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CASE REPORT

Intramedullary Schwannomas: A Rare Case Report*

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ABSTRACT

This case report aims to review a very rare case of intramedullary schwannoma with literature review. A 52-year-old male patient presented with complaints of difficulty walking and leg spasms. Radiological examinations revealed an intradural lesion at the C7-T3 level, and gross total resection was performed. No postoperative neurological deterioration was observed. The patient's preoperative ASIA and McCormick scores were recorded as C and 3, respectively; while they were recorded as 6th months and 1st year D, E and 2, 1 postoperatively, respectively. Histopathological examination reported schwannoma. Our patient has been under outpatient clinic follow-up for four years. Intramedullary schwannoma is a rare spinal cord tumor with 198 cases reported in the literature. The exact etiopathogenesis of this rare condition is still under debate. Due to the low recurrence rate, microsurgical gross total excision is the gold standard in the treatment of intramedullary schwannomas.

Keywords: Schwannoma. Intramedullary tumor. Spinal cord. Intradural tumor.

İntramedüller Schwannoma: Nadir Olgu Sunumu

ÖZET

Bu olgu sunumu, oldukça nadir görülen intramedüller yerleşimli schwannoma olgusunun literatür eşliğinde incelenmesini amaçlamaktadır. 52 yaşında erkek hasta, yürüme güçlüğü ve bacaklarda kasılma şikayetiyle başvurdu. Radyolojik incelemelerde C7-T3 seviyesinde intradural yerleşimli kitlesel lezyon saptanması üzerine gross total rezeksiyon yapıldı. Postoperatif nörolojik kötüleşme gözlenmedi. Hastanın ameliyat öncesi ASIA ve McCormick skorları sırasıyla C ve 3, ameliyat sonrası ise sırasıyla 6. ay ve 1. yılda D, E ve 2, 1 olarak kaydedildi. Histopatolojik inceleme, schwannoma olarak rapor edildi. Hastamız 4 yıldır poliklinik takibimizdedir. İntramedüller schwannoma, literatürde 198 vaka bildirilen nadir bir omurilik tümörüdür. Bu nadir durumun kesin etyopatogenezi konusunda tartışmalar devam etmektedir. Düşük nüks oranı nedeniyle intramedüller schwannomaların tedavisinde altın standart mikroşirurjikal gross total eksizyondur.

Anahtar Kelimeler: Schwannoma. İntramedüller tümör. Spinal kord. İntradural tümör.

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Primary spinal cord tumors constitute approximately 15% of central nervous system tumors[1]. Spinal tumors are categorized based on their location in three main groups: extradural, intradural extramedullary, and intramedullary. Schwannomas are primary nerve sheath tumors with intradural extramedullary localization. Intramedullary schwannomas (IMS) are extremely rare, with only 198 reported cases in the literature[1-6]. Among these cases, 13 cases were reported with special pathological subtypes (intramedullary melanocytic schwannoma) and very rare localizations, including 6 cases at the cervicomedullary junction and 13 cases at the conus[1,3,6-9].

Our study aims to conduct a retrospective examination and compare our case with the existing literature on IMS. By collecting data, reviewing and analyzing the literature in the PubMed database, we aimed to examine a very rare case of intramedullary

schwannoma in the light of the literature. By doing so, we hope to shed more light on this uncommon condition and contribute to the collective knowledge in this area.

Case Report

A 52-year-old male patient presented to our outpatient clinic with complaints of difficulty in walking and spasms in the legs. Neurological examination revealed spastic paraparesis. The patient's ASIA score was classified as C, indicating motor incomplete impairment and the McCormick score indicated moderate disability[10,11]. Radiological studies showed the presence of intramedullary lesions at the C7-T3 level; spinal magnetic resonance imaging (MRI) shows heterogeneous and hyperintense on T2-weighted images and hypointense on T1-weighted images, with heterogeneous contrast enhancement (Figure 1 A-E). The lesion was radiologically evaluated as intradural extramedullary. However, our intraoperative findings indicated that the tumor was not purely extramedullary. It was seen to be an intramedullary tumor with a subpial location originating from the T2 dorsal root with exophytic component. Intraoperative neuromonitoring (ION) was not used in the case. Microsurgical resection was performed on the patient, and as a result, the hemorrhagic, purple tumor tissue was dissected from the pial-medullary adhesions and the root from which it originated was cut macroscopic complete resection of the mass was achieved (Figure 2 A-C). Histopathological examination revealed a well-circumscribed tumoral lesion with regions of high and low cellularity, varying cellular densities, and randomly distributed cells with oval and spindle nuclei. S100 immunohistochemical staining was used to identify diffuse strong positivity in the immunohistochemical evaluation (Figure 3 A-C). The preoperative and postoperative ASIA scores remained at level C, and McCormick scores were 3. After a compatible rehabilitation process at the 6th month and 1st year follow-up, the patient's ASIA and McCormick scores became D and E, 2 and 1, respectively. No additional adjuvant treatment was administered to our patient. Our patient has been under outpatient clinic follow-up for four years.

