J Turgut Ozal Med Cent 2017;24(1):54-8
ORIGINAL ARTICLE DOI: 10.5455/jtomc.2016.11.123

Intradural extramedullary nerve sheath tumors: 37 cases clinical series

Yurdal Gezercan, Emre Bilgin, Gokhan Cavus, Vedat Acik, Ali Ihsan Okten

Adana Numune Training and Research Hospital, Neurosurgery Clinic, Adana, Turkey.....

Abstract

Aim: The nerve sheaths are generally called as schwannomas and they are developed from dorsal nerve roots. They form 25% of the intradural tumors. They are observed more frequently in 4th and 5th decades. They are generally intradural however 20% of them develop as dumbbell shaped and become intradural-extradural components. The treatments of them are laminectomy or laminoplasty and total excision.

Materials and Methods: 37 spinal schwannoma cases operated in our clinic between the years of 2005-2015 and are investigated in our study in terms of age, sex, complaint, duration of complaints, radiological localizations, neurological examination findings, relation with the dura, surgical method implemented and the effects of these parameters on the clinical course.

Results: 19 of 37 cases were males (51.4%), 18 were females (48.6%). Age average was 44.9 (20-79). The most frequent complaint was pain (72.9%) and weakness and numbness in the legs. They had lumbar (59.5%), thoracic (24.3%), cervical (16.2%) localizations. 18.9% of the cases revealed extraforaminal extension to abdomen or thorax in the shape of dumbbell. 80% total mass excision was provided by performing 80% laminectomy and 20% laminoplasty for the patients. 3 CSF (Cerebrospinal fluid) leakage, 2 superficial infection, 1 meningitis, 3 temporary neurological deficit and 1 permanent neurological deficit developed as postop complication.

Conclusion: Nerve sheath tumors are the most frequent type of intradural extramedullary tumors and they have benign characteristics. They absolutely provide good results in early diagnosis, early period surgery, without extraforaminal extension and in total resection.

Keywords: Spinal Schwannoma; Total Resection; Intradural Extramedullary Tumors.

INTRODUCTION

Schwannomas are benign tumors originating from myelin sheath of the peripheral nerves called as schwannoma cell tumor, neuroma, neurinoma, neurilemmoma. They may occur by originating from cranial nerves in the head and peripheral nerves from any part of the body. (1) Spinal schwannomas mostly originate from dorsal branch and rarely from ventral branch of the nerve root. (2) The frequency in general population is 0.3-0.5/100.000. They form 1/3 of all spinal tumors. They are the most common among the İntradural Extramedullary (ID-EM) tumors. They are common in 3rd-5th decades and it is notified that they are more frequently observed in females than males in various series (3,4).

They are localized in posterior and posterolateral part of the spinal cord. They reveal localizations in thoracic region the most and then in cervical and lumbar regions. 58% of these tumors reveal intradural-extramedullary, 27% is extradural and 15% intra/extradural in dumbbell-shaped localizations. They rarely reveal intramedullary localizations (3,5-8).

Received: 13.12.2016
Accepted: 18.12.2016

Correspondence
Gökhan Çavuş
Adana Numune Training and Research Hospital, Department of Neurosurgery, Adana, Turkey
E-mail: gokhanctf@yahoo.com

Schwannomas are 90% benign and they have very slow growing patterns. The period passing between diagnosis and emerging of the symptoms is nearly 2-3 years. Segmental and radicular pain frequently intense at nights is the first revealed symptom. Findings of cord compression in the advanced delaying cases are revealed. There are 80% pain, 10% motor weakness, 10% sphincter and sensation disorders (5,7,9-11).

In terms of the tumors developed in a long period of time, 50% rate of expansion of interpeduncular distance, cavitation in the posterior contour of the neuro-centrum, interforaminal expansion may be observed in X-Ray (CR). Sharply circumscribed mass and neighbouring bone erosion may be observed in CT. The golden standard in the diagnosis of intradural extramedullary tumors is MRI in the present time. Isointense or slightly hypo intense cord according to the nerve roots is observed in T1, 75% hyper intense, 40% cyst, 10% hemorrhage may be observed in T2 (5,7,9,11,12).

