ABSTRACT

Background: Cervical sub-axial dumbbell neurofibromas (NFs) account for nearly 20% of all NFs, with prognosis depending on the extent of excision. When majority of tumor is extra-foraminal (Tomaya’s Type IIb and c), certain unconventional anterolateral or posterolateral neck approaches are used for maximum safe excision. In our article, we provide a brief review of the literature regarding various surgical approaches, emphasizing the utility of posterolateral or combined anterior and posterior neck approaches for such giant NF.

Methods: We performed a retrospective analysis of prospectively maintained surgical databases, from our hospital discharge codes, for all cervical Type IIb-c dumbbell NF patients, who underwent surgery at our institution between 2015 and 2019. Clinical variables included age at admission, clinical presentation, and surgical difficulties, and the outcome was analyzed.

Results: Four patients of age ranging from 22 to 45 years (M:F 3:1) were operated by posterolateral (n = 3) and combined anteroposterior (n = 1) approach. Three patients underwent near-total excision and one patient had total excision. One patient with tumor capsule attached to roots of upper brachial plexus had motor deficit, who was re-admitted for neurotization. There was no intraoperative complication.

Conclusion: Posterolateral approach for cervical dumbbell giant NFs is safe, effective, and promises maximum safe excision. The major blood vessels including carotid artery or jugular veins in neck and vertebral artery at foraminal portion are directly under vision and control of surgeon. Despite near-total excision, wherein small part of adhered capsule is left, recurrence rate is low.

Keywords: Cervical intradural extramedullary, combined anteroposterior single-stage approach, dumbbell neurofibromas, posterolateral triangle of neck

INTRODUCTION

The variability in terms of location, extent, and size of a dumbbell cervical spinal neurofibroma (NF) possesses unique technical challenges for the surgeon. It is a generally accepted belief that the nerve rootlet/root attached is not separable from the tumor, and complete excision is nearly impossible without associated deficit. The dumbbell NF is classified into nine types on the basis of extraforaminal extension.[1] Similar classification was given by Goel et al. for C2 peripheral nerve sheath tumors based on the location and extent.[2] The Type I (within spinal canal) and Type IIa (extension up to intervertebral foramen) NF are usually approached through conventional hemi-laminectomy or open-door laminoplasty.[1] The Type IIb-c possesses a surgical challenge because majority of tumor is extra-foraminal.
of tumor lies in the extraforaminal region. The anatomical knowledge of posterolateral neck triangle and surgical corridors is needed for maximal safe excision. Being a benign disease, adjuvant radiotherapy is not recommended, therefore surgical expertise offers cure. The part of tumor extending in posterolateral or anterolateral triangles of neck demands multidisciplinary approach for maximum safe excision. In delayed presentations, when the extraforaminal part becomes giant cosmetically disfiguring with vertebral artery (VA) nearly encased, the approach demands extra vigilance. The surgeon should understand the nuances and sequelae of these unconventional corridors. Although the proximity to carotid artery, vagus nerve, and internal jugular vein increases complexity level, the anterolateral or posterolateral corridors obviate the need of facetectomy and therefore additional stabilization procedure is not required.\[3\] In this article, we provide a brief review of the literature supporting the importance of revisiting the neck approaches for giant cervical Type IIb-c dumbbell NF.

METHODS

We performed a retrospective analysis of prospectively maintained surgical databases, from our hospital discharge codes, for all cervical Type IIb-c dumbbell NF patients, who underwent surgery at our institution between 2015 and 2019. Clinical variables included age at admission, clinical presentation, and surgical difficulties, and the outcome was analyzed. Informed consent was obtained from all individual participants included in the study.