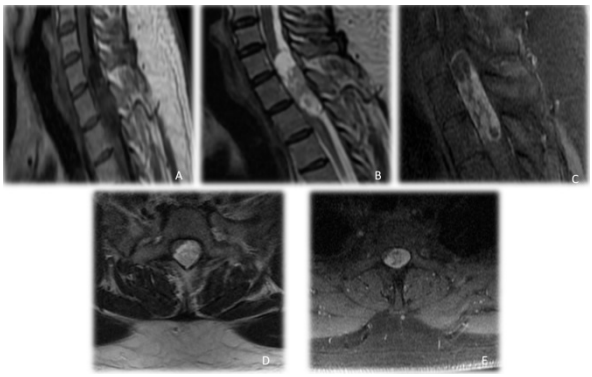

Figure 1.
Preoperative T1-weighted sagittal image (A), T2-weighted sagittal image (B), and T1-weighted contrast-enhanced sagittal image (C) of the patient; T2-weighted axial image (D) and T1-weighted contrast-enhanced axial image (E) at the cervicothoracic level. The lesion contains a cystic component, appears as an intradural, extramedullary mass pushing the cord laterally and stains heterogeneously in post-contrast images.

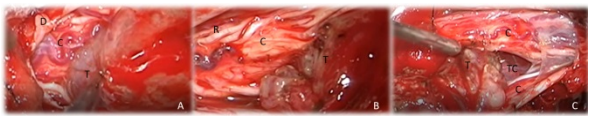

Figure 2.
Intraoperative microscopic images (A, B, C) are shown. We observed that the tumor, which was thought to be intradural extramedullary radiologically, showed subpial intramedullary invasion intraoperatively, originating from the left T2 dorsal root. After severing the dorsal root connection and opening the pial-medullary adhesions, macroscopic complete resection of the mass was achieved. D; Duramater, C; Cord, T; Tumor, R; Roots, TC; Tumoral Cavity

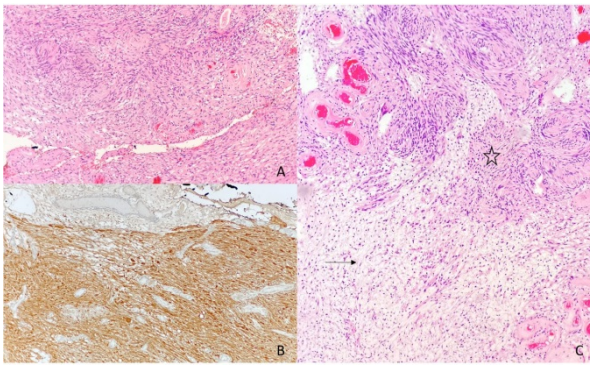

Figure 3.
Photomicrograph illustrating (A) tumor cells with narrow, spindle-shaped and wavy, monotonous appearance, without cytological atypia (Hematoxylin and eosin, x 100), (B) diffuse, strong S100 immunohistochemical staining (x100); and (C) typical of compact hypercellular Antoni A tissue (star), and hypocellular with Antoni B tissue (arrow) (Hematoxylin and eosin, x 100) seen.

Discussion and Conclusion

Intramedullary schwannoma is a rare spinal cord tumor, first described by Kernohan in 1931[12]. Since its initial discovery, 199 cases, including ours, have been reported in the literature, highlighting its uncommon nature[1-6]. The general characteristics of the last 6 publications in the literature are summarized in Table I. These cases show that IMS is more common in the cervical region and is lowest in the lumbar region. It is more common in males. The mean age of onset is 45.2 years. All cases underwent surgery.