Total removal of the tumor should be aimed in the surgery. As it is originated from the nerve root, it should be dissected and if needed, the root should be cut and prognosis is quite good as well as there will be nearly no problem. There may be recurrence in the subtotal post-operative period and there is no radiotherapy indication in the post-operative period. There are complications such as arachnoiditis, vertebral deformity in the literature (3,7-10,13,14).

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MATERIALS and METHODS

The investigation of treatment results and the factors affecting the treatment results of 37 patients with schwannoma pathology results and operated with spinal tumor diagnosis in our clinic between the years 2005 and 2015 is aimed.

Clinic, radiological and histo-pathological investigations were performed for the diagnosis. The cases were scanned retrospectively and the files, imaging results and operation notes were investigated. The cases were investigated and evaluated by taking into consideration the ages, sexes, complaint findings, complaint periods, imaging methods, spinal canal localization regions, neurologic examination findings, relationships with dura, surgical methods and early surgical period results. Early period surgical results are achieved by comparing the neurological examination performed on the 10th day in the postoperative period and first time neurological examination results.

RESULTS

The age average of our cases were 44.9 (20-79) and 51.4% (19 cases) were males and 48.6% (18 cases) were females.

The most frequent first application complaint was pain 59.5% rate (22 cases). Extremity weakness was 10.8% (4 cases). The rate, for which pain and weakness were observed together, was 13.5% (5 cases).

3 of 5 cases, who had motor deficit in the neurological examination, were para-paretic and had 2/5 strength, 2 cases were mono-paretic and had 3/5 strength. Hypoesthesia was 10.8% (4 cases), paraplegia was 2.7% (1 case) and the case rate coming with sphincter disorder such as urine incontinence was 2.7% (1 case).

In 37 cases, 91.9% (34 cases) was used with MRI together with CR, 8.1% (3 cases) was used with CR, MRI and Computerized Tomography (CT). And all of the cases were taken under surgery with MRI and CR.

Of the cases, 59.5% (22 cases) was lumbar localization, 24.3% (9 cases) was thoracic localization and 16.2% (6 cases) was cervical region localization (Figure 1).

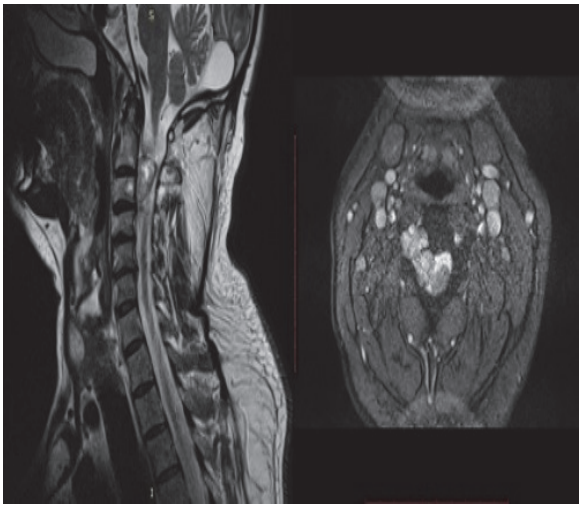

Figure 1. T2 sagittal and axial contrasted C2-3 dumbbell shaped schwannoma.

81.1% was ID-EM (30 cases), 18.9% (7 cases) was ED. Total resection was performed on 89.2% (33 cases) of the cases, gross total resection was performed on 10.8% (4 cases) and when early period surgical results were evaluated, partial recovery was observed in 54.1% (20 cases), no change was observed in 37.8% (14 cases). Recovery was observed as 4/5 para-paresis in 2 cases, who had 3/5 para-paretic and recovery was observed as 3/5 mono paresis in 1 case, who had 2/5 mono-paretic. In the post-operative period, CSF leaks were observed in 3 cases, superficial skin infections were observed in 2 cases and they were taken under control with appropriate treatments. Temporary neurological disorder was observed in 8.1% (3 cases) and neurological examination got better within a week (Table 1, table 2).

Table 1. Distribution of sex, complaint findings, imaging methods, spinal channel levels and localizations, neurological findings.

