Interdisciplinary Neurosurgery 18 (2019) 100481

Contents lists available at ScienceDirect

Interdisciplinary Neurosurgery

journal homepage: www.elsevier.com/locate/inat

Case Reports & Case Series

Excision of dumbbell shaped ganglioneuroma of cervical spine using a facet preserving inside out approach - What we know and what we learnt

Harsh Deora (MCh)[a], Ashutosh Kumar (MS)[b], Kuntal Kanti Das (MCh)[b,*], Bitan Naik (MD)[c], Priyadarshi Dikshit (MCh)[a], Arun Srivastava (MCh)[b], Sanjay Behari (MCh)[b]

[a] Dept. of Neurosurgery, National Institute of Mental Health and Neurosciences (NIMHANS), Bangalore 560029, India
[b] Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical sciences, Lucknow, Uttar Pradesh 226014, India
[c] Department of Pathology, Sanjay Gandhi Postgraduate Institute of Medical sciences, Lucknow, Uttar Pradesh 226014, India

ARTICLE INFO

Keywords:
Ganglioneuroma
Dumbbell cervical tumors
Hemi-laminectomy
Primitive neuroectodermal tumors

ABSTRACT

Introduction: Cervical benign lesions present a unique opportunity to provide significant improvement in the quality of life of the patient along with the promise of a cure. Critical neurovascular structures are nearby and must be kept in mind along with the minimal bone loss to avoid postoperative instability. Ganglioneuroma of the cervical spine presents such a unique opportunity where complete excision often results in a cure.

Case report: We report a case of 57 year old male with a dumbbell-shaped ganglioneuroma with an intra-dural extension which was operated via a C6–C7 hemilaminectomy and facet preserving inside-out technique under neuro-physiological monitoring and also report the all the previous such reported cases via a thorough literature search. With only 15 such cases previously reported, the traditional teaching of a rare intradural extension has now been reversed with majority of the cases having an intradural extension and dumbbell-shaped tumors.

Conclusion: Although childhood in origin they present in adulthood due to their slow growth rate and multiplicity and recurrence are rare unless associated with familial syndromes. Knowledge of this rare pathology increases the index of suspicion and thus dictates the outcome of the case.

1. Introduction

Ganglioneuromas are rare forms of benign tumors that are pathologically the mature forms of neuroblastoma and can occur anywhere where sympathetic tissue exists. Consequently, they are commonly found in the adrenal medulla, retroperitoneum, posterior mediastinum, pelvis and, like in our case, the neck. The cervical spine is the third most common location of this rare tumor after the retroperitoneum and posterior mediastinum. Still, there have been only 15 such cases reported in the literature in the cervical spine [1–15] and thus the knowledge reserve is scarce for this lesion. There are cases in association with neurofibromatosis [2] and some that are dumbbell shaped with intra-dural extension too. We attempt to present a snapshot of all such reported cases and the myriad of treatments offered along with a similar one we were able to treat.

2. Case report

A 57 year old male, right-handed, was admitted with complaints of gradually progressive quadriparesis for 3 years with pain in nape of the neck. The weakness started from the right upper limb about six months ago when he reported difficulty in writing or holding a pen and gradually progressed to the right lower limb and eventually involved all four limbs. On examination, there was difficulty in the extension of the right elbow and grip weakness if the right hand. The triceps and supinator jerks were absent on the right side while all the

Abbreviations: MRI, magnetic resonance imaging; MRC, Medical Research Council; NF, neurofibromatosis

* Corresponding author.
E-mail address: kkdas@sgpgi.ac.in (K.K. Das).

https://doi.org/10.1016/j.inat.2019.100481
Received 22 April 2019; Received in revised form 5 May 2019; Accepted 18 May 2019