Table I. General characteristics of the last 6 intramedullary schwannoma studies published between 2020 and 2024.

Study	Patients (n)	Mean age (years)	M/F Ratio	Surgery (n)	Most Common lesion segments (n, %)
Swiatek et al. (1) 2020	166	40.5	92/74	N/A**	Cervical (92, 55)
Li et al. (2) 2024	11	48.7	4/7	GTR (8) STR (3)	Servical (4, 36) Thoracic (4, 36)
McCann et al. (3) 2023	1	40	1/0	GTR (1)	Thoracic (1, 100)
Hara et al. (4) 2023	11	50.2	7/4	GTR (9) STR (2)	Servical (6, 54)
Adji et al. (5) 2024	1	43	0/1	GTR (1)	Thoracic (1, 100)
Salamah et al. (6) 2024	8	42,2	7/1	N/A**	Cervical (4, 50) Thoracic (4, 50)
Dogan et al.*	1	52	1/0	GTR (1)	Cervical (1, 100)
Total	199	45.2	112/87	199	Cervical (107, 53.7)

* Current research to compare other studies. ** All cases underwent surgery, but the subtype was not specified. M: Male, F: Female, GTR: Gross total resection, STR: Subtotal resection, N/A: Not available

The exact etiopathogenesis of IMS remains unclear, and various theories are being discussed. Notably, Schwann cells are not typically found in the central nervous system, which adds to the mystery surrounding the intramedullary localization of this pathology. To date, no definitive explanation has been established for this unique occurrence. The literature focuses on five main etiopathogenesis[2-6,13-17]:

1) Subpial dissemination

2) Schwann cells in the dorsal root entry zone

3) Medullary invasion via perivascular nerve plexus

4) Originating from pial mesodermal/neural crest cells

5) Chronic inflammation/trauma

The medullary location of the tumor could be explained by the subpial dissemination theory. The intramedullary tumor also did not have an extradural component and was notable for its subpial intramedullary location, confirming the subpial spread theory described in the literature. To date, five cases using the terminology of subpial schwannoma have been reported in the literature[13-17]. The mechanism by which this pathology causes intramedullary spread is still debatable. The most accepted theory of subpial spread is intramedullary dissemination of Schwann cells through pial invasion from the critical area where the sheath on the dorsal root is lost[13]. Although it explains the cases of pial dissemination in the literature, it is insufficient to describe other cases. Other theories have suggested that the tumor origin may originate from different regions and therefore its invasion. However, these theories are also insufficient to determine the genetic and functional mechanism by which etiopathogenesis occurs[13-17].

The recent IMS series reported by Li et al.[2], which included the largest case series in the literature (11 patients), discussed tumor location, radiological findings, and treatment strategies. All tumors were dorsal and even dorsolaterally located and had the characteristic of pushing the spinal cord to one side. In our case, the radiological features of the tumor were similar to other cases in the literature. Unilateral compression effect may be an important finding to suspect IMS. Another study in Japan by Hara et al.[4] the results of a multicenter study conducted by and presenting 11 cases also yielded similar results. The study focused on GTR/STR (gross total resection/subtotal resection) and follow-up options in the case of medullary invasion and mentioned the effect of the region where the lesion originated on tumor spread. It was emphasized that STR option should be reviewed in cases where GTR is not possible in the presence of subpial spread and medullary invasion. Schwannomas have a good dissection plan and have low recurrence rates with GTR. In our case, since pial dissemination and medullary invasion were not in our preoperative surgical plan, difficulties were encountered in the resection of this tumor. In addition, since it was evaluated as extramedullary, preoperative ION was not used. If there is a preliminary diagnosis of schwannoma in this intradural tumor, it is useful to prepare for ION.

All these discussions do not change the treatment method of the disease, and the gold standard treatment is still surgical total excision. On the other hand, more comprehensive research and development of IMS classification and definitions in preoperative preparation, postoperative prognosis and follow-up continue to be needed. Larger case series and more comprehensive studies are needed to understand the pathophysiology better and optimize treatment strategies. Research studies are of great importance to uncover the mysteries surrounding IMS and provide better outcomes for patients diagnosed with this rare spinal cord tumor.

