10.5005/jp-journals-10039-1188

CASE REPORT

Bilateral Symmetric Dumbbell C1-C2 Ganglioneuroma in Neurofibromatosis Type 1 Patient Causing Spastic Quadriparesis

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ABSTRACT

Introduction: Ganglioneuroma is a very rare tumor and rarely found bilaterally in association with neurofibromatosis patient. Very few cases of dumbbell ganglioneuroma in the cervical region have been reported in the literature.

Materials and methods: A 36-year young male presented to us with a complaint of progressive spastic quadriparesis for 4 years. We did MRI of the cervical spine with contrast and found bilateral symmetrical dumbbell mass at the C1-C2 level severely compressing the spinal cord. Rest of the spine and brain imaging showed normal findings.

Result: We operated the patient. Bilateral decompression of the cervical spine at the corresponding level was achieved by complete removal of tumor. Patient's motor power of all limbs gradually improved. Histopathology was done which revealed ganglioneuroma of both the resected tumors.

Conclusion: Bilateral symmetric dumbbell ganglioneuroma is extremely rare in C1-C2 region and can be associated with neurofibromatosis Type 1.

Keywords: Dumbbell, Ganglioneuroma, Neurofibromatosis Type 1, Spastic quadriparesis

How to cite this article: Chaurasia B, Chowdhury D, Ansari A, Khan RA, Chaurasia R, Khan AH, Chaurasiya RK, Barua KK, Chaurasia R, Lopa NA, Chaurasia SK, Biswas P, Islam MM. Bilateral Symmetric Dumbbell C1-C2 Ganglioneuroma in Neurofibromatosis Type 1 Patient Causing Spastic Quadriparesis J Spinal Surg 2018;5(3):138-143.

Source of support: Nil

Conflict of interest: None

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INTRODUCTION

Ganglioneuromas are very rare, well circumscribed, a benign tumor which is derived from neural crest cells that normally migrate into the adrenal medulla and sympathetic ganglia.[1-6] They occur in both children and young adults generally below 20 years of age, having female preponderance.[7]

Ganglioneuromas are peripheral neuroblastic tumors like neuroblastoma and ganglioneuroblastoma which consists of ganglion cells and Schwann cells.[8,1] They are most commonly located in posterior mediastinum (41.5%), followed by retroperitoneum (37.5%) and adrenal gland (21%). Only 8% of these tumors are located in the cervical region, and most of them have a dumbbell shape due to the extradural growth of tumor into spinal canal.[3,8-12] Most of the cervical ganglioneuromas are usually single.

Multiple ganglioneuromas are rare and as far as we know only four cases of bilateral symmetric dumbbell ganglioneuroma has been described in the literature. This is our 5th case which we will describe in relation with neurofibromatosis Type 1.[2,8,9] There are 30 cases of cervical ganglioneuromas in association with neurofibromatosis Type 1 have been described in the literature of which only four cases were located in the C1-C2 region.[1,9] The purpose of this report is to present an additional case of bilateral symmetric dumbbell ganglioneuroma presenting as spinal cord compression in association with neurofibromatosis Type 1. Here we will briefly describe the clinically relevant features, management, approach to the surgery and pathological findings.

CASE REPORT

A 36-year-old male with neurofibromatosis Type 1 presented to us with the complain of progressive weakness of all four limbs. He also had tingling and numbness more marked in both the upper the limb and weakness was more pronounced in upper limb than lower limb. For 15 days he became unable to walk freely and sometimes

needed support to walk. On general examination of the patient, we found multiple café au lait spot all over the body and multiple subcutaneous nodules all around the body and limbs. Other features of NF1 like iris lisch nodule, axillary freckling, optic nerve glioma, sphenoid dysplasia and history of NF1 in the family were absent. Neurological examinations of the patient revealed he had spastic gait, all modalities of sensation reduced below C-3 dermatome, bilateral planter extensors, all deep tendon jerks in both upper and lower limbs exaggerated and presence of ankle clonus.

All the muscle power of both upper limbs and lower limbs on the left side were three, and right side were four.

Magnetic resonance imaging of cervical spine with contrast showed two bilateral symmetrical extradural dumbbell masses at a c1-c2 level which were extending outside through neural foramina. In T1WI it was isointense, slight hyperintense on T2WI, and there was slight uptake of contrast by the tumors. Spinal cord at that level was severely compressed bilaterally (Fig. 1). Rest of the screening of spine showed normal findings. MRI of the brain revealed normal findings. We did not do CT scan and vertebral angiography due to financial problems of the patient.