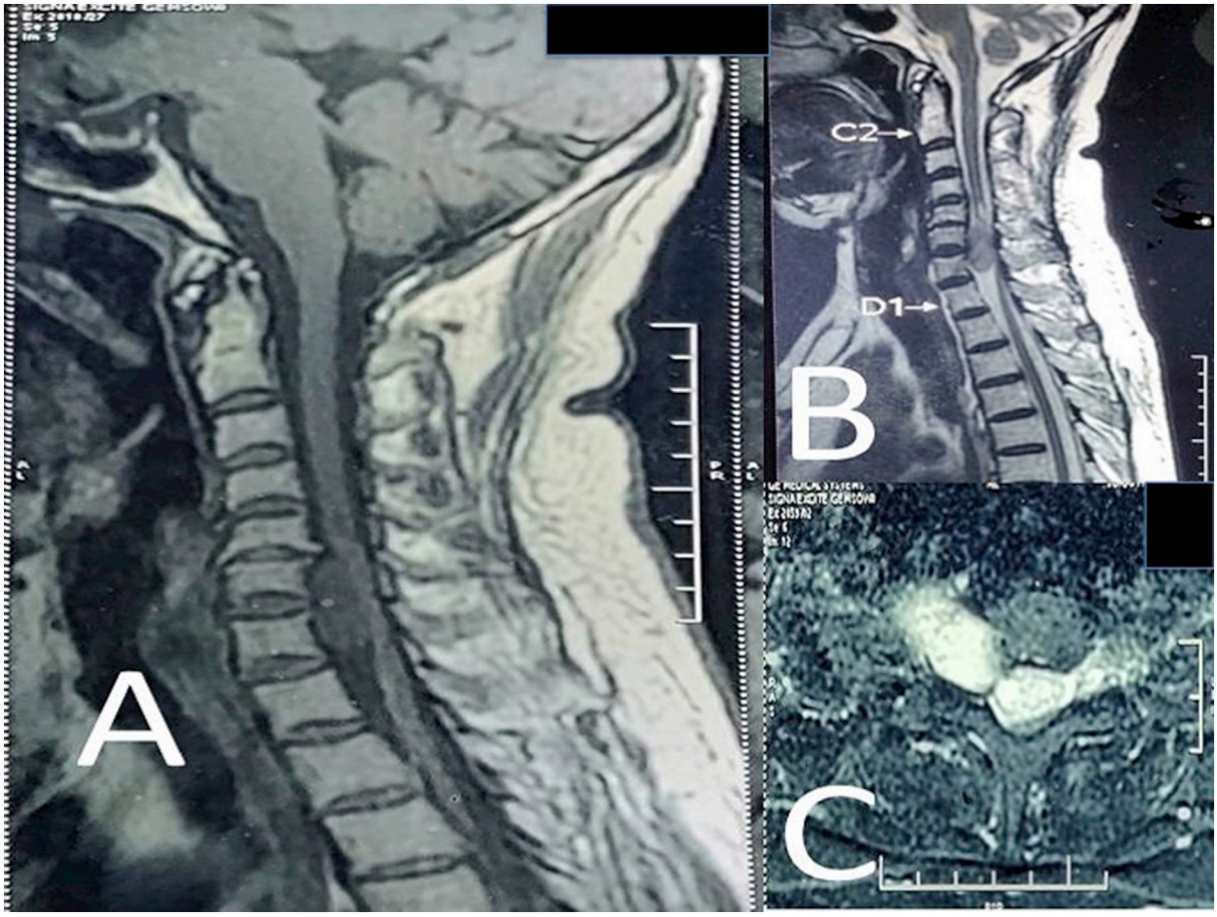

Fig. 1. T1 (A) and T2 (B) weighted saggital cuts of magnetic resonance imaging without contrast showing a mass in the cervical spine with severe compression of the cord and meniscus sign suggestive of an intradural extramedullary location. (C) T1 weighted axial cut of Magnetic resonance imaging with contrast showing extension of the lesion through the neural foramina and uniform contrast enhancement of the lesion.

other jerks of bilateral lower limbs were exaggerated and plantars were extensor. There was severe pain and sensory loss in the C6–C7 dermatomes but there were no disturbances in micturition or bowel movements. There were no Caif-au-lait spots in the body and there was no family history of neurofibromatosis. When the patient presented to us he was able to walk only with someone's help (Nurick Grade 4 and mJOA score-9). MRI with contrast of the cervical spine was suggestive of a well-defined intra-dural-extra-medullary lesion with trans-foraminal extension at C6 C7 level (Fig. 1). It was brilliantly contrast enhancing and T1 iso-intense, T2 hetro-intense. Based on clinical features and radiological findings, a working diagnosis of C6 neurofibroma was made and the patient was posted for surgery.

