Single root multiple spinal schwannomas: Case report, treatment strategy and review of literature

Augustinas Fedaravičius a, d, 1, Avner Michaeli b, 1, Victor Diomin c, Tehila Kaisman Elbaz a, Arimantas Tamašauskas d, Israel Melamed a, ∗

a Department of Neurosurgery, Soroka University Medical Center, Be’er Sheva, Israel
b Surgical Monitoring Services, Israel
c Institute of Pathology, Soroka University Medical Center, Be’er Sheva, Israel
d Department of Neurosurgery, the Hospital of Lithuanian University of Health Sciences, Lithuania

ABSTRACT

INTRODUCTION: Schwannomatosis is defined as multiple schwannomas without presence of neurofibromatosis and is a rare pathology. In vast majority of cases the schwannomas grow from different nerve roots or peripheral nerves.

PRESENTATION OF CASE: A 52-year-old woman presented with multiple intradural schwannomas arranged in a chain along the spinal canal causing significant compression. The lesions were successfully removed using a left side en-bloc hemilaminectomy technique in order to preserve maximal stability of the posterior column. Back and leg pain resolved completely. Tendon reflexes returned to normal shortly. There was decreased pain sensation in the distribution of the left L3 spinal root.

DISCUSSION: The traditional surgical strategy for posterior approach by laminectomy or laminotomy is sometimes complicated with instability or deformation of the vertebral column that requires surgical stabilization. We performed a one side en-bloc hemilaminectomy thus maintaining the integrity of the muscles and ligaments on the opposite side and preserving maximal stability of the vertebral column. Densely adherent tumors required careful sharp dissection and separation under neurosurgical monitoring and stimulation for recognition and preservation of spinal roots. An additional tumor was discovered by exploration of the spinal canal using an endoscope.

CONCLUSION: Multiple spinal cord schwannomas that are growing along the same part of the vertebral column can be safely removed by one-sided hemilaminectomy with preservation of the integrity of the muscles and ligaments on the opposite side and thus maintain spinal stability. The 3D endoscope can be a good tool for visual exploration of the spinal canal.

© 2020 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

1. Introduction

Spinal schwannomas or neurinomas are the most common benign, slow growing nerve sheath tumors [1–13]. Usually multiple spinal schwannomas are associated with neurofibromatosis type 1 (NF1 - von Recklinghausen’s disease) or type 2 (NF2) [1–13]. In the last three decades significant number of cases of multiple schwannomas have been described that were not associated with any characteristic features of neurofibromatosis [1–13]. This group of patients with multiple schwannomas in the absence of typical signs of NF1 and NF2 syndromes was defined as schwannomatosis [1–13]. Our literature analysis revealed a case of multiple schwannomas in a single peripheral (sciatic) nerve with family history of schwannomas, but we didn’t find a note in described cases on a single spinal nerve root as a source of multiple spinal schwannomas.

We present a rare case of spinal schwannomatosis in which the multiple schwannomas have grown from a single spinal root. This work has been reported in line with the SCARE criteria [15].

2. Case report

A 52-year-old woman with a 3 year history of low back and leg pain treated in the pain clinic presented with 2 months of worsening pain especially when in a supine position as well as neurologic claudication. The pain was so severe that she was unable to sleep. There was a slight relief in the pain when the patient took a sitting position. Furthermore, the patient experienced frequent falls due to sudden onset leg weakness.

Neurological examination revealed normal muscle tone and normal power in all muscle groups in both legs. The patella and Achilles tendon reflexes were symmetrically decreased. Pain and
Fig. 1. 1– MRI images demonstrated multiple schwannomas at lumbar levels. 2 – left side hemilaminectomy without dissection and maintaining the integrity of the muscles and ligaments on the opposite side. 3 – the schwannomas hidden anterior to the cauda equina nerve roots. 4 & 5 – tEMG for preservation of motor nerve roots.

Fig. 2. 1 – Endoscopic view of the up-seated schwannoma with surrounding structures from inside of spinal canal. 2 – Neurophysiological guidance by spinal roots stimulation. 3 – Sharp dissection of the motor root from schwannoma by diamond knife. 4 – Coagulation of the sensory part of the left L3 spinal root before resection. 5 – Decompressed cauda equina spinal roots after complete removal of the schwannomas.

deep sensation was preserved. The Lasegue's signs were positive at an angle 60 degrees on both sides. Sphincter tone was normal.