SCHWANNOMA		NUMBER	PERCENTAGE %
SEX	MALE	19	51.4%
	FEMALE	18	48.6%
FIRST COMPLAINT	PAIN	22	59.5%
	WEAKNESS	4	10.8%
	WEAKNESS WITH PAIN	5	13.5%
	PARAPLEGIA	1	2.7%
	LOSS OF SENSE	4	10.8%
	SPHINCTER PROBLEM	1	2.7%
IMAGING METHODS	DG and MRI	34	91.9%
	DG,MRI and CT	3	8.1%
CHANNEL LEVELS	LUMBAR	22	59.5%
	THORACIC	9	24.3%
	SERVICAL	6	16.2%
LOCALIZATIONS	INTRADURAL EXTRAMEDULLARY	30	81.1%
	EXTRADURAL	7	18.9%
NEUROLOGICAL EXAMINATION	MOTOR DEFICIT	5	13.5%
	SENSATION DISORDER	4	10.8%
	PARAPLEGIA	1	2.7%
	SPHINCTER DISORDER	1	2.7%

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Table 2. Method of the surgery performed and early surgery results.

SCHWANNOMA		NUMBER	PERCENTAGE (%)
SURGERY PERFORMED	TOTAL EXCISION	33	89.2%
	GROSSTOTAL EXCISION	4	10.8%
EARLY SURGERY RESULTS	PARTIAL RECOVERY	20	54.1%
	NO CHANGE	14	37.8%
	TEMPORARY DISORDER	3	8.1%

DISCUSSION

Spinal schwannomas generally originated from dorsal nerve roots are frequently intradural-extramedullary localized. They may be extradural or intra-medullary dumbbell shaped in less rates (15).

Schwannomas are mostly specified as extramedullary in literature. They may be observed as dumbbell shaped both intradural and extradural in 10-15%, extradural in 10% and intramedullary 1%. Rasmusen et al. evaluated histopathological investigation of 163 of 557 intraspinal neoplasm cases as schwannoma and notified only 1 of them as intramedullary localized schwannoma (16). Conti, Klekamp, Seppala specified 83.6%, 49.3%, 66% of schwannomas as intradural-extramedullary, 7.4%, 27.7%, 13% as extradural, 1.3%, 0% as intradural intramedullary in their own series. In terms of the cases, 81.1% (30 cases was found as intradural extramedullary, 18.9% (7 cases) as extradural, 0% as intramedullary in our own series. The rates of our series were observed as compliant with literature (4,7,8-10,18).

In general for schwannomas, there was no sex differentiation found as well as the age averages changed in 3rd and 5th decades. When sex distribution rates of schwannomas were considered, it was observed in a study conducted by Conti et al. that 61.1% was male and 38.8% was females and the average age was 44.3 and the age range changed between 7 and 80. On the other hand, Mc. Cormick et al. highlighted the equality of female and male rates and the frequency in 4th and 6th decades. In our series, 51.4% (19 cases) was male and 48.6% (18 cases) was female. The age average was 44.9, the age range was found as 20 and 79. The age and sex rates of our series are considered as compliant with the literature (4,9,10,13).

When the literature is investigated, it is observed that pain is the most frequent complaint. Seppala et al. specified in their study that the pain existed in nearly half of the cases. It was specified that it was localized in thoracic region most frequently, then in cervical and lumbar regions. Mc Cormick et al. defended that this localization order involved all intradural extramedullary tumors. Conti et al. specified the most frequent localization positions as 48.6% lumbar, 32.9% thoracic and 18.4% cervical region. In our series, lumbar region was observed as 59.5%, thoracic region was observed as 24.3% and cervical region was observed as 16.2%. Results compliant with the literature was achieved in our series in terms of this aspect (4,5).