RESULTS

Case 1
A 22-year-old male patient was admitted with complaints of painless swelling in the right side of neck, which was gradually increasing in size for the last 3 years. On examination, there was a 6 cm × 5 cm swelling, in the posterior triangle of the neck. The swelling was nodular, firm in consistency, and there was restricted mobility in vertical direction. There was no neurological deficit on examination. The incisional biopsy, done elsewhere, was suggestive of benign mesenchymal tumor. Magnetic resonance imaging (MRI) showed a well-defined, T1-weighted imaging (WI) isointense, 6 cm × 5 cm × 3 cm mass lesion, in the paraspinal region [Figure 1a]. The lesion was extending from C2-C3 intradural region, through neural foramina, to the posterior triangle of neck (Type IIc). The strap muscles were draped around the tumor [Figure 1b-e]. The contrast images showed peripheral enhancement with central necrotic areas. The tumor was moderately vascular, approximately 6 cm × 4 cm × 3 cm in size, firm, nodular whitish mass in the right posterior triangle area, extending into C2-C3 neural foramina. There were dense adhesions between previous scar of biopsy site and soft tissues. Complete tumor excision was achieved [Figure 1f]. The patient was discharged on postoperative day (POD) 5 and is under follow-up for 8-months without any deficit.

Case 2
The second patient was a 43-year-old male patient, who had similar complaint of gradually progressive painless swelling in the right side of the neck for the last 4 years, along with tingling paresthesia along the distribution of the right median nerve. Fine-needle aspiration cytology was suggestive of benign mesenchymal tumor. There was no neurological deficit on examination. Computed tomography showed a well-defined, oval, isodense to hypodense swelling of size 4.5 cm × 3.8 cm × 4.1 cm, in the right neck, beneath the right sternocleidomastoid (SCM). The carotid artery and internal jugular vein were pushed anteromedially, and the swelling extending superiorly till thyroid cartilage and inferiorly till thoracic inlet (Type IIc). The surgical excision was planned electively, wherein, a 4 cm × 4 cm × 3.8 cm swelling, arising deep into the clavicular head of SCM, extending from the posterior trunk of brachial plexus, was dissected out in total [Figure 2a-h]. The near-total excision of tumor was done leaving behind a capsule along the posterior trunk of brachial plexus (from which it was arising). Postoperatively, the patient developed motor deficit. A second-stage neurotization procedure was done to restore functions. The patient is under 14-month follow-up.

Case 3
Our third patient, a 25-year-old female, was admitted with the complaint of gradually progressive painless swelling in the left side of the neck for the last 2 years (Type IIc). There was no neurological deficit on examination. On examination, there was a single swelling, approximately 8 cm × 10 cm size, over the left lateral aspect of the neck, behind the posterior border of SCM. The swelling was firm and tender, mobile in anteroposterior direction but not in superoinferior direction. Intraoperatively, the tumor was seen lateral to the carotid sheath, in the posterior triangle of the neck [Figure 3a-f]. The tumor was well encapsulated, yellowish red in color, and moderately vascular. The phrenic nerve was identified and was stretched over the capsule. The upper trunk of brachial plexus was seen in the inferomedial relation and invested in the capsule. A part of tumor capsule was left behind along the phrenic and vagus nerve, and rest of the tumor along with the capsule was excised. The patient was discharged on POD 5 and is doing well after the 7-month follow-up.
Case 4
A 40-year-old male was previously operated for C4-5 NF 10 years back. The patient was admitted with complaints of gradually progressive painless swelling in the left side of the neck and tingling paresthesia in the fifth cervical dermatome for the last 4 years. On neurological assessment, the contour of the left shoulder was lost with atrophy of the left thenar and hypo-thenar eminences of the wrist. The tone was increased in all the four limbs (modified Ashworth grade 2), and power in the left upper/lower limb was slightly less (4/5), with left grip being 70%–80% compared to the right side. The MRI was suggestive of a well-defined dumbbell-shaped lesion of size 5 cm × 7 cm × 2.5 cm in the left paravertebral aspect of C4 and C5 vertebra level with widening of neural foramen (Type IIb). The lesion was T1-WI hypointense and T2-WI hyperintense and had homogenous intense contrast enhancement. The lesion was extending into the spinal canal, leading to cord compression. A combined anteroposterior approach was planned [Figure 4a and b]. In the initial part of surgery, the patient was positioned prone, and previous laminectomy incision was re-opened [Figure 4c].

