Spinal Intradural Extramedullary Nerve Sheath Tumors and Meningiomas: Surgical Management and Outcome

Ali Kotb Ali, Khaled El Bahy, Omar Yousef Hammad, Ashraf G. Al- Abyad and Adel Hussein El Hakim
Neurosurgery Department, Ain Shams University, Cairo, Egypt

ABSTRACT

Objective: *This study was done on 40 patients having extramedullary cord tumor (nerve sheath tumor and meningiomas) to evaluate the clinical presentation, investigation, surgery, post-operative outcome and recurrence.* ***Patients:*** *This study was carried out on 40 patients having diagnosis of spinal nerve sheath tumors or meningiomas and treated in the period between 1999 and 2005. The medical records of patients, such as their clinical presentations, neuroradiological findings, operative reports, functional outcome, complications and recurrence, were evaluated. The average age of these 40 patients was 38±13 years. Female sex predominated slightly in meningiomas while male sex slightly predominated in nerve sheath tumors.* ***Results:*** *The average duration of symptoms until admission for surgery was significantly shorter for patients with nerve sheath tumors (22.1±23 months) than for patients with meningiomas (25±15 months). Complete tumor removal was achieved in 60% of the patients; 74.1% in nerve sheath tumors and 30.8% in meningiomas. Resection of a clinically relevant nerve root was done in 14 cases of nerve sheath tumors; 2 (14.3%) in the cervical, 5 (35.7%) in the thoracic and 7 (50.0%) in the lumbar regions. Post-operative complications occurred were CSF leakage in 7 cases (25.9%) of nerve sheath tumors and in 4 cases (30.8%) of meningiomas, while wound infection was observed in 1 patient (3.7%) of nerve sheath tumors and 2 patients (15.4%) of meningiomas.* ***Conclusion****: Early diagnosis and surgery is the preferred management in cases of spinal nerve sheath tumors and meningiomas due to its associated excellent functional improvement and low recurrence rates. Surgical transsection of the nerve root carrying nerve sheath tumors will lead to radical removal and less incidence of recurrence; but this must be weighed against the risk of spinal cord damage.*

INTRODUCTION

Spinal cord tumors account for about 15% of central nervous system neoplasms[11]. About two thirds of spinal cord tumors in adults are extramedullary[9,11]. Nerve sheath tumors, meningiomas and filum terminale ependymomas account for most extramedullary neoplasm[11]. Nerve sheath tumors are categorized as schwannomas or neurofibromas[4] and account for about 25% of intradural spinal cord tumors in adults[9]. Spinal meningiomas represent 25 to 46% of tumors of the spine[5]. Typically, they are located in the intradural extramedullary space, grow slowly and spread laterally in the subarachnoid space until they induce symptoms. They most frequently occur in the thoracic region in middle aged women[2,3,7,8,10,12].

Nerve sheath tumors, in particular, carry nearly the best prognosis of all spinal tumors. However, their propensity to recur if not resected completely, their extradural and extraspinal extensions and special features in patients with neurofibromatosis offer points of interest for a more detailed study[6]. Advances in radiological and surgical devices (MR imaging, neuro-monitoring, intra-operative ultra-sonography, operative microscope and ultrasonic surgical aspirator) have resulted in earlier diagnosis and aided in obtaining a total resection[3].

In this retrospective study, data from 40 patients (operated upon for excision of spinal nerve sheath tumors

or meningiomas) were collected and analyzed to evaluate the presenting clinical pictures, duration of symptoms, surgical management, post-operative outcome and recurrence in these cases.

PATIENTS & METHODS

The study was carried out on 40 patients with histologically diagnosed spinal nerve sheath tumors or meningiomas during the period between 1999 and 2005. These patients were operated in the Neurosurgery Department, Ain Shams University, Egypt. Evaluation of patients was based on their medical records, as regard their clinical presentations, neuroradiological findings, operative reports, functional outcome, complications and recurrence. The mean follow-up period was 20 months (range 3-72 months).

The evaluation of the clinical course was determined by using the Functional Independence Measure (FIM) 7 rating scale of disability[(1)].