In the light of the literature, it is seen that pain is the most common complaint. Seppala and colleagues reported that almost half of their cases were painful. In our study, the most frequent complaint of our cases was pain by 59.5% (22 cases) and this was followed by pain and weakness by 13.5% (5 cases). The case rate, who applied only due to weakness, was 10.8% (4 cases). Our cases applied to our clinic with paraplegia by 2.7% (1 case), sensation loss 10.8% (4 cases) and sphincter problem 2.7% (1 case). Segmental and radicular pain frequently intense at nights is the first revealed symptom when the literature is investigated. Application to clinic is observed with cord compression as a result of the cord compression of the grown tumor in advanced delayed cases. Also there are 80% radicular pain, 10% motor weakness, 10% sphincter and sensation disorders in the literature. Seppala stated that 46% localized pain, 31% para-paresis, 27% findings due to cord compression are observed (5,6,7,11,18**).** Pain complaint in our series was found as fundamental complaint compliant with the literature and the rates of our cases applying with weakness are observed as compliant with literature.

CR and CT has limited place in the diagnosis of schwannoma. Expansion of interpeduncular distance, cavitation in the posterior contour of the neurocentrum, foraminal expansion may be observed in direct graphy in terms of some benign tumors. CT has no superiority compared to MRI and it may assist for diagnosis in cases MRI is contraindicated. However, the golden standard diagnosis method at the present time is MRI. As we can achieve various plane sections, it is supportive for revealing definite limits, localization of the mass, invasion findings of neighboring structures. It enables performing diagnosis and definitive diagnosis as it reveals information of histological structure features. It is typical for classical heterogeneous sharp-edged contrast involvement schwannomas, thus this appearance is observed more commonly than other intradural extramedullary tumors. We used Direct graphy and MRI in 91.9% (34 cases), Direct graphy, MRI and CT together in 8.1% of our cases in our series. We used CT in 3 of our cases, who had extraforaminal extension, in order to evaluate the bone structure. We observed heterogeneous sharp-edged contrast involvement in contrasted MRI images of our cases and we recognized that our findings are compliant with the literature (5,17,9,19,20).

Mc Cormick et al. specified average complaint period as 2 years in their series and Conti et al. specified average complaint period over 2 years in their own

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series. This period is between 10 and 240 weeks and average complaint period is 37.6 weeks in our series. We operated many of our cases with the early diagnosis (7,10).

Laminectomy was performed in 80% (30 cases) of our cases and in 20% (7 cases) of them, laminoplasty was performed. Possible upper and lower extremity and at the same time sphincter complications were prevented during surgery by using Motor and Sensory stimulated potential monitorization (MEP-SEP) in 80% cases (21).

In 6 months-4 years follow-up of our cases, no recurrence or relapse was observed, no malign transformation was observed. Post-laminectomy kyphosis developed in long-term follow-ups in 2 of our cases with lumbar localizations, who were performed laminectomy, and deformity surgery was performed by operating for the second time. That was not observed in the cases that we performed laminoplasty. The region, in which post-laminectomy kyphosis is observed most commonly, is lumbosacral region in the literature and the long-term results of laminoplasty are better (22,23).

In the series of Conti et al., 83.53% of the ones with lumbosacral localization and 72.22% of the ones localized in cervical and thoracic regions revealed recovery. 22.22% of the ones in thoracic regions, 13.84% of the ones in lumbosacral regions and 13.63% of the ones in cervical region revealed partial recovery. No change was detected in 4.61% of the ones localized in lumbosacral regions, 4.64% of the ones localized in cervical region and 3.7% of the ones localized in thoracic regions. While positive results are observed in lumbosacral region, negative results are observed in cervical and thoracic regions.

When investigated in our series, no change was observed in our cervical and especially thoracic localized series while partial recovery was observed in our lumbar localized neurological deficit cases. Our findings were compliant with the literature. In the post-operative early period, CSF leak in 3 cases, superficial infections in 2 cases, meningitis in 1 case, temporary neurological deficit in 3 cases and permanent neurological deficit in 1 patient was observed. Their treatments were completed during the hospitalization periods and they are discharged.

CONCLUSION

We submitted a series including 37 cases in this study. Schwannomas are the most commonly observed intradural extramedullary benign spinal tumors. Diagnosis of intradural extramedullary tumors, hence schwannomas are performed easily by the modern imaging methods of our day. The main purpose of the treatment should be total resection as well as treatment approach in the lesions associated with the genetic diseases may differ. Early diagnosis before the occurrence of the neurological findings and removal of the tumor through totally as soon as possible are important factors affecting the prognosis and life quality of the patient.

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