We tried to be minimalistic in our bone removal and thus a right C6–C7 hemilaminectomy was done to reach the lesion. Intraoperative electrophysiological monitoring was used while operating this case as the tumor was compressing the spinal cord dorsally at the C6–C7 level. Comprehensive multi-modal monitoring of MEP, SSEP and nerve-root-specific monitoring using EMG for the biceps (C6), brachioradialis (C6) and triceps (C7) muscles were done. The tumor was found in extradural space with an intra-dural and trans-foraminal extension. The extradural component was completely excised by opening the dura directly over it and the entire tumor was removed from within without removing the facet. Tilting the microscope allowed us an oblique corridor to chase this part of the tumor. Subsequenty, the dural incision on the extradural part of the tumor was extended on the dural sac over the cord. Then the intradural, spinal cord abutting tumor was removed. There was no change in the MEP or SSEP although the EMG in triceps did not trigger even on nerve stimulation suggesting motor root involvement by the tumor. Dura was closed using interrupted prolene 4-0 sutures and there was no leak intraoperatively.

Post-operatively, the patient had an improvement in spasticity and power in all limbs while the weakness in elbow extension and grip did not improve immediately in the right upper limb. The histopathological report was that of an encapsulated lesion with spindle-shaped cells with thin, curved, elongated nuclei with scant cytoplasm along with scattered mature ganglion cells. On IHC, the ganglion cells were positive for synaptophysin, S-100 and NSE. The spindle cells were positive for S-100. Ki67 index was < 1%. It was reported as ganglioneuroma (Fig. 2).

