and also allows better definition of the tumor from associated edema (1,2,8). In our patient, Gd-enhanced MRI was not available, making differentiation between tumor and edema difficult. This, along with the clinical urgency of the case, led us to perform an extended laminectomy instead of laminoplasty.

Total surgical removal should be attempted because of the benign nature of Schwannomas (4,5). However, due to rarity, they may be difficult to differentiate from gliomas preoperatively or during surgical exploration (2,8). Since complete surgical removal is not attempted in infiltrating gliomas (too much aggressive approach may add deficit without any benefit), correct pre- or intraoperative diagnosis is crucial (2,8). Our case further highlights this fact, as an intraoperative diagnosis of schwannoma enabled us to do a complete surgical removal that resulted in good recovery with noticeable improvement of his neurological symptoms.

To conclude, pediatric intramedullary schwannoma

without neurofibromatosis is extremely rare. Nevertheless, it must be kept in mind when a child presents with a solitary intramedullary tumor since its total resection can be achieved, improving surgical outcome.

References

1. Colosimo C, Cerase A, Denaro L, Maira G, Greco R. Magnetic resonance imaging of intramedullary spinal cord schwannomas. Report of two cases and review of the literature. *J Neurosurg* 2003;99(1 Suppl):114-7. Kodama Y, Terae S, Hida K, Chu BC, Kaneko K, Miyasaka K. Intramedullary schwannoma of the spinal cord: Report of two cases. *Neuroradiology* 2001;43(7):567-71.
2. Kodama Y, Terae S, Hida K, Chu BC, Kaneko K, Miyasaka K. Intramedullary schwannoma of the spinal cord: Report of two cases. *Neuroradiology* 2001;43(7):567-71.
3. Binatli O, Ersahin Y, Korkmaz O, Bayol U. Intramedullary schwannoma of the spinal cord. A case report and review of the literature. *J Neurosurg Sci* 1999;43(2):163-8.
4. 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(1):34-44.
5. Ogungbo BI, Strachan RD, Bradey N. Cervical intramedullary schwannoma: Complete excision using the KTP laser. *Br J Neurosurg* 2000;14(4):345-8.
6. Jallo GI, Freed D, Epstein F. Intramedullary spinal cord tumors in children. *Childs Nerv Syst* 2003;19(9):641-9.
7. Darwish BS, Balakrishnan V, Maitra R. Intramedullary ancient schwannoma of the cervical spinal cord: Case report and review of literature. *J Clin Neurosci* 2002;9(3):321-3.
8. Nadkarni TD, Rekate HL. Pediatric intramedullary spinal cord tumors. Critical review of the literature. *Childs Nerv Syst*;1999;15(1):17-28.