Pre-operative imaging consisted of plain X-rays of the affected spinal segment. MRI enhanced with gadolinium was the modality of choice for the pre-operative imaging and post-operative study. All patients underwent classic posterior laminectomies after intra-operative fluoroscopic localization of the correct level. In cases where the lesion was located anteriorly, the laminectomy was extended laterally toward the articular process to provide sufficient exposure and avoid displacement of the spinal cord. After dural opening, inspection of the arachnoid was done to detect any scarring or spinal cord tethering then the arachnoid was dissected off the tumor capsule leaving the arachnoidal plane between tumor, spinal cord and nerve roots intact. In cases with nerve sheath tumors, small posterior tumors were generally removed in Toto. For large, lateral or anteriorly placed nerve sheath tumors, the tumor was decompressed first before attempting to dissect and remove the tumor capsule. If the tumor originated from a sensory nerve, the nerve root was cut and removed with the tumor to ensure radical removal. With clinically relevant nerve roots, the involved root was dissected proximal and distal to the tumor in an attempt to isolate the fascicle harboring the tumor. If this fascicle could be isolated and the remainder of the nerve was not infiltrated by tumor, only the affected fascicle was resected with the tumor. In cases of meningiomas, a plane was developed between the arachnoid and the tumor. The tumor was then internally debulked using suction and microscissors. After debulking in the majority of cases, the tumor was rolled away from the spinal cord and toward its dural attachment. The tumor was then removed from its dural attachment. Dura with remaining tumor was coagulated using bipolar diathermy.

Post-operatively, every patient underwent plain radiography. All patients underwent gadolinium enhanced MRI at 12 months post-intervention. Further scans were performed if clinical symptoms worsened again post-operatively or if tumor removal was incomplete.

Clinical assessment was performed before discharge and after 3 months. Additional follow-up information was obtained during further outpatient visits to detect any clinical deterioration in the form of development of a new neurological deficit or an aggravation of a pre-existing symptom without subsequent recovery.

RESULTS

In the period from 1999 to 2005, a total of 40 patients with spinal nerve sheath tumors or meningiomas were treated surgically in the Neurosurgery Department, Ain Shams University, Egypt. The average age of the patients was 38 ± 13 years (age ranged from 14 to 66 years). Female sex predominated slightly in meningiomas (7 females vs. 6 males), while male sex slightly predominated in nerve sheath tumors (14 males vs. 13 females). The average duration of symptoms until admission for surgery was significantly shorter for patients with nerve sheath tumors (22.1±23 months) than for patients with meningiomas (25±15 months)

Most patients demonstrated a slowly progressive course. One patient presented with a history of 10 years duration. For the majority of the patients, the first symptom noted was pain (in 92.6% of nerve sheath tumor and in 84.6% of meningiomas cases). Gait affection was present in 81.5% of nerve sheath tumor cases and in 84.6% of meningioma cases. Dysethesias or hypothesia was present in 88.9% of nerve sheath tumor and in 92.3% of meningioma cases. Sphincteric disturbance was recorded in 37.0% in nerve sheath tumor cases and in 69.2% of meningiomas cases.

Motor power was assessed pre- and post-operatively using the Medical Research Council Scale (MRCS). The muscle power was graded for each muscle group in the affected limb(s) and the mean was determined. As shown in table (1), the motor power in patients with nerve sheath tumors was grade 2 in 3.7%, grade 3 in 29.6%, grade 4 in 55.6% and grade 5 in 11.1% while in meningioma patients, it was grade 3 in 53.8%, grade 4 in 38.5% and grade 5 in 7.7%.

Table (1): Pre-operative motor power grading using the MRCS

Grade	Pre-operative motor power					
	Meningioma		Nerve Sheath Tumors		Total	
	No.	%	No.	%	No.	%
2	0	0	1	3.7	1	2.5
3	7	53.8	8	29.6	15	37.5
4	5	38.5	15	55.6	20	50.0
5	1	7.7	3	11.1	4	10.0
Total	**13**	**100**	**27**	**100**	**40**	**100**

Functional Independence Measure (FIM) 7 rating scale was used to evaluate the patients pre- and post-operatively [(1)].

