A CASE REPORT: MULTIPLE SCHWANNOMAS OF THE SPINE

Nguyen Xuan Phuong¹, Nguyen Thanh Bac^{1*}

Abstract

Neurinoma is the most common benign nerve sheath tumor originating from Schwann cells. Most benign neurinomas are single. Patients with multiple tumors can be seen in conditions such as neurofibromatosis (NF) type 2, whose typical sign is bilateral vestibular schwannomas. Some authors have described it as a form of schwannoma, which presents multiple schwannomas without associating with NF factors. Studies have recognized this condition as the third major form of NF [1, 2, 3]. We describe a case of multiple schwannomas in the spine associated with schwannomas by imaging and surgical features to clarify the characteristics of this disease.

Keywords: Multiple schwannomas; Microsurgery.

INTRODUCTION

Neurinoma, originating from Schwan cells and occurring in the spine, has an incidence of 0.3 - 0.5/100,000 people contracting annually. The disease incidence in men and women is the same. Schwannomas are usually detected at 40 - 50 years of age, and the most common location is in the lumbar region and cauda equina of the spinal cord. Tumors are usually benign and round in shape with clear boundaries [1, 2]. Clinical manifestations are discreet;

magnetic resonance imaging enables the detection of the disease and the making of appropriate treatment plans.

CASE REPORT

A 66-year-old female patient with a history of dull, vague back, and leg pain for 3 months was treated as an outpatient and admitted to the hospital with pain that gradually worsened over the past 1 month, especially when in a supine position and having intermittent claudication. The pain gradually increases,

*Corresponding author: Nguyen Thanh Bac (bacnt103@gmail.com)

Date received: 10/01/2024

Date accepted: 18/3/2024

¹Department of Neurosurgery, Military Hospital 103

http://doi.org/10.56535/jmpm.v49i4.684

making it impossible for the patient to sleep. It can be relieved when in a sitting position, and the patient sometimes falls due to sudden leg weakness.

Clinical examination showed normal muscle tone and strength in all muscle groups of both legs. Patellar and Achilles tendon reflexes reduced symmetrically. There were no abnormalities in pain or sensation of depth. There was a positive Lasegue sign at 60 degrees on both sides. There was no sphincter disorder. The patient did not have any lumps under the skin.

Magnetic resonance imaging of the lumbar spine showed multiple intradural lesions of varying sizes, hypointensity on T2 and T1 at the D12 - L3 level, and relatively homogeneous and hyperintensity after injecting contrast agent. The lesions were arranged in a series along the spinal canal and caused compression of the conus and cauda equina to the right, with the most significant compression of the roots at the L1 level (*Figure 1*). MRI scans of the brain, cervical spine, and thoracic spine did not detect any abnormal mass.

The patient was not diagnosed with NF type 1 or 2 and had no relevant family or medical history.

The tumors were removed by hemilaminectomy and microsurgical

excision of the tumor. Schwannomas hid anterior to the conus and cauda equina nerve roots (*Figure 1*). During surgery, tumors were dissected and removed with the help of microneurosurgery instruments to preserve the function of the nerve roots. Five tumors that pressed the cone and adhered to the cauda equina roots were completely removed (*Figure 3*).

The postoperative progress was stable. Symptoms of back pain and leg pain were completely resolved. Tendon and bone reflexes returned to normal in a short time. The patient only had a slight decrease in pain sensation in the distribution area of the left L1 root.

Histopathology (Figure 2) demonstrated focal cellular tumors and showed wellcircumscribed discrete nodules with compact (A) and loosely textured (B) infiltrative areas, minor focal lymphocytic inflammation, and hyalinized blood vessels. The lesions were surrounded by a thin fibrous capsule, showed markedly increased mitotic activity (1 M F/10 HPF) without significant nuclear size change, nuclear hyperchromia, or necrosis, and were classified as schwannoma. Immunoassay was positive for the S100 marker (cytoplasmic and nuclear staining) and negative for EMA and PR.



Figure 1. Tumors on magnetic resonance imaging.



Figure 2. Histopathology of schwannom, ×100, stained Hematoxilin & Eosin (H&E) (A). Typical Verocay type ×400, H&E stain (B).



