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Research Article

Spinal Neurofibromas: Literature Review and Surgical Outcome

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Abstract

Objective: To determine the overall clinical outcome, specific features of spinal neurofibromas following surgery.

Background: Among all primary spinal neoplasms, approximately one-third are neurofibromas. Its incidence, clinical presentation, operative techniques and outcome of surgical resection need be studied thoroughly.

Methodology: This Retrospective study was conducted in department of Neurosurgery Lady Reading Hospital Peshawar, Pakistan from January 2016 to December 2016. Medical records of patients was revised and searched. Inclusion criteria was all symptomatic newly diagnosed and recurrent spinal neurofibromas. Exclusion criteria was asymptomatic spinal neurofibromas found as incidental finding. Patients were followed upto 3 months post operatively. The demographic data, clinical, radiological and histological details of patients were analyzed to assess and evaluate the predictive factors that affect post-operative clinical outcome. Diagnosis was made on clinical features, contrast enhanced spinal magnetic resonance imaging (MRI) and confirmed by histopathology of the lesion. Gross Total Microsurgical resection was carried out in all cases through a posterior approach. Post-Operative assessment was done in terms of clinical, neurological improvement of symptoms and surgical complications.

Results: Total number of patients were (15) harboring spinal neurofibroma. Mean age was (55 years). There were (8) males and (7) females patients. Neurofibroma was found in cervical spine in 11 cases (72%) and in thoracic spine in 4 cases (28%). Fresh cases were (13) and recurrent cases were (2).

Clinical presentations of patients were gait disturbance 14 cases (93%), Motor deficit/weakness 12cases (80%), sensory deficit 12 cases (80%), radicular pain 9 cases (60%), local spinal pain in 7 cases (47%), sphincter dysfunction in 9 cases (60%). The most common site of spinal neurofibromas was cervical region. All tumors were intramural extramedullary. All patients were thoroughly observed for 12 months. Post-operative neurological improvement was observed in 13 patients (87%). There was no operative mortality. None of the patient was subjected to adjuvant radiotherapy.

Conclusion: Spinal neurofibromas are benign tumors. Surgical excision can be safely done without having any post-operative deficits. Availability of many different tools and good microscope has made complete removal of tumors with no further recurrences.

Keywords: *Neurofibroma; Spinal Tumors; Surgical Outcome*

Introduction

Spinal schwannomas are slow-growing, benign lesions that occur throughout the spinal canal. These schwannomas represent 25% of all spinal tumors [1] and most occur sporadically and are solitary. Spinal schwannomas may also occur in the setting of neurofibromatosis type 2 and are an important pathological feature of this disease. Although schwannomas typically are small, 11% of them may be defined as giant, meaning that they extend over multiple vertebral levels, have a large extraspinal extension, or extend into myofascial planes [2]. Only 3% to 4% of patients presenting with a schwannoma are determined to have multiple lesions [3].

Schwannomas originate from Schwann cells of the dorsal nerve roots but in approximately 23% of cases, they may arise from the ventral nerve roots [4]. The tumor arises from a single nerve fascicle, with the remaining fascicles either displaced to one side or located around the tumor. Schwannomas appear grossly as a smooth globoid mass attached to a nerve, do not produce nerve enlargement, and are suspended eccentrically from the nerve [4]. They are firm, encapsulated neoplasms and can be cystic, hemorrhagic, or fat-containing. Histologically, they are composed of compact and interwoven bundles of long, spindle-shaped Schwann cells (Antoni type A tissue), which often are intermingled with sparse areas of more polymorphic Schwann cells embedded in a loose eosinophilic matrix (Antoni type B tissue) [4].

Most schwannomas are entirely intradural, but some extend intraforaminal region as a dumbbell mass or are purely extradural. Dumbbell tumors are spinal tumors with contiguous intraspinal, foraminal and extraforaminal components. It has been observed in one study that (49% to 84%) of schwannomas were intradural, (8% to 32%) were completely extradural, (1% to 19%) were both intradural and extradural, (6% to 23%) were dumbbell, and (1%) were intramedullary [5]. Intradural schwannomas were more common in the thoracic and lumbar regions, whereas dumbbell tumors occurred predominantly in the cervical spine [6,10].

Spinal schwannomas usually present in the fifth or sixth decades of life. Men and women are affected equally. Main presentations of this tumor is usually localized pain with sign and symptoms associated with nearest neural compressions. Neurological deficits develop in the course of time. Due to slow growing nature of this tumor, the symptoms may be subtle initially then worsening gradually. Localized pain, gait ataxia, motor weakness, bladder paresis, and dysesthesia may be presenting complains initially [7]. Duration of presentations of this disease may be varied from 2 - 3 years to as much as over 15 years.