Pre-operative FIM was shown in table (2) as grade 3 in 3.7%, grade 4 in 7.4% , grade 5 in 40.7%, grade 6 in 37.0% and grade 7 in 11.1% in nerve sheath tumors, while it was grade 3 in 15.4%, grade 4 in 23.1%, grade 5 in 38.5%, grade 6 in 15.4% and grade 7 in 7.7% in meningiomas. The FIM scale score was significantly increased post-operatively in both groups of patients

Table (2): Pre and post-operative FIM evaluation

Grade	Pre-operative FIM					
	Meningioma		Nerve Sheath Tumors		Total	
	No.	%	No.	%	No.	%
3	2	15.4	1	3.7	3	19.1
4	3	23.1	2	7.4	5	30.5
5	5	38.5	11	40.7	16	79.2
6	2	15.4	10	37.0	12	52.4
7	1	7.7	3	11.1	4	18.8
Total	**13**	**100**	**27**	**100**	**40**	**100**
	Post-operative FIM					
3	1	7.7	1	3.7	2	5
4	0	0	0	0	0	0
5	2	15.4	2	7.4	4	10
6	4	30.8	8	29.6	12	30
7	6	46.2	16	59.3	22	55
Total	**13**	**100**	**27**	**100**	**40**	**100**

As shown in table (3), 22.5% of the operations were performed in the cervical region (7 cases of nerve sheath tumors and 2 cases of meningiomas), 50.0% in the thoracic region (11 cases of nerve sheath tumor and 9 cases for meningiomas) and 27.5% in the lumbar region (9 cases for nerve sheath tumor and 2 cases for meningiomas).

Intradural nerve sheath tumors were more common than dumbbell-shaped tumors (with extradural extension) as it was 81.5% vs. 18.5%, respectively, whereas all meningioamas were totally intradurally.

Table (3): Distribution of lesions according to their levels

Lesion level	Meningioma		Nerve Sheath Tumors		Total	
	No.	%	No.	%	No.	%
Cervical	2	15.4	7	25.9	9	22.5
Thoracic	9	69.2	11	40.7	20	50.0
Lumbar	2	15.4	9	33.3	11	27.5
Total	**13**	**100**	**27**	**100**	**40**	**100**

Table (4): Site of lesion and its leve

Lesion site	Lesion level						Total	
	Cervical		Thoracic		Lumbar			
	No.	%	No.	%	No.	%	No.	%
Dumbbell-shaped	6	37.5	2	6.5	2	10	10	14.9
Intradural	10	62.5	29	93.5	18	90	57	85.1
Total	**16**	**100**	**31**	**100**	**20**	**100**	**67**	**100**

Table (5): Different pathology and degree of excision

Degree of excision	Meningioma		Nerve Sheath Tumors		Total	
	No.	%	No.	%	No.	%
Complete	4	30.8	20	74.1	24	60
Partial	9	69.2	7	25.9	16	40
Total	**13**	**100**	**27**	**100**	**40**	**100**

Table (4) shows that intradural lesions were slightly more common in the thoracic and lumbar spine with percentages of 93.5% and 90%, respectively, whereas the dumbbell tumors were encountered predominantly in the cervical area (37.5%).

Complete tumor removal was achieved in 60% of the patients; 74.1% in nerve sheath tumors and 30.8% in meningioma (table 5), (figure 1, 2, 3)

The complications occurred post-operatively were CSF leakage in 7 cases (25.9%) of nerve sheath tumors and 4 cases (30.8%) of meningiomas. Wound infection occurred in 1 patient (3.7%) of nerve sheath tumors and 2 patients (15.4%) of meningiomas. Meningitis and post-operative hematoma were observed in 1 patient (3.7%) for each complication in cases of nerve sheath tumors.

Four patients were operated for recurrence within 5 years; two for nerve sheath tumors and two for meningiomas. The first surgery was not done in our institute for all patients with recurrence.

Table (6): Different pathology and associated root resection.

Root resection	Meningioma		Nerve Sheath Tumors		Total	
	No.	%	No.	%	No.	%
Intact root	13	100	13	48.1	26	65
Resected root	0	0	14	51.9	14	35
Total	**13**	**100**	**27**	**100**	**40**	**100**

Table (6) shows that resection of a clinically relevant nerve root was done in 14 cases of nerve sheath tumors, 2 (14.3%) in the cervical, 5 (35.7%) in the thoracic and 7 (50.0%) in the lumbar regions. In the cervical or lumbosacral compartment, this resulted in a transient neurological deficit which lasted less than 3 months in 5 of these 14 patients (35.7%). Permanent motor deficit was observed in 1 patient while 2 patients suffered permanent sensory deficit. No nerve root was resected in any of the meningiomas cases.