Figure 1: Magnetic resonance imaging sagittal view showing a well-defined, T1-weighted imaging heterointense mass lesion extending from the C2-C3 intradural region, through neural foramina, to the posterior triangle of neck (a). The patient was positioned in supine lateral with previous biopsy incision mark seen (b). Intraoperative photographs showing how sternocleidomastoid and strap muscles draped the tumor (c) and capsular dissection was done all around (d). The tumor was excised in-total (e). Follow-up magnetic resonance imaging showing total excision of tumor (f).

Figure 2: The patient was positioned supine-lateral with head slightly extended (a) and skin incision along the posterior border of sternocleidomastoid was made extending laterally (b) The sternocleidomastoid was drapped around the tumor and pushed superiorly (c) which was dissected (d) and the lateral border of tumor lifted off the skin (e) Then, sternocleidomastoid was dissected along its inferior surface and pulled laterally to make space between tumor and muscle (f). The carotid artery and internal jugular vein were pushed anteromedially, and the swelling extending superiorly till thyroid cartilage and inferiorly till thoracic inlet (g). The near-total excision of tumor was done leaving behind a capsule along the posterior trunk of brachial plexus (h).
The intradural part of the tumor was adherent to the adjacent nerve roots and was pushing the cord to the right side. The tumor was removed in piecemeal fashion, and near-total excision was done. The tumor was also extending extradurally through the left fifth cervical neural foramina. The tumor was dissected off the surrounding tissue and VA. The part of the tumor which was lateral to the VA was excised, and Gelfoam® was placed there for identification of the last part of the tumor when we would approach it form the anterolateral approach. After excision of the posterior part of the tumor, the patient was turned supine and cervical incision was given. Left-sided “Boomrang skin incision” was given, and the tumor was identified posterior to the SCM [Figure 4d]. The tumor was splaying scaleneus anterior and scaleneus medius muscles and was pushing the brachial plexus anteriorly. Postoperatively, there was improvement in spasticity, and the
patient was discharged on POD7. The patient is doing well in the outpatient department at 9-month follow-up, without any deficit [Figure 4e].

**DISCUSSION**

Spinal NF (SNF) is a benign, well-defined, encapsulated tumor, usually arising from the dorsal nerve roots. It is commonly present in the fourth and fifth decades of life. Nearly 80%–85% of them remain intradural, but an unfavorable subset spreads extradurally and forms a dumbbell-shaped mass. McCormick described dumbbell SNF as tumors with contiguous intraspinal, foraminal, or extraforaminal spread. The narrow spinal canal diameter and close proximity to VA, ranks cervical SNF as technically challenging and demanding surgery. Surgery is indicated for symptomatic SNF or tumors associated with neurofibromatosis Type II. The common posterior laminctomy approach for SNF has been well described before, however, surgical nuances and corridors for ventrally located tumors or extensive extradural SNF remain relatively untouched. With the rising acceptance of minimally invasive spine surgery, even small size SNFs have gained re-popularity. In our series, we intend to describe and review the unconventional corridors for sub-axial cervical SNF.

The surgical strategy depends on the epicenter of the tumor, relation to the denticulate ligament, and extent or size of the extradural part. Small-sized, dorsally situated SNFs are approached posteriorly by the conventional laminectomy or hemi-laminectomy approach. A tumor having ventral epicenter needs facetectomy or other corridors. The intervertebral foramen forms the bottleneck for extradural SNF, and the disconnecting intra- and extra-dural part at this site forms one of the critical intraoperative steps in combined approach. Asazuma et al.’s classification of the tumors, in three-dimensional plane, forms the backbone in preoperative planning. Another important point to be taken care of is the cranio-caudal spread, i.e., number of intervertebral foramen involved. It has been proved that complete excision of SNF, although challenging, results in better prognosis and disease-free survival. Dumbbell tumors have more propensity for recurrence owing to inadequate excision. Complex approaches, even like vertebrectomy, have been described in literature, but had serious complications such as vascular injury or postoperative pain. Complete excision of SNF has been achieved using anterior or lateral approach with/without facetectomy, lateral approach with VA ligation, and combined anteroposterior approach for cervical SNF Type IIb, IIC, IIIb, and VI.