Methodology

Study design: Retrospective descriptive study.

Place and duration of study: This study was conducted in department of Neurosurgery Lady Reading Hospital Peshawar, Pakistan from January 2016 to December 2016.

Inclusion criteria

All symptomatic newly diagnosed and recurrent spinal neurofibromas patients were included.

Exclusion criteria

Asymptomatic spinal Neurofibroma patients found as incidental finding were excluded.

Methods and patient: Total fifteen patients were studied after taking consent from patients and approval from ethical committee of hospital. Medical records of patients was revised and searched through questionnaire. Mean duration of post-operative follow-up was 3 months. The demographic data, clinical and radiological details of patients were analyzed to assess and evaluate the predictive factors

that affect post-operative clinical outcome. Diagnosis was made on clinical features, contrast enhanced spinal magnetic resonance imaging (MRI) and confirmed by histopathology of the lesion. Complete resection of tumors were done through posterior approach in all cases. Post-Operative assessment was done in terms of clinical, neurological improvement of symptoms and surgical complications.

Statistical analysis

Analysis was done by SPSS vs 20. All the categorical data was presented in frequency and percentages and continues data was evaluated in mean and standard deviation.

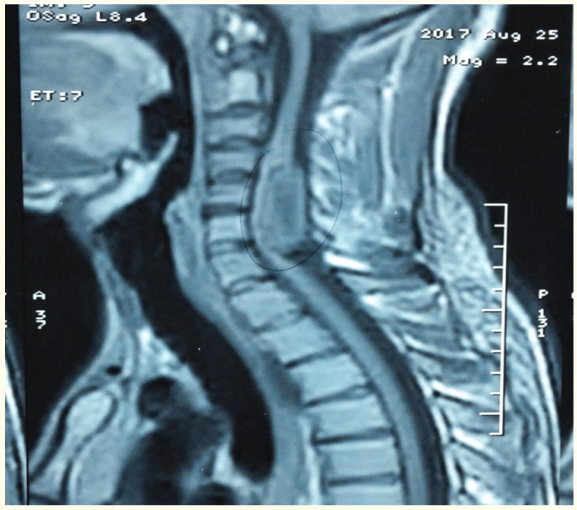
Results

Variable (N = 15)	
Age (Mean ± S.D)	55 ± 1.01
Female (%)	7 (46.66)
Male (%)	8 (53.33)
Region of neurofibroma (N = 15)	
Cervical spine	11 cases (72%)
Thoracic spine	4 cases (28%)
Clinical presentation (N = 15)	
Fresh cases	13 (86.66%)
Recurrent cases	2 (13.33%)

Table 1: *Demographic profile and baseline data of patients.*

Total number of patients were (15) harboring spinal Neurofibroma. The demographic profile and baseline data of patients were represented in table 1.

Figure 1 clinical presentation of patients.



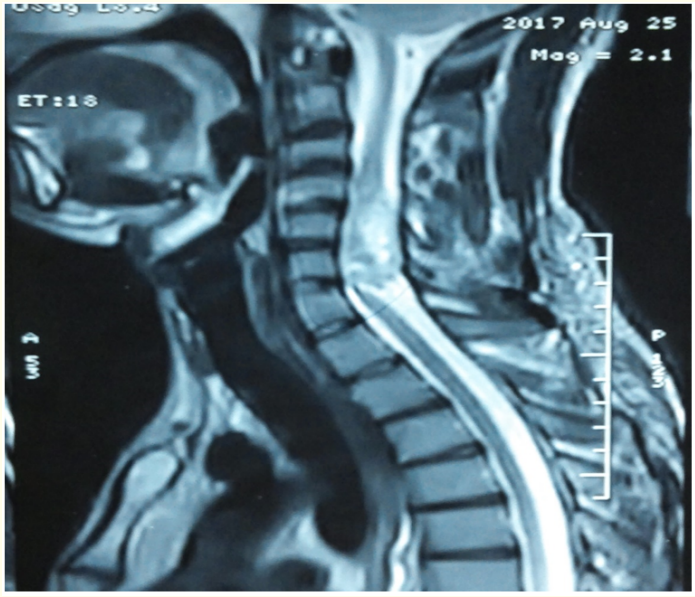


Figure 1: *MRI cervical spine with T1W and T2W images showing neurofibroma at C4, C5, C6 levels.*

Out of 15 patients most of them (93%) were represented gait disturbance at clinical presentation.