In total, 4 patients demonstrated recurrences (2 patients with nerve sheath tumors and 2 with meningioma) and all of these 4 cases not operated for recurrence in the present study.

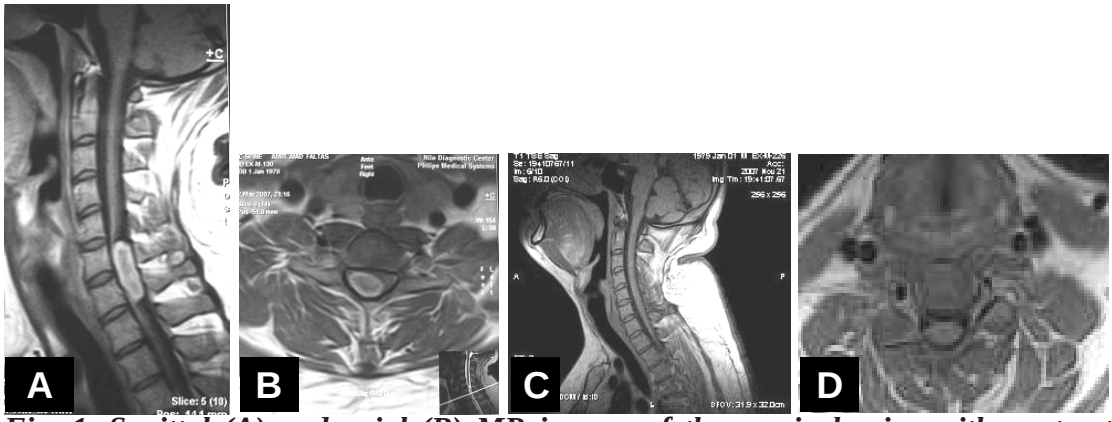

Fig. 1. Sagittal (A) and axial (B) MR images of the cervical spine with contrast showing enhanced neurofibroma. Sagittal (C) and axial (D) T1-weighted images after contrast showing total excision of the tumor.

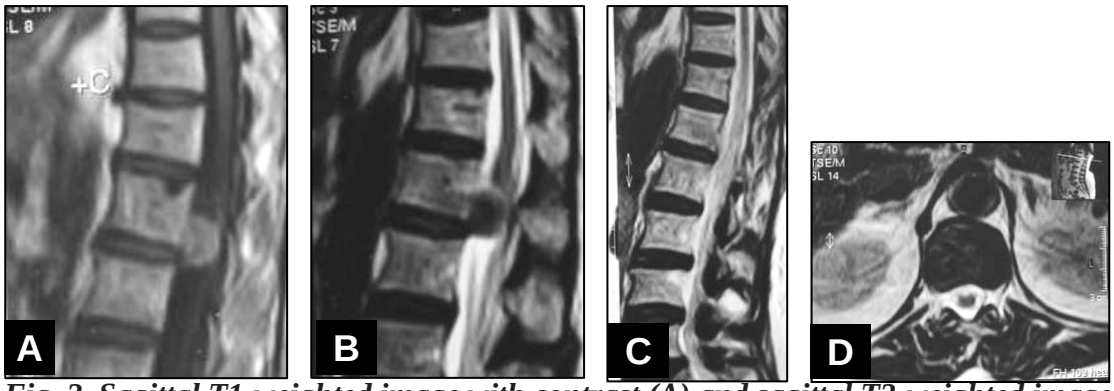

Fig. 2. Sagittal T1-weighted image with contrast (A) and sagittal T2-weighted image (B) of dorsolumbar spine showing enhanced meningioma. Sagittal T2-weighted image (C) and axial T2-weighted image (D) showed total excision of the tumor.