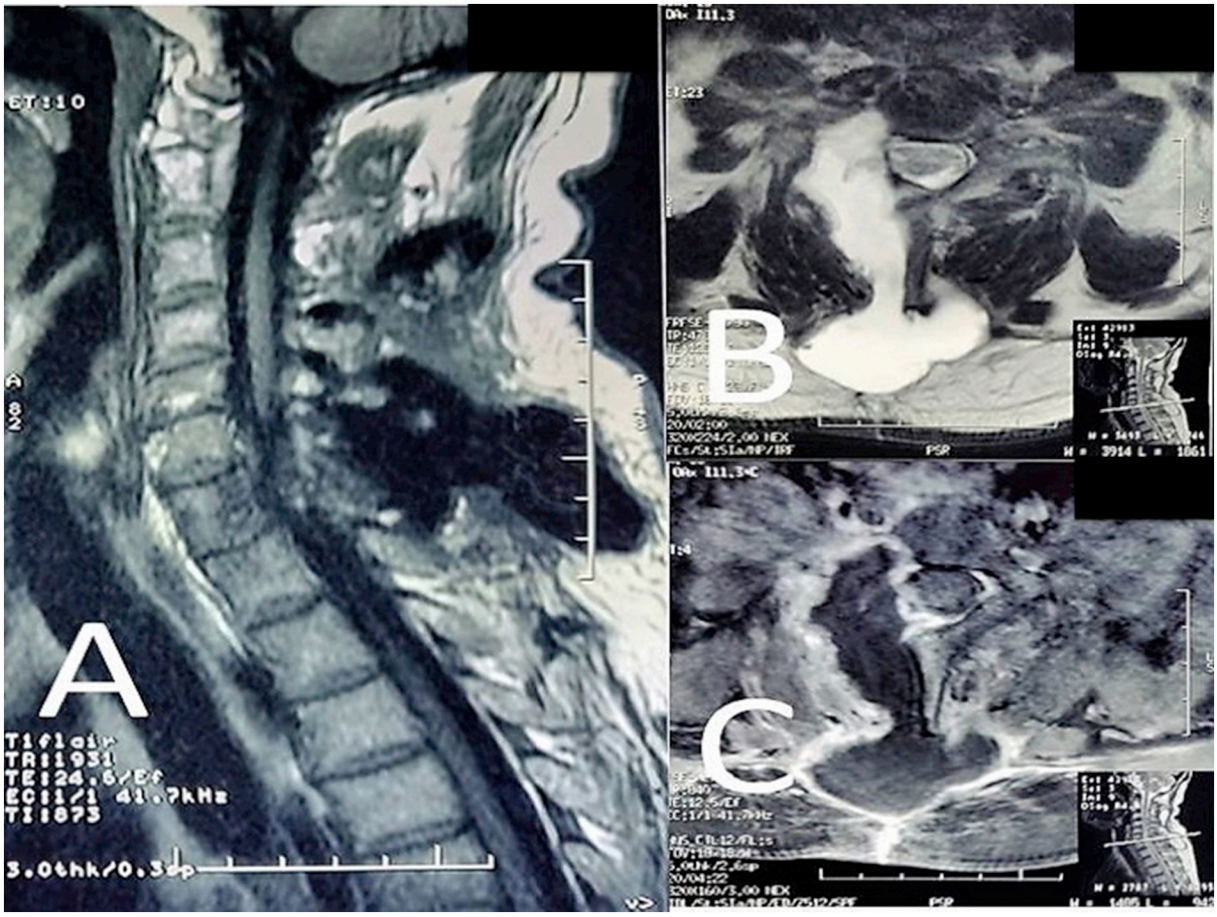

Fig. 2. Postoperative T1 (A) and T2 (B) weighted saggital images of magnetic resonance imaging without contrast showing complete excision of the lesion with formation of pseudomeningocoele. (C) T1 weighted axial cut of Magnetic resonance imaging with contrast showing no residual enhancement of the lesion indicating completeness of excision.

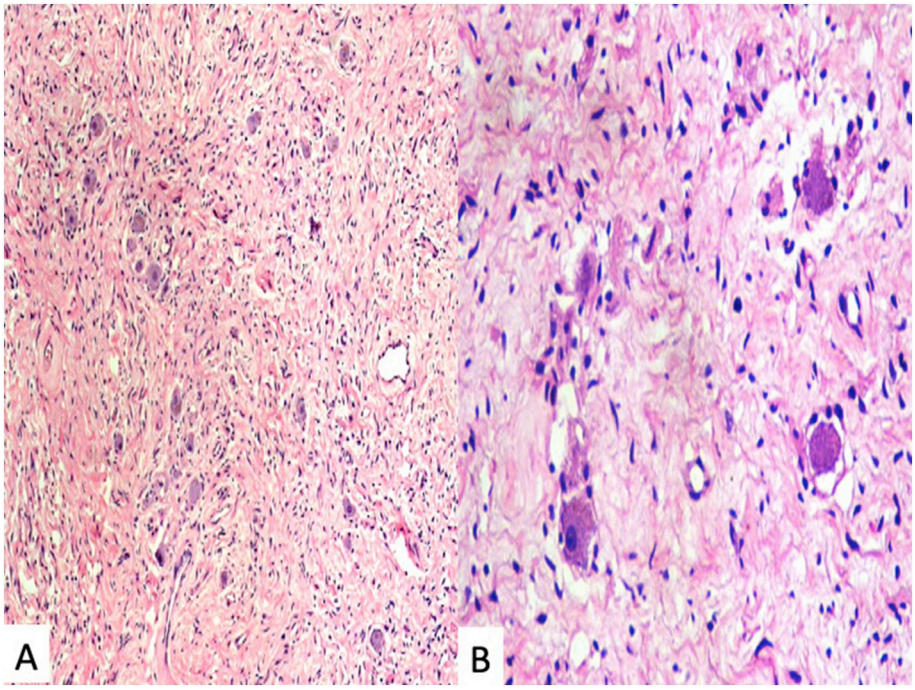

Fig. 3. H&E (×100) stained section shows tumor displaying predominantly spindle shaped cells with thin, curved to elongated nuclei and scant cytoplasm along with fair number of scattered mature ganglion cells (A). High power (×400) showing large number of spindle shaped cells with no evidence of mitosis indicating benign pathology (B).