The principles in traditional approach for dorsal SNF have not changed since years, although the endoscopic modification transformed the surgical outcome. Zhu et al. used similar approach for lumbar SNF. The incision was made on fluoroscopic guidance, and disc space was identified. Then, Destandau’s endoscopic sheath is introduced and docked over the lamina at desired level. NF was dissected all around the arachnoid-capsule plane, and the nerve root attachment was coagulated and cut. Patient selection and familiarity with narrow endoscopic corridor is a prerequisite. St Clair et al. reported endoscopic-assisted spinal cord decompression in metastasis, where they did vertebrectomy and spinal stabilization also. In a series of 18 cases, Parihar et al. operated four cases similarly, where they excised the tumor completely. The complex approaches for dumbbell SNF are technically demanding and must be balanced on risks and possible morbidity. The original lateral approach to SNF, as described by Verbiest, defined corridor lateral to carotid sheath. Even detachment of SCM muscle has been mentioned. However, in our experience, we believe that SNF with large extraforaminal extension actually splays SCM and scalene muscles and grows away from the carotid sheath. We took corridor lateral to SCM muscle and anterior margin of trapezius. In a series of 57 cases, Asazuma et al. described combined anteroposterior approach in seven cases, five out of which were Type IIIb. They described the skin incision along the posterior margin of SCM muscle for anterior approach, but did not discuss the intraoperative nuances and rationale. We took a modified curvilinear incision along the posterior margin of SCM with inferior edge curving over the clavicle. These SNFs are usually attached or arise from a single rootlet, and cutting these nerve root, which is usually sensory, does not increase the chance of postoperative neurological deficit. Even some authors believe that the rate of recurrence is higher on leaving the attached nerve root. In our cases II and III, we were able to identify the attached nerve root extracapsular and then made incision anterior to the attachment, for decompression. We preserved the attached root and even the capsule of tumor adhered, but still one of our patients developed postoperative transient weakness. Intraoperative neuromonitoring is a suitable adjunct, and its use might further reduce the risk of postoperative deficits. In another series of 367 patients, 5 patients in the subaxial cervical region were operated using anteroposterior approach, while one case in the thoracic region needed transthoracic approach. We have focused our discussion on sub-axial cervical SNF only to prevent dilution of topic. In our case IV, we resected intradural and intraspinal part initially till intervertebral foramen and then turned the patient supine and went between corridor described above. Here, we want to emphasize that in the superior part of the posterior triangle of the neck, individual scalene muscles, originating from different cervical vertebra, are seen, which
continues as a single bundle, in the inferior part of the triangle. Therefore, when tumor grows, SCM along with single bundle of scalene is pushed anteromedially. In the superior part of triangle, scalene muscle needs blunt dissection to delineate the tumor capsule.

The limits of the tumor capsule is not always obvious and may be confused with the periosteal sheath, therefore both proximal and distal control of VA is necessary. The VA is controlled by resecting the anterior part of the transverse processes situated below and above the involved level. In a series by Lot et al., the authors exposed the involved cervical nerve root at the tip of the transverse process, distal to the lateral part of the tumor.\(^{12}\) In this, VA was occluded in a case of neurofibromatosis.\(^{12}\) We believe that this part is not so tricky, if one maintains the dissection over the capsule of NF. The lower part of tumor generally pushes brachial plexus trunk inferiorly, as in our case II, but still some branches from the root of C5–6, may be found draped over the tumor capsule anteriorly.