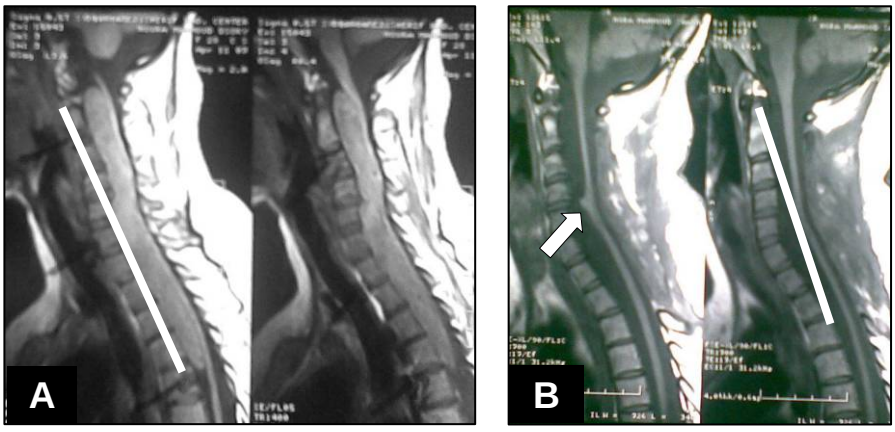

Fig. 3. Sagittal MR image of the cervical spine with contrast (A) showing enhanced neurofibroma. The white line denotes the extension of the tumor. Sagittal T1-weighted image after contrast (B) showed total excision of the tumor except very small piece attached to the nerve root (small arrow). The white line in image B denotes the space after tumor excision.

DISCUSSION

The annual incidence of primary intraspinal neoplasm is approximately five per million for females and three per million for males. Spinal intradural extramedullary tumors account for two thirds of all intraspinal neoplasms.[1]. Nerve sheath tumors are the most common spinal tumors and they frequently occur at an age between 25 and 50 years with no clear discrepancy between men and women[1,11,14,17,19,21,23]. Meningiomas are the second most common intradural spine tumors after neuromas[9]. Spinal meningiomas occur less frequently than intracranial ones and account for approximately 7.5 to 12.7% of all meningiomas[24]. Spinal neuromas and neurofibromas carry an excellent prognosis after complete removal[2,8,9,10,17,21].

There was typically a delay between the onset of symptoms and diagnosis. The mean duration of symptoms prior to presentation was 1 to 2 years[13, 15, 16, 20]. This study also shows slowly progressive course, and one patient presented with a history of 10 years. The first symptom noted was pain (92.6% in nerve sheath tumors and 84.6% in meningiomas). Gait affection was present in 81.5% with nerve sheath tumors and in 84.6% with meningiomas. Dysthesias was observed in 88.9% with nerve sheath tumors and in 92.3% with meningiomas. Sphincteric disturbances were recorded in 37.0% in nerve sheath tumors and in 69.2% with meningiomas. In other studies, some patients showed pain symptoms 15 to 20 years prior to diagnosis of spinal meningioma and most patients demonstrated a slowly progressive course[16]. The patients with spinal nerve sheath tumors in Klekamp and Samii study[14] showed that the first symptom noted was pain in 74% of cases, gait ataxia in 8% and dysthesias in 8% of cases. Jinnai and Koyama[11] reported in their studies of 149 cases with spinal nerve sheath tumors that pain was present in 36.9%, motor weakness in 24.2% and dysthesias and/or numbness in 25.6% of cases. On the other hand, in studies of spinal meningiomas, weakness was present in 64% and 32% of the patients were non-ambulatory at the time of presentation.

Levy et al.[16] reported that the rate of misdiagnosis in their pre MR imaging studies was 33% and the errors in diagnosis delayed treatment, which sometimes leads to inappropriate surgery. Seven of their patients underwent inappropriate surgeries including lumbar disc exploration and knee surgery. In the present study, instead of the presence of MRI to diagnose this pathology, there was a radiological misdiagnosis of prolapsed inter-vertebral disc in two patients with lumbar nerve sheath tumors and one patient with dorsal meningiomas. After having full neurological assessment and proper investigations, the correct diagnosis of those patients was reached.

Magnetic resonance imaging is the best imaging technique for diagnosing spinal meningiomas. It clearly delineates the level of the tumor and its relation to the cord, which is useful in planning surgery[6,16,24]. Klekamp and Samii[15] reported that MR imaging led to early diagnosis of spinal meningiomas that helped the patients to have less severe neurological deficits at the time of surgery. Typically, spinal meningiomas were isointense to the normal spinal cord on T1- and T2-weighted images, and they displayed intense enhancement after gadolinium injection[5].