Table 1
Comprehensive review of all cases of ganglioneuroma of the cervical spine and their management.

Author	Age (years)/sex	Site of origin	Dumbbell shape	Intraspinal extension	Multiplicity	NF-1	Surgical approach	Extent of excision
Bhand et al. [1]	22/F	Unilateral C3	+	Extradural	–	–	Cervical laminectomy from C-3 to C-7 level	Subtotal
Kyoshima et al. [2]	35/M	B/L C2 and C3	+	Intradural	+	+	Suboccipital partial craniectomy and wide laminectomy of C1 to C4	Subtotal
Shepherd and Sutton [3]	35/M	Unilateral C2–C7	+	Intradural	+	+	Laminectomy	Subtotal
Sinclair and Yang [4]	44/F	Unilateral C2–C5	+	Intradural	+	+	Suboccipital craniectomy with C1 to C5 laminectomy	Subtotal
Strang and Nordenstam [5]	63/F	Unilateral C2–C4	+	Intradural	–	–	C1–4 laminectomy	Total
Maggi et al. [6]	1.5/F	Unilateral C2–C6	+	Extradural	–	–	Laminectomy	Total
Radulovi et al. [7]	39/M	Unilateral C5	+	Extradural	–	–	C4 and C5 laminectomy followed by left lateral cervical approach	Subtotal
Tei et al. [8]	51/F	Unilateral C1	–	Intradural	–	–	Suboccipital craniectomy and C1–2 hemilaminectomy	Total
Ugarriza et al. [9]	53/M	B/L C2	+	Extradural	+	–	C1 arch and C2 lamina were removed	Total
Naohisa et al. [10]	15/M	B/L C2, unilateral C4	+	Extradural	+	+	Multilevel laminectomy	Subtotal
Zhang et al. [11]	24/F	Unilateral C6-C7	+	Extradural	–	–	C7 hemilaminectomy	Total
Son et al. [12]	13/M	Unilateral C2C3	+	Intradural	–	+	Hemi-laminectomy of C2 was done without arthrodesis.	Subtotal
Hioki et al. [13]	72/M	Bilateral C1C2	+	Intradural	–	–	Posterior arch at C1 and cranial portion of the C2 lamina was removed	Subtotal
Ando et al. [14]	20/M	Bilateral multiple C1TO C4	+	Intradural	+	+	Resection of the C1 posterior arch and double open-door laminoplasty of C2, C3, and C4	Subtotal
Badri et al. [15]	41/F	Unilateral C5/6	+	Intradural	–	+	C4–C6 laminectomy	Total
Present case	57/M	Unilateral C6/7	+	Intradural	–	–	C6–C7 hemi-laminectomy	Total

The patient was discharged with advice to follow up in OPD after a month. Post-operative MRI showed complete excision of the tumor and an expanded cervical cord along with a pseudo-meningocoele formation (Fig. 3). The patient had a gradual improvement in power of the elbow extension and at 12 months improved to 4+/5 (MRC scale) power with a 90% handgrip and was able to write again. Sensory loss remained static. There were no new deficits or development of kyphosis.

3. Discussion

Ganglioneuromas are essentially tumors of the neuroectodermal cells derived from the neural crest and are classified as childhood embryonal tumors. These neuroblastic tumors form a separate class, which comprises of Neuroblastoma; ganglioneuroblastoma, intermixed; ganglioneuroma and ganglioneuroblastoma, nodular tumors. These are essentially solid tumors extracranial in location and are primarily found in children. They vary in their relative population of Schwann cells and neuroblasts and thus can form a spectrum from the benign ganglioneuroma to the intermediate ganglioneuroblastoma to the malignant neuroblastoma, according to International Neuroblastoma Pathology Classification [10–15].

