ORIGINAL ARTICLE

# Management of Cervical Neurofibroma Type 1

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## *ABSTRACT*

***Objective:*** *To determine the clinical presentation and postoperative outcome of cervical neurofibroma type 1.*

***Study design:*** *Retrospective study.*

***Place and Duration of Study:*** *Hayatabad Medical Complex Hospital Peshawar Khyber pakhtoonkhawa from 1st February 1999 to 31st July 2011.*

***Material and Methods:*** *A total 41 patients with symptomatic cervical spine neurofibromas who underwent surgical decompression and tumor resection were included in this study. Both gender (male and female) and patients in the age range of 20 – 70 years were included in this study. These patients were operated through posterior approach of the cervical spine and then followed for six months for postoperative outcome .The patients' demographic details and clinical manifestation were entered into a semi structured proforma. Data was analyzed through statistical program SPSS version 11.*

***Results:*** *Out of 41 patients, there were 22 (53.6%) males and 19 (46.3%) females. The age of patients ranged from 20 to 70 years. In this study the overall mean age was 39.2 years. Majority of patients 19 (46.3%) were in the age range of 31 – 40 years. Most common clinical presentation of patient were quadrapresis in 23 (56.1%) pain neck 9 (22%) paraparesis 9 (22%) patients. Postoperatively most of the patients recovered from their preoperative symptoms.*

***Conclusions:*** *Quadraparesis and pain neck were the common clinical presentation of cervical cord neurofibroma type 1. Surgical outcome of cevical neurofibroma type 1 is good.*

***Key words:*** *Cervical neurofibroma type 1, spinal tumor, cervical tumor.*

***Abbreviations:*** *NFI: Neurofibromatosis type 1, NF2: Neurofibromatosis type 1, CNS: Central nervous system.*

## INTRODUCTION

The neurofibromatosis are genetic disorders that give rise to tumors of the nerve sheath, glia, and meninges. **Neurofibromatosis type 1 ($NF_1$)**, previously known as von Recklinghausen's or peripheral neurofibromatosis, is characterized by multiple cafe-au-lait spots, neurofibromas, axillary freckling, and iris hamartomas. It is an autosomal – dominant disorder affecting about one in 4000 individuals. **Neurofibromatosis type 2 (NF2)**, previously known as bilateral acoustic or central neurofibromatosis, is characterized by multiple spinal and intracranial tumors, particularly bilateral eighth nerve schwannomas (acoustic neurinomas). It is also an autosomal dominant disorder with a high penetrance, but it occurs less frequently.[1-3]

The gene for **neurofibromatosis type 1** is located near the centromere on the long arm of chromosome **17**, and the **neurofibromatosis type 2** locus is near the center of the long arm of chromosome **22**.[2]

**Benign nerve sheath tumors** of the spinal nerve roots commonly occur sporadically in both types of neurofibromatosis. These tumors may not be distinguished radiologically or at the time of surgery, but are classified pathologically either as neurofibromas or schwannomas. Although the precise cell of origin is uncertain, both tumors have prominent Schwann cell

components.[2,4,5]

$NF_1$ has a better prognosis with a lower incidence of CNS tumors than $NF_2$. However, morbidity and mortality rates in $NF_1$ are not negligible.[5,6] Some of the more severe complications are visual loss secondary to optic nerve gliomas, spinal cord tumors, scoliosis, vascular lesions, and long – bone abnormalities, which sometimes necessitate amputation.[5-7]

The purpose of our study was to determine the clinical presentation and postoperative outcome of cervical neurofibroma type 1.

## MATERIAL AND METHODS

This was a retrospective review of 41 cases involving NF-1 patients with symptomatic cervical spine neurofibromas who underwent surgical decompression and tumor resection, from 1st February 2001 to 31st July 2011, at Hayatabad Medical Complex Hospital Peshawar Khyber Pakhtoonkhawa. These patients were operated through posterior approach of the cervical spine and then followed for six months for postoperative outcome. The patients' demographic details; clinical manifestation and postoperative outcome were entered into a semi structured proforma.