Lot et al. described the technique of oblique drilling for dumbbell tumors.\(^{12}\) In their series of 57 cases, 38 patents of sub-axial spine were operated by anterolateral corridor, while posterolateral corridor was described for C1–C2 SNF. For anterolateral approach, Lot et al. preferred the incision along the anterior margin of SCM. They preferred the corridor between SCM and internal jugular vein.\(^{12}\) The transverse process was exposed by resecting paravertebral muscles, and sympathetic chain was preserved rolling along the aponeurosis. An early description by McCormick for anterior approach also describes the division of anterior scalene and longus capitus muscle to expose the anterior tubercle of transverse process and costal process.\(^{20}\) They included all spinal tumors, so the plane of dissection might be different; fortunately, in all our four cases, we found good capsular plane. We decompressed the tumor using an ultrasonic aspirator, and then the capsule was dissected further to find VA. The drilling of vertebral body was not needed in any of our case, despite the giant size of the tumors. The shifted relation of VA needs meticulous dissection and prior radiological assertion. Lot et al. also described the excision of intradural part of dumbbell SNF in anterolateral approach by oblique drilling of vertebral body, as done for cervical spondylisis. Ariff et al. reported a similar case of dumbbell sub-axial SNF, which was operated in two stages, 1 month apart.\(^{13}\) They preserved interspinous and supraspinous ligaments, and decompressed intraspinal part initially, but did not describe the details of anterior approach. The dumbbell SNF most often originates from the nerve root inside the intervertebral foramen; consequently, along the tumor development, the VA is displaced anteromedially. However, it may happen that probably due to the origin of the tumor from the distal and extraspinal parts of the nerve root, the VA is pushed inside the intervertebral foramen.\(^{14}\) Sometimes, a radicular artery, branch of VA, supplies the tumor and enhances the difficulty for surgeon. Some authors prefer preoperative angiogram to define all feeders. The paravertebral plexus bleed is time-consuming and sometimes frustrating. However, this is a technique that is not familiar to most neurosurgeons, who need some training before feeling comfortable with it.

We believe that in the anterolateral approach, the exposure of all parts of dumbbell tumors, no matter whatever is their extensions (extraspinal, extradural, and intradural), can be excised, with added advantage that the spinal stability is not compromised as the joints are fully preserved. Considering the long intraoperative time and blood loss, some authors propose lateral corridor that combines laminectomy and sectioning of the paraspinal muscles through a transverse incision, which is considered a simpler technique with excellent exposure of the lateral aspects of the spine, facilitating complete removal of tumor.\(^{15}\)

Another approach described is the posterior subscapular approach, wherein the trapezius is transacted and the levator scapulae with rhomboids are dissected in a segmental fashion. The thicker portion of the trapezius can be split in a medial direction. In addition, some of the serratus posterior muscle may also be divided. The first rib is removed, and Weitlaner retractor is placed under the second rib and soft tissue of the superior neck to open up the supraclavicular space posterior to the plexus.\(^{13}\) For lesions extending intraforaminally, a posterior foraminotomy is performed. The facet joint can also be removed to expose the intraforaminal course of nerve and tumor.

In the case of Types I–III, wherein, with limited development into the extraforaminal part, the posterolateral approach is considered a good option. In contrast, extraspinal tumor with limited extension into the foramen, anterior-lateral is considered as better approach.\(^{14}\) The posterolateral approach, described in literature, reported joint destruction and the additional needs of stabilization. The combined anterior and lateral approach as described by Hakuba et al. needs a discectomy and the drilling of the adjacent vertebral bodies followed by bone grafting.\(^{16}\) Jiang et al. reported that patients who undergo facetectomy without fusion have a 50% risk of scoliosis or kyphoscoliosis, with or without neurological deficit.\(^{17}\) Our case (case IV), Type IIb, the extraspinal part, was extending from the neural foramen, directly widening it but not de-stabilizing it. Hence, the
corridor was lateral, and the incision was given over the major bulk of the tumor. After dissecting the fat and Level V lymph nodes, the capsule was seen and the tumor was detached at the attachment to dura. We left a small part of tumor, but need of second-stage stabilization or facetectomy obviated. The patient had significant improvement in radicular pain. Complete excision is not always possible and neither needed; unless the proliferation rate is high (MIB-1 index, >5%), a second-stage surgery in not required.\(^\text{[6]}\)