Care should be taken to preserve the arachnoidal plane between tumor

capsule, spinal cord and neighboring nerve roots[14]. The arachnoid provides the best safeguard against inadvertent injury of these structures or their blood supply[14,21]. On the other hand, arachnoid scarring after previous operations may cause significant problems in preserving important structures and concurrently removing the tumor completely[7,14,21].

Klekamp and Samii[14] recommended the transsection of the nerve root carrying the neuroma to ensure radical removal, whenever justifiable. In the present study, root resection was done in 14 cases of nerve sheath tumors; 2 in cervical, 5 in thoracic and 7 in lumbar regions. Transient neurological deficits were observed in 5 patients who have gradually improved. Permanent sensory deficits were observed in two patients and motor deficit in one patient. Other studies[2,8,14,17,22] documented that transsection of a motor root in the cervical or lumbar area will not be followed by additional and permanent neurological deficits in the overwhelming number of cases. Destruction of the root by the neuroma and concomitant innervation from neighboring nerve roots seem to offer the best explanations for this observation[14,22]. Although definitive conclusion cannot be drawn, sacrifice of the ventral nerve root in critical areas may not lead to such disastrous outcomes as reported previously[11,12].

Although total resection of spinal nerve sheath tumors has been considered to be feasible[11,14], some cases in which total resection was not achieved have been reported[3,18,25]. Klekamp and Samii[14] documented complete tumor removal in 86% of their patients with spinal nerve sheath tumors .In meningiomas, complete resection was achieved in 82 to 99% of cases [5,13,15,16,20,24]. There may be two obstacles to do total resection in these tumors; one is adhesion to the spinal cord due to hemorrhage, inflammation or subpial localization and the other is the critical structures attached to extradural components outside the spinal canal in the cervical region, such as the vertebral artery[11]. In this study, total resection was achieved in 74.1% of cases of nerve sheath tumors and 30.8% of cases of meningiomas. Partial resection was done in 4 cases of dumbbell-shaped nerve sheath tumors out of five cases. In general, the benefits of complete resection need to be weighed against the potential for spinal cord damage[6,15,16,20,24].

In the present study, the improvement of the neurological status after surgery was found to be 74.1% in cases of nerve sheath tumors and 76.9% in cases of meningiomas. In other studies, the improvement of neurological status was approximately 80% of patients having nerve sheath tumors[3,11,14] but in cases of spinal meningiomas, the functional improvement occurred in 53 to 95% of cases in different studies[5,13,15,16,20,24].

CONCLUSION

Early diagnosis and surgery is the preferred management in cases of spinal nerve sheath tumors and meningiomas due to its associated excellent functional improvement and low recurrence rates. Surgical transsection of the nerve root carrying nerve sheath tumors will lead to radical removal and less incidence of recurrence; but this must be weighed against the risk of spinal cord damage.

REFERENCES

1. **Albanese, V. and Platania, N.:** Spinal intradural extramedullary tumors. Personal experience, J. Neurosurg. Sci., 46,18-24, 2002.