Data was analyzed through statistical program SPSS version 11. All the qualitative variables like gender, quadrapresis, paraparesis, neck pain and urinary incontinence were analyzed for percentages and frequencies. Mean + standard deviation was calculated for quantitative variable like age. The results were presented through tables.

## RESULTS

### Sex Incidence

Out of 41 patients, there were 22 (53.6%) males and 19 (46.3%) females.

**Table 1:** Sex Incidence.

| Sex | No. | % |
|---|---|---|
| Male | 22 | 53.6 |
| Female | 19 | 46.4 |
| Total | 41 | 100 |

### Age Incidence

The age of patients ranged from 20 to 70 years. In this study the mean age was 39.2 years. Majority of patients 19 (46.3%) were in the age range of 31 – 40 years (Table 2).

**Table 2:** *Age Distribution.*

| S. No. | Age Group | Number of Patients | Percentages % |
|---|---|---|---|
| 1. | 20 – 30 | 8 | 19.5 |
| 2. | 31 – 40 | 19 | 46.3 |
| 3. | 41 – 50 | 7 | 17.1 |
| 4. | 51 – 60 | 5 | 12.2 |
| 5. | 61 – 70 | 2 | 4.9 |
| | Total | 41 | 100 |

### Clinical Presentation

Most common clinical presentation of patient was quadrapresis in 23 (56.1%) paraparesis 9 (22%) pain neck 9 (22%) patients (Table 3).

**Table 3:** *Clinical Presentation.*

| S. No. | Clinical Presentation | Number of Patients | Percentages (%) |
|---|---|---|---|
| 1. | Quadrapresis | 23 | 56.1 |
| 2. | Paraparesis | 9 | 22 |
| 3. | Neck pain radiating to upper limb | 9 | 22 |
| 4. | Urinary incontinence | 2 | 4.9 |

## DISCUSSION

NF-1 is one of the most common autosomal disorders of the central nervous system. The genetic locus of the disease is on the long arm of chromosome 17. The incidence of central nervous manifestations in NF-1 is about 15%. The spinal manifestations of the disease are scoliosis, intramedullary tumors (mostly astrocytoma), dural dysplasia, meningoceles, and nerve sheath tumors. $NF_1$ has been considered be an alternative form of neurofibromatosis, showing multiple spinal tumors and cafe´-au-lait spot, occuring in several members of the same family.[3,5,7,8]

These lesions present as slowly progressive masses that can either compress to the adjacent vital struc-

tures or interfere with normal physiology. Common musculoskeletal impairments associated with neurofibromatosis type 1 ($NF_1$) include cervical pain, muscle weakness, muscle stiffness, headaches, and postural deviations.Cervical cord compression due to spinal neurofibroma is a rare but important complication. The early diagnosis is important for preventing irreversible quadriplegia.[6,7,9]

In our study age range was from 20 to 70 years. The mean age of presentation was 39.2 years. A study was conducted by Taleb FS showed mean age of 42.5 years.[10] Leonard JR showed the mean age 29.9 years.[11]

In this study most of the patients were male 22 (53.6%). Ma J also reported male predominance in his study.[12] Scheithauer BW also documented male predominance.[13]

Clinical presentation of the patients in our study were quadrapresis in 23 (56.1%) patients, paraparesis in 9 (22%) patients, neck pain radiating to upper limb in 9 (22%) patients and urinary incontinence in 2 (9.2%) patients. This means that the common clinical presentation was quadrapresis. Taleb FS showed the same result.[10] Matti T reported that quadraparesis is the common clinical presentation of cervical neurofibroma type 1.[14]

Postoperatively most of the patient's 31 (75.6%) symptoms improved fully, only nine (21.9%) patients having some neurological deficit and one (2.4%) patient died. Halliday AL showed good recovery postoperatively.[15] Matti T showed the good outcome of surgery but the mortality was 15.7%, this might be due to long follow-up of their study.[14] Leonard JR documented that majority of their patients improved postoperative from their symptoms.[11]

## CONCLUSIONS

Quadraparesis and pain were the common clinical presentation of cervical cord neurofibroma type 1. Surgical outcome of cervical neurofibroma type 1 is good.

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