The most common site of spinal Neurofibroma was cervical region. All tumors were intradural extramedullary. Patients were followed upto 3 months. Neurologically 13 patients (87%) had significant improvements in post-operative periods. There were zero mortality in intraoperative periods. None of the patient was subjected to adjuvant radiotherapy.

Clinical presentation	Percentage improvement
Gait disturbance	11/14 (78%)
Motor deficits (Weakness)	10/12 (83%)
Sensory deficits (Numbness/Paranesthesia)	12/12 (100%)
Radicular pain	9/9 (100%)
Local spinal pain	7/7 (100%)
Sphincter dysfunction	3/9 (33%)

Table 2: *Post-operative outcome at F/U visits.*

All patients were operated via a midline posterior approach. Preoperatively, the corresponding spinal level was marked with a Coin and plain XRAY AP VIEW of the corresponding spinal level was taken, which helped in identification of correct spinal segment where Neurofibroma was diagnosed after skin incisor.

Discussion

Spinal neurofibromas account for about 20% of primary spinal cord tumors in adults. There is no significant prevalence difference between males and females. The incidence of spinal neurofibromas varies with the age of affected patients who are between the 40 and

60 years. In our case, mean age was 55 years and peak incidence was also between the same age groups. In our study, the higher incidence was seen in the cervical spine in 11 cases (72%) and in thoracic spine in 4 cases (28%). According to studies, local pain, radicular pain, motor and sensory deficits due to a spinal cord lesion and Bladder/bowel disfunction found to be Common clinical presentations [8]. Similarly, our study shows the similar trend of presentations (Figure 1).

In our study, we found that the most common site of spinal neurofibromas was cervical region (Table 1). Similarly, recent studies showed that tumors were located mainly in the cervical region and tend to grow both extra- and intradurally [8]. Another data from institutional neuropathology found that most cases of neurofibromas were found in cervical region [9]. All tumors were intradural extramedullary. Patients were followed upto 3 months. Postoperative neurological improvement was observed in 13 patients (87%). There was no operative mortality. None of the patient was subjected to adjuvant radiotherapy. Less than 1% of those affected with neurofibromas may have malignant tumors and may depend upon histological analysis [10]. Total removal of neurofibromas that are not associated with neurofibromatosis is generally curative. However, tumors with extensive Para spinal involvement that are sub totally resected have a definite propensity to recur. Deficits resulting from sacrifice of the involved nerve roots are usually minor and well tolerated. Otherwise, we experienced two recurrences among 15 cases by subtotal removal to avoid the nerve roots injury.

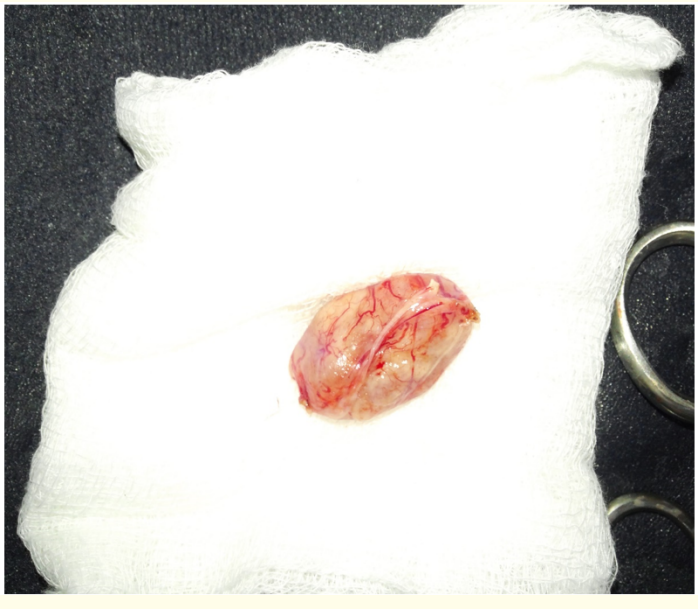


Figure 2: *Neurofibroma after removal along with involved nerve root.*

Conclusion

Spinal neurofibromas are benign tumors. Surgical excision can be safely done without having any post-operative deficits. Availability of many different tools and good microscope has made complete removal of tumors with no further recurrences.

Ethics Approval and Consent to Participants

Not applicable.

Consent for Publication

An informed written consent was obtained from the patient.

Availability of Data and Materials

Not applicable.

Competing Interest

None.

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Authors Contribution

Conception, Diagnosis and design, Radiological diagnosis and Final approval of manuscript; - Adnan Khaliq

Manuscript preparation, Technical revision, Manuscript editing and revision; Farooq Azam, Adnan Khaliq, Mumtaz Ali, Bipin Chaurasia.

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