Ethics Committee Approval Information:
The study is a case report and does not require ethics committee approval. Written consent was obtained from the patient for the publication of the case report and accompanying images.

Researcher Contribution Statement:
Idea and design: Ş.D.; Data collection and processing: A.İ.O., M.Ö.; Analysis and interpretation of data: A.İ.O., P.E.; Writing of significant parts of the article: Ş.D.
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References

1. Swiatek VM, Stein KP, Cukaz HB, et al. Spinal intramedullary schwannomas-report of a case and extensive review of the literature. Neurosurg Rev 2021;44(4):1833-1852.
2. Li J, Song Z, Shang A. Clinical Features and Surgical Management of Intramedullary Schwannoma of the Spinal Cord. World Neurosurg 2024;185:e1280-e1286.
3. McCann MM, Hain JD. Intramedullary melanotic schwannoma: a rare presentation of a rare tumor. Illustrative case. J Neurosurg Case Lessons 2023;5(7):CASE22391.
4. Hara T, Mizuno M, Hida K, et al. Investigators of Intramedullary Spinal Cord Tumors in the Neurospinal Society of Japan. Intramedullary Schwannoma of the Spinal Cord: A Nationwide Analysis by the Neurospinal Society of Japan. Neurospine 2023;20(3):747-755.
5. Adji NK, Putri KYW, Indreswari L, Gunawan R, Nugraha MY. Multiple thoracic intramedullary schwannoma: A case report. Int J Surg Case Rep 2024;116:109330.
6. Salamah HM, Eltokhy AG, Ezzat M, Alkheder A, Taha MM. Spinal intramedullary schwannoma: Report of two cases with review of the literature. Int J Surg Case Rep 2024;117:109479.
7. Tahta A, Cetinkal A, Cakir A, Sekerci Z. Intramedullary schwannoma of cervicomedullary junction: A case report. Neurochirurgie 2022;68(3):331-334.
8. Singh R, Chaturvedi S, Pant I, Singh G, Kumari R. Intramedullary schwannoma of conus medullaris: rare site for a common tumor with review of literature. Spinal Cord Ser Cases 2018; 6;4:99.
9. Cheng X, Liu J, Le J, Huang S, Chen H, You C. Invasive intramedullary melanotic schwannoma: case report and review of the literature. Eur Spine J 2018;27(Suppl 3):303-308.
10. McCormick PC, Stein BM. Intramedullary tumors in adults. Neurosurg Clin N Am 1990; 1(3):609-30.
11. Maynard FM Jr, Bracken MB, Creasey G, et al. International Standards for Neurological and Functional Classification of Spinal Cord Injury. American Spinal Injury Association. Spinal Cord 1997;35(5):266-274.
12. Kernohan JW WH, Adson AW: Intramedullary tumors of the spinal cord. A review of fifty-one cases, with an attempt at histologic classification. Arch Neurol Psychiatry 1931;25:679–701.
13. Decharin P, Suvithayasiri S, Nivatpumin P, Trathitephun W. Subpial Schwannoma of the Cervical Spinal Cord: A Case Report and Its Intraoperative Finding Supporting a Theory of the Pathogenesis of an Intramedullary Schwannoma. Asian J Neurosurg 2022;17(1):108-111.
14. Ozawa N, Tashiro T, Okamura T, Koyama K, Ohata K, Inoue Y. Subpial schwannoma of the cervical spinal cord mimicking an intramedullary tumor. Radiat Med 2006;24(10): 690–694.
15. Houkin K, Abe H, Iwasaki Y, Tsuru M, Miyasaka K. Spinal sub-pial schwannoma. Case report [in Japanese]. Neurol Med Chir (Tokyo) 1983;23(05):370–374.
16. Kim SD, Nakagawa H, Mizuno J, Inoue T. Thoracic subpial intramedullary schwannoma involving a ventral nerve root: a case report and review of the literature. Surg Neurol 2005;63(04): 389–393, discussion 393.
17. Suematsu Y, Tsuji O, Nagoshi N, et al. Concurrent dorsal subpial schwannoma and ventral meningioma arising at the same upper cervical level: a case report. Spinal Cord Ser Cases 2020;6(01):64.