2. **Cervoni, L., Celli, P., Scarpinati, M. and Cantore, G.:** Neurinomas of the cauda equina clinical analysis of 40 surgical cases, Acta Neurochir. (Wien), 127, 199-202, 1994.
3. **Conti, P., Pansini, G., Mouchaty, H., Capuano, C. and Conti, R.:** Spinal neurinomas: Retrospective analysis and long term outcome of 179 consecutively operated cases and review of the literature, Surg. Neurol., 61, 34-43, 2004.
4. **Ditunno, J.F., Jr.:** Functional assessment measures in CNS trauma, J. Neurotrauma, 9 (suppl. 1), S301-S305, 1992.
5. **Gezen, F., Kahraman, S., Canakci, Z. and Beduk, A.:** Review of 36 cases of spinal cord meningioma, Spine, 25, 727-731, 2000.
6. **Gottfried, O.N., Gluf, W., Quinones, H.A., Kan, P. and Schmidt, M.H.:** Spinal meningiomas: Surgical management and outcome, Neurosurg. Focus, 14, 2, 2003.
7. **Halliday, A.L., Sobel, R.A. and Martuza, R.L.:** Benign spinal nerve sheath tumors: Their occurrence sporadically and in neurofibromatosis types 1 and 2, J. Neurosurg., 74, 248-253, 1991.
8. **Hanakita, J., Suwa, H., Nagayasu, S., Nishi, S., Iihara, K. and Sakaida, H.:** Clinical features of intradural neurinomas in the cauda equina and around the conus medullaris, Neurochirurgia (Stuttg), 35, 145-149, 1992.
9. **Helseth, A. and Mork, S.J.:** Primary intraspinal neoplasms in Norway, 1955 to 1986, A population based survey of 467 patients, J. Neurosurg., 71, 842-845, 1989.
10. **Hori, T., Takakura, K. and Sano, K.:** Spinal neurinomas: clinical analysis of 45 surgical cases, Neurol. Med. Chir. (Tokyo), 24, 471-477, 1984.
11. **Jinnai, T. and Koyama, T.:** Clinical characteristics of spinal nerve sheath tumors: Analysis of 149 cases, Neurosurgery, 56, 510-515, 2005.
12. **Kim, P., Ebersold, M.J., Onofrio, B.M. and Quast, L.M.:** Surgery of spinal nerve schwannoma. Risk of neurological deficit after resection of involved root, J. Neurosurg., 71, 810-814, 1989.
13. **King, A.T., Sharr, M.M., Gullan, R.W. and Bartlett, J.R.:** Spinal meningiomas: A 20 years review, Br. J. Neurosurg., 12, 521-526, 1998.
14. **Klekamp, J. and Samii, M.:** Surgery of spinal nerve sheath tumors with special reference to neurofibromatosis, Neurosurgery, 42, 279-289, 1998.
15. **Klekamp, J. and Samii, M.:** Surgical results for spinal meningiomas, Surg. Neurol., 52, 552-562, 1999.
16. **Levy, W.J., Jr., Bay, J. and Dohn, D.:** Spinal cord meningioma, J. Neurosurg., 57, 804-812, 1982.
17. **Levy, W.J., Latchaw, J., Hahn, J.F., Sawhny, B., Bay, J. and Dohn, D.F.:** Spinal neurofibromas: A report of 66 cases and a comparison with meningiomas, Neurosurgery, 18, 331-334, 1986.
18. **Lot, G. and George, B.:** Cervical neuromas with extradural components: Surgical management in a series of 57 patients, Neurosurgery, 41, 813-820, 1997.
19. **McCormick, P.C., Post, K.D. and Stein, B.M.:** Intradural extramedullary tumors in adults, Neurosurg. Clin. N. Am., 1, 591-608, 1990.
20. **Roux, F.X., Nataf, F., Pinaudeau, M., Borne, G., Devaux, B. and Meder, J.F.:** Intraspinal

meningiomas: Review of 54 cases with discussion of poor prognosis factors and modern therapeutic management, Surg. Neurol., 46, 458-463, 1996.

21. **Salah, S., Horcajada, J. and Perneczky, A.:** Spinal neurinomas: A comprehensive clinical and statistical study on 47 cases, Neurochirurgia (Stuttg), 18, 77-84, 1975.
22. **Schultheiss, R. and Gullotta, G.:** Resection of relevant nerve roots in surgery of spinal neurinomas without persisting neurological deficit, Acta Neurochir. (Wien), 122, 91-96, 1993.
23. **Schwartz, T.H. and Mccormick, P.C.:** In Neurological surgery, Spinal cord tumors in adults, Winn, H.R. (ed), Youmans, Saunders, p. 4817-4834, 2004.
24. **Solero, C.L., Fornari, M., Giombini, S., Lasio, G., Oliveri, G., Cimino, C. and Pluchino, F.:** Spinal meningiomas: Review of 174 operated cases, Neurosurgery, 25, 153-160, 1989.
25. **Tatagiba, M., Bini, W., Sepehrnia, A., Eichhorn, R., Kleider, A. and Samii, M.:** Involvement of spinal nerves in neurofibromatosis, Neurosurg. Rev., 17, 43-49, 1994.