Ganglioneuromas form the most benign end of the disease and hence bony preservation and maximizing the quality of life remains the primary objective along with complete excision leading to cure. We conducted a thorough search using the PubMed and Medline database using the keywords “Ganglioneuroma” and “Cervical spine” and prepared a detailed account of all such cases reported and their characteristics along with operative approaches (Table 1). Certain conclusions that can be drawn from these searches are: the most common are of origin is the upper cervical spine (above C3), which is seen in 75% of all cases reported. Recurrences have been reported but are mostly confined to familial cases and those with von Recklinghausen's disease (NF). Its association with von Recklinghausen's disease is seen in 20% of cases [1–3].

In the cervical spine a dumbbell shape (93.75%) and intradural extension are common (62.5%), a fact, which was further, corroborated again in our case. This is against the initially reported literature of rare dumbbell shaped extension and few intradural extension cases [2]. Though being childhood in origin most of these cases present in adulthood (81.25%) after the age of 18 years. This is due to their slow growth and consequently later presentation [7–10].

These cases generally recover their motor power as the lesion originates from the sensory root. This is in contradiction to their ventral extension, which has often been reported and may be explained by the root exit being a natural corridor for growth. Although the prognosis of these cases is uniformly good if excised completely local recurrence, regional lymph node metastasis, and malignant transformation is a possibility and surveillance at 1 year intervals is warranted [4–7]. Location dictates precaution as a number of critical neurovascular structures are in the vicinity and must be preserved. We describe this case to highlight the hemi-laminectomy approach (Fig. 4) in these cases and the importance of opening dura to look for the residual tumor that might be missed otherwise.

4. Conclusion

Often benign tumors with a good prognosis are where we should be most vigilant. If performed safely it can lead to the cure and good quality of life, otherwise, the deficit will have to be borne for a long period of time. A ganglioneuroma of the cervical spine presents such a possibility and thus knowledge of this rare benign disease is paramount. With only 15 such cases reported the pattern of growth and intradural extension are been increasingly seen, thus changing the knowledge database of this disease.

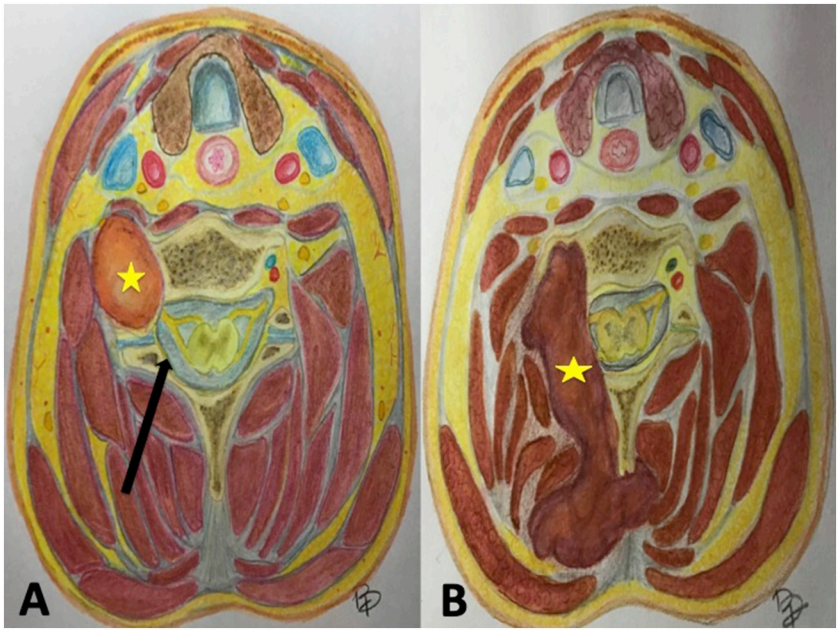

Fig. 4. (A): Schematic representation of the lesion (yellow asterisk) and the hemi-laminectomy approach taken for this case. (B): Schematic representation of the postoperative cavity and pseudomeningocoele formation (yellow asterisk) in this case. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

Disclosure of funding/source of support including

1) names of the granting organizations
2) applicable grant or contract numbers
3) names of each author who received specific funding
4) specific material support given: Nil.