Figure 3. Five tumors were removed and a postoperative MRI of the spine.

DISCUSSION

Neurinoma is a common condition, often associated with NF1 or NF2. whereas multiple Swchanomas are rarer and may not be associated with these factors. [1, 2, 4, 5]. Symptoms of the disease often manifest as dull lumbar spine pain and vague pain, gradually increasing over time, and despite aggressive multimodal medical treatment, most patients do not have pain relief. There are 62% of patients who do not have pain relief after intensive medical treatment [6]. Depending on the location of the injury and the degree of nerve root compression, sensory disorders appear in the corresponding dominant area. Symptoms of sensory disorders often occur first due to tumors growing from sensory roots.

According to the classification of Ibrahim S (2017), neurinomas are divided into 4 types based on the location of the tumor with the dura mater and spinal canal. The tumors in our study are type I which are localized exclusively intradurally. This is the most common type, accounting for 51%. Symptoms do not improve much after surgery for type I and IV patients. However, in our case, the patient's pain symptoms were almost completely relieved [7, 8]. Neurinomas are removed by posterior hemisection and microsurgery. In cases of spinal anesthesia, neuromuscular electrical monitoring can be performed during surgery to avoid damage to functional roots and nerves. The 30-degree endoscope can be used in combination to visually explore the spinal canal, avoiding missed lesions [9]. In our patient, the largest tumor was near the foramen, so we used an endoscope during surgery to observe the foramen to remove the tumor thoroughly and avoid missing it.

Neurinomas are usually benign and can recur. According to author Gerganov, the recurrence rate of neurofibromas is generally about 0.5 - 5%, and longterm postoperative monitoring with imaging diagnostics is necessary to detect tumor recurrence [10]. Our patient was examined with magnetic resonance imaging 6 months after surgery, and no tumor recurrence was detected.

CONCLUSION

Multiple neurinomas develop along the nerve roots. Therefore, the spinal cord must be examined with an MRI before surgery. Radical tumor removal surgery preserves nerve function and brings good long-term results to the patient. As the tumor has the ability to recur, long-term monitoring and examination are required after surgery. Acknowledgments: We would like to thank Military Hospital 103 for its professional support. We claim our research is conducted in an absolutely objective manner and have no conflicts of interest.

REFERENCES

1. Chen SL, Liu C, Liu B, et al. Schwannomatosis: A new member of neurofibromatosis family. *Chin Med J (Engl)*. 2013; 126(14):2656-2660.

2. Seppälä MT, Sainio MA, Haltia MJ, et al. Multiple schwannomas: schwannomatosis or neurofibromatosis type 2? *J Neurosurg*. 1998; 89(1):36-41.

3. Gonzalvo A, Fowler A, Cook RJ, et al. Schwannomatosis, sporadic schwannomatosis, and familial schwannomatosis: A surgical series with long-term follow-up. *Clinical article*. *J Neurosurg*. Mar 2011; 114(3):756-62.

4. Koontz NA, Wiens AL, Agarwal A, et al. Schwannomatosis: The overlooked neurofibromatosis? *AJR Am J Roentgenol*. 2013; 200(6):646-653.

5. Qin S, Yang C, Ren H. Cerebral hernia caused by a thoracic surgery for multiple schwannomas in a patient with Neurofibromatosis type 2. *Turk Neurosurg*. 2013; 23(2):245-248.

6. Merker VL, Esparza S, Smith MJ, et al. Clinical features of schwannomatosis: A retrospective analysis of 87 patients. *Oncologist*. 2012; 17(10): 1317-1322.

7. Ioannidis P, Mamouli D, Foroglou N. Expanding schwannomatosis phenotype. *J Neurooncol*. 2015; 122(3):607-609.

8. Sun I, Pamir MN. Non-Syndromic Spinal Schwannomas: A novel classification. *Front Neurol*. 2017; 8:318.

9. Fedaravičius A, Michaeli A, Diomin V, et al. Single root multiple spinal schwannomas: Case report, treatment strategy and review of literature. *Int J Surg Case Rep.* 2020; 74:113-116.

10. Gerganov V, Petrov M, Sakelarova T. Schwannomas of Brain and Spinal Cord. *Adv Exp Med Biol.* 2023; 1405:331-362.