A lumbar spine MRI revealed the multiple intradural, different sized lesions hypointense on T2 and T1 at L1–L3 levels with significant post-contrast homogenous enhancement. The lesions were arranged in a chain along the spinal canal and caused a shift of the conus and the cauda equina roots to the right with significant compression of the roots at the L3 level (see Fig. 1). Brain, Cervical and Thoracic Spine MRI were unremarkable.

The patient was not diagnosed with neurofibromatosis type 1 or 2, and had no relevant drug and family history.

Multiple schwannomas were successfully removed using a left side en bloc hemilaminectomy with a laminotome (ANSPACH) for exposure, in order to preserve maximal stability of the posterior column. The surgery was performed by a senior neurosurgeon. The schwannomas were hidden anterior to the conus and the cauda equina nerve roots (Fig. 1). During careful dissection and removal of the tumors under neurophysiological guidance for preservation...
of motor nerve roots, it was found that all schwannomas grew from the sensory part of the same spinal root (Fig. 2). Tumors (in the amount of 6) that were compressing the conus and were densely adherent to the cauda equina roots, were gross total removed (Fig. 2). After removal of the biggest tumor and several small tumors beneath it, the spinal canal was explored using a 30° endoscope (AESCU LAP) and an additional small tumor was discovered cranially relative to the largest tumor (Fig. 21). The source of the schwannomas was the sensory part of the left L3 spinal root. This root, crumpled and stratified into fibers by schwannomas, was sacrificed and excised with the tumors.

The postoperative course was uneventful. The back and leg pain completely resolved. The tendon reflexes returned to normal shortly. There was only depressed pain sensation in the distribution of the left L3 spinal root.

Pathology study (Fig. 3) demonstrated focally cellular tumors and shown separate well circumscribed nodules with compact (Antony A) and loosely textured (Antony B) areas, focal scant lymphocytic inflammatory infiltration, hyalinized blood vessels. The lesions were surrounded by thin fibrous capsule, shown focally increased mitotic activity (1 M F/10 HPF) with no significant nuclear size variability, nuclear hyperchromasia or necrosis and, therefore were classified as cellular schwannomas. Immunostaining for S100 markers was positive (cytoplasmatic and nuclear stain); EMA and PR immunostains were negative.

3. Discussion

Multiple spinal schwannomas not associated with neurofibromatosis are rare [1,3–8,12,13]. The schwannomas can be located in different parts of spinal column as separate lesions [3,4,12], or could present as multiple lesions clustered in the same part of the spine [1,2,4,6,9,10]. In vast majorities of cases described previously in the literature, the sources of schwannomas were different spinal roots [1,3–8,12,13] and multiple schwannomas that grew from one single root were present only in peripheral nerves [10]. In contrast, in our case all schwannomas grew from the same spinal root along the three levels of lumbar spine canal L1-L3. Traditionally the posterior approach by laminectomy or laminotomy is preferred for surgical resection of schwannomas [2,3,6,12,13]. But this surgical strategy is in some cases complicated with instability or deformation of the vertebral column that requires surgical stabilization [13,14]. In the presented case we performed for exposition of the spinal canal a one side en bloc hemilaminectomy technique without dissection and maintaining the integrity of the muscles and ligaments on the opposite side in order to preserve maximal stability of the vertebral column. Tumors densely adherent to the cauda equina roots required careful sharp dissection or separation by peel away techniques under neurosurgical monitoring and repeated stimulation for recognition and preservation of spinal roots. The endoscope is a valuable tool for exploration of the spinal canal, in our case it facilitated the discovery of an additional tumor.

4. Conclusion

Multiple spinal cord schwannomas that are growing along the same part of the vertebral column can be safely removed by one side hemilaminectomy with preservation of the integrity of the muscles and ligaments on the opposite side and thus maintain spinal stability. Neuroradiology is mandatory for the surgical procedure to allow successful results and to prevent any neurological damage.

Fig. 3. 1 – H&E stain shows compact (Antony A) in the right and loosely textured (Antony B) areas in the left. 2 – Different nodule – well-formed fibrous capsule and hyalinized blood vessels, H&E, *200. 3 – S100 immunostaining – cytoplasmatic and nuclear stain, *400. 4 – Fibrous capsule collagen stains blue with Masson trichrome special stain (left, *200).
The 30° endoscope can be a good tool for visual exploration of the spinal canal.

**Conflicts of interest**

No conflicts.

**Sources of funding**

No sponsors.

**Ethical approval**

This case report presentation is exempt from ethical approval.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

**Author contribution**

**Fedaravičius**: Study conception, data collection, data analysis, manuscript writing/editing, final approval of manuscript.