Declaration of Competing Interest

Nil.

Acknowledgement

Nil.

References

[1] A.A. Bhand, Ganglioneuroma of the cervical spine, J. Coll. Physicians Surg. Pak. 15 (2005) 114–116.
[2] K. Kyoshima, K. Sakai, M. Kanaji, S. Oikawa, S. Kobayashi, A. Sato, et al., Symmetric dumbbell ganglioneuromas of bilateral C2 and C3 roots with intradural extension associated with von Recklinghausen's dase: case report, Surg. Neurol. 61 (5) (2004) 468–473.
[3] R.H. Shepherd, D. Sutton, Dumb-bell ganglioneuromata of the spine with a report of four cases, Br. J. Surg. 45 (1958) 305–317.
[4] J.E. Sinclair, Y.H. Yang, Ganglioneuromata of the spine associated with Von Recklinghausen's disease, J. Neurosurg. 18 (1961) 115–119.
[5] R.R. Strang, H. Nordenstam, Dumb-bell ganglioneuroma of the cervical spine, Acta Neurol. Scand. 38 (1962) 60–66.
[6] G. Maggi, P. Dorato, V. Trischitta, A. Varone, F. Civetta, Cervical dumbbell ganglioneuroma in an eighteen month old child. A case report, J. Neurosurg. Sci. 39 (1995) 257–260.
[7] D.V. Radulovi, D. Branislav, M.K. Skender-Gazibara, N.M. Igor, Cervical dumbbell ganglioneuroma producing spinal cord compression, Neurol. India 53 (2005) 370–371.
[8] R. Tei, T. Morimoto, K. Miyamoto, S. Aketa, T. Shimokawara, Y. Shin, et al., Intradural extramedullary ganglioneuroma associated with multiple hamartoma syndrome, Neurol. Med. Chir. (Tokyo) 47 (2007) 513–515.
[9] L.F. Ugarriza, J.M. Cabezudo, J.M. Ramirez, L.M. Lorenzana, L.F. Porras, Bilateral and symmetric C1–C2 dumbbell ganglioneuromas producing severe spinal cord compression, Surg. Neurol. 55 (2001) 228–231.
[10] N. Miyakoshi, M. Hongo, Y. Kasukawa, A. Misawa, Y. Shimada, Bilateral and symmetric C1–C2 dumbbell ganglioneuromas associated with neurofibromatosis type 1 causing severe spinal cord compression, Spine J. 10 (2010) e11–e15.
[11] J. Zhang, R. Shrestha, J. Li, S. Jiang, Giant cervicothoracic ganglioneuroma, Neurol. India 59 (3) (2011) 465.
[12] D.W. Son, G.S. Song, Y.H. Kim, S.W. Lee, Ventrally located cervical dumbbell ganglioneuroma producing spinal cord compression, Kor. J. Spine 10 (4) (2014) 246.
[13] K. Miyamoto, K. Fushimi, Y. Hirose, K. Shimizu, A. Hioki, Y. Kito, Cervical symmetric dumbbell ganglioneuromas causing severe paresis, Asian Spine J. 8 (1) (2014) 74.
[14] T. Matsumoto, S. Imagama, N. Ishiguro, K. Ando, Z. Ito, H. Matsui, et al., Cervical myelopathy caused by bilateral C1–2 dumbbell ganglioneuromas and C2–3 and C3–4 neurofibromas associated with neurofibromatosis type 1, J. Orthop. Sci. [Internet] 19 (4) (2012) 676–681, https://doi.org/10.1007/s00776-012-0315-8.
[15] M. Badri, G. Gader, K. Bahri, I. Zammel, Cervical ganglioneuroma: clinical and radiological features of a rare tumour, BMJ Case Rep. (2018), https://doi.org/10.1136/bcr-2017-223412 (bcr-2017-223412).