**CONCLUSION**

In the Type II SNF, there is giant extraforaminal position of NF. The surgical access to these tumors should be taken from posterolateral or anterolateral triangles of neck. Posterior approaches utilize facetectomy and de-stabilize the spine with additional need of instrumentation. The posterolateral approach for cervical dumbbell giant NF is safe and effective and promises maximum safe excision. The major blood vessels including carotid or jugular in neck and vertebral at foraminal portion are directly under vision and control of surgeon. Even with near-total excision, wherein a small part of the adhered capsule is left, the recurrence rate quoted in literature is low.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Asazuma T, Toyama Y, Maruwa H, Fujimura Y, Hirabayashi K. Surgical strategy for cervical dumbbell tumors based on a three-dimensional classification. Spine (Phila Pa 1976) 2004;29:E10-4.
2. Goel A, Muzumdar D, Nadkarni T, Desai K, Dange N, Chagla A. Retrospective analysis of peripheral nerve sheath tumors of the second cervical nerve root in 60 surgically treated patients. J Neurosurg Spine 2008;8:129-34.
3. Zhu YJ, Ying GY, Chen AQ, Wang LL, Yu DF, Zha LL, et al. Minimally invasive removal of lumbar intradural extramedullary lesions using the interlaminar approach. Neurosurg Focus 2015;39:E10.
4. Lenzi J, Anichini G, Landi A, Piecoci A, Passacantilli E, Pedace F, et al. Spinal nerves schwannomas: Experience on 367 cases-historic overview on how clinical, radiological, and surgical practices have changed over a course of 60 years. Neurol Res Int 2017;2017:3568359.
5. McCormick PC. Surgical management of dumbbell tumors of the cervical spine. Neurosurgery 1996;38:294-300.
6. Nakamura M, Iwanami A, Tsuji O, Hosogane N, Watanabe K, Tsuji T, et al. Long-term surgical outcomes of cervical dumbbell neurinomas. J Orthop Sci 2013;18:8-13.
7. Mahore A, Chagla A, Goel A. Giant ventral midline schwannoma of cervical spine: Agonies and nuances. J Korean Neurosurg Soc 2010;47:454-7.
8. Mahore A, Muzumdar D, Chagla A, Goel A. Pure ventral midline long segment schwannoma of the cervicodorsal spine: A case report. Turk Neurosurg 2009;19:302-5.
9. St Clair SF, McLain RF. Posterolateral spinal cord decompression in patients with metastasis: An endoscopic assisted approach. Surg Technol Int 2006;15:257-63.
10. Parihar VS, Yadav N, Yadav YR, Ratre S, Bajaj J, Kher Y. Endoscopic management of spinal intradural extramedullary tumors. J Neurol Surg A Cent Eur Neurosurg 2017;78:219-26.
11. Verbiest H. A lateral approach to the cervical spine: Technique and indications. J Neurosurg 1968;28:191-203.
12. Lot G, George B. Cervical nerves with extradural components: Surgical management in a series of 57 patients. Neurosurgery 1997;41:813-20.
13. Mohd Ariff S, Joehainey J, Ahmad Sabri O, Abdul Halim Y. Two-stage surgery for a large cervical dumbbell tumour in neurofibromatosis 1: A case report. Malays Orthop J 2011;5:24-7.
14. George B, Lot G. Surgical treatment of dumbbell neurinomas of the cervical spine. Crit Rev Neurosurg 1999;9:156-60.
15. Onesti ST, Ashkenazi E, Michelsen WJ. Transapical spinal canal exposure of dumbbell tumors of the spine. Report of two cases. J Neurosurg 1998;88:106-10.
16. Hakuba A, Komiyama M, Tsujiimoto T, Ahn MS, Nishimura S, Ohta T, et al. Transcudiscal approach to dumbbell tumors of the cervical spinal canal. J Neurosurg 1984;61:1100-6.
17. Jiang L, Lv Y, Liu XG, Ma QF, Wei F, Dang GT, et al. Results of surgical treatment of cervical dumbbell tumors: Surgical approach and development of an anatomic classification system. Spine (Phila Pa 1976) 2009;34:1307-14.