**Michaeli**: Study conception, data collection, data analysis, manuscript writing/editing, final approval of manuscript.

**Dionin**: Data collection, data analysis, manuscript editing, final approval of manuscript.

**Kaisman Elbaz**: Data collection, data analysis, manuscript editing, final approval of manuscript.

**Tamašauskas**: Data collection, data analysis, manuscript editing, final approval of manuscript.

**Melamed**: Study conception, data collection, data analysis, manuscript writing/editing, final approval of manuscript.

**Registration of research studies**

Presented case report is not considered a research study.

**Guarantor**

Israel Melamed.

**Provenance and peer review**

Not commissioned, externally peer-reviewed.

**References**

[1] M. Daras, B.S. Koppel, C.W. Heise, M.J. Mazzeo, T.P. Poon, K.R. Duffy, Multiple spinal intradural schwannomas in the absence of Von Recklinghausen’s disease, Spine (Phila Pa 1976) 18 (December (16)) (1993) 2556–2559.

[2] S. Narayan, N. Jain, H.A. Patil, Rare case of spinal schwannomatosis presenting as conus-cauda syndrome, J. Spinal Surg. 3 (July–September (3)) (2016) 108–111.

[3] M. Seppala, M. Halita, R. Sankila, J. Jaaskelainen, O. Heiskanen, Long-term outcome after removal of spinal schwannoma: a clinicopathological study of 187 cases, J. Neurosurg. 83 (1995) 621–626.

[4] S.W. Kim, S.M. Lee, Multicentric spinal schwannomas without any evidence of neurofibromatosis Type2, J. Korean Neurosurg. Soc. 36 (2004) 508–509.

[5] A. Landi, D.E. Dugoni, N. Marotta, C. Mancarella, R. Delfini, Spinal schwannomatosis in the absence of neurofibromatosis: a very rare condition, Int. J. Surg. Case Rep. 2 (2011) 36–39.

[6] S.H. Lee, S.H. Kim, B.J. Kim, D.J. Lim, Multiple schwannomas of the spine: review of the schwannomatosis or congenital neurilemmomatosis; a case report, Korean J. Spine 12 (2) (2015) 91–94.

[7] C. Kayaoglu, G. Sengul, I. Aydin, Multiple schwannomas of cauda equina in the absence of Von Recklinghausen’s disease, Audi. Med. J. 29 (12) (2007) 1907–1909.

[8] M. Seppala, M. Sainio, M. Halita, J. Kimmunen, K. Setala, J. Jaaskelainen, Multiple schwannomas: schwannomatosis or neurofibromatosis type 2? J. Neurosurg. 89 (1998) 36–41.

[9] M. Altsas, J. Aylan, G. Silav, R. Sarsib, K. Coskun, N. Balak, N. Isik, I. Elmaci, Microsurgical management of non-neurofibromatosis spinal schwanna, Neurocirugia 24 (6) (2013) 244–249.

[10] N. Satoh, Y. Ueda, M. Koizumi, T. Takehima, J. Iida, K. Shigematsu, H. Shigematsu, H. Matsumori, Y. Tanaka, Assessment of pure single nerve root resection in the treatment of spinal schwannoma: focus on solitary spinal schwannomas located below the thoracolumbar junction, J. Orthop. Sci. 16 (2011) 148–155.

[11] V. Javalkar, T. Pigott, P. Pale, G. Findlay, Multiple schwannomas: report of two cases, Eur. Spine J. 16 (Suppl. 3) (2007) S287–S292.

[12] A. Ogose, T. Hotta, T. Morita, H. Otuka, Y. Hirata, Multiple schwannomas in the peripheral Nerves, J. Bone Jt. Surg. 80-B (July (4)) (1998) 657–661.

[13] J. Lenzi, G. Anichini, A. Landi, A. Picciochi, E. Passacantilli, F. Pedace, R. Delfini, A. Santoro, Spinal nerves schwannomas: experience on 367 cases—historic overview on how clinical, radiological, and surgical practices have changed over a course of 60 years, Neurol. Res. Int. 2017 (2017) 1–12.

[14] A. Inoue, T. Ikata, S. Kato, Spinal deformity following surgery for spinal cord tumors and tumorous lesions: analysis based on an assessment of the spinal functional curve, Spinal Cord 34 (1996) 536–542.

[15] B.A. Agha, M.R. Borrelli, R. Farwana, K. Koshy, A. Fowler, D.P. Orgill, For the SCARE Group, The SCARE 2018 statement: updating consensus surgical Case Report (SCARE) guidelines, Int. J. Surg. 60 (2018) 132–136.