Patient was operated through posterior approach in the prone position. We did a slight big incision extending from slightly below the external occipital protuberance up to slightly below the mid-cervical region. The posterior arch of both c1 and c2 were intact. We exposed it by separating muscular attachment of it. The tumor was seen as avascular, solid and smooth and was covered with the capsule. Dural margins was also seen and the capsule was slowly and carefully separated from tumor and was removed en-block. First right then left side of the tumor was removed without doing any bone works. Tumors were not attached to dura so it did not pose any difficulty in removing it. The microscope was finally used to see the completeness of the tumor. Hemostasis was secured and wound closed in layers with drain kept in situ.

Postoperative image showed complete removal of both tumors (Fig. 2). Histopathology of the tumor was done (Figs 3 and 4). Both the resected specimens (Fig. 5) showed benign neoplasm composed of cluster of large cells with interlacing bundle of spindle cells suggesting the presence of ganglion cells. The ganglion cells were large cells with one or two nuclei having abundant cytoplasm schwann cells. No mitotic activity was found nor was cellular pleomorphism seen.

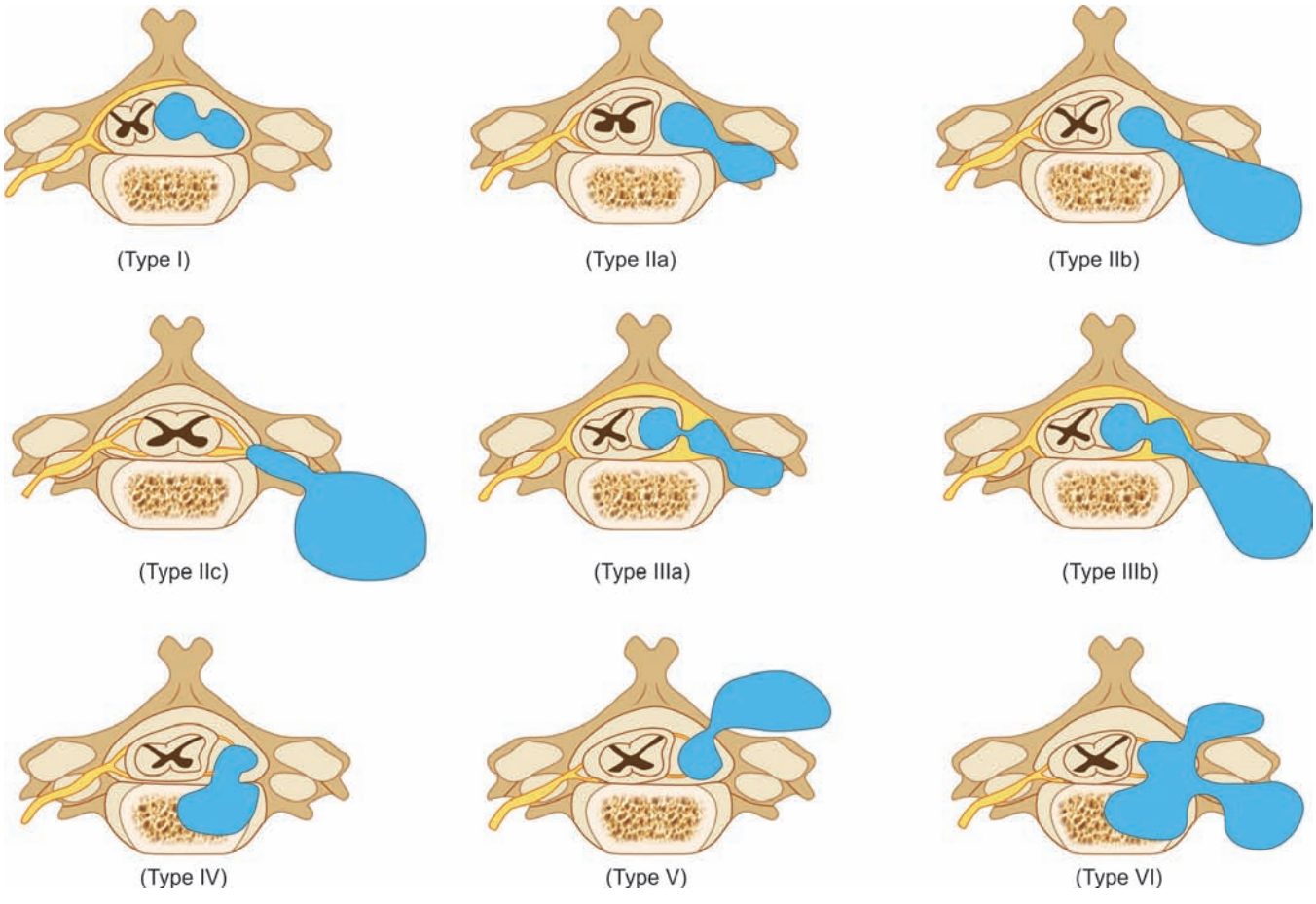

Fig. 1: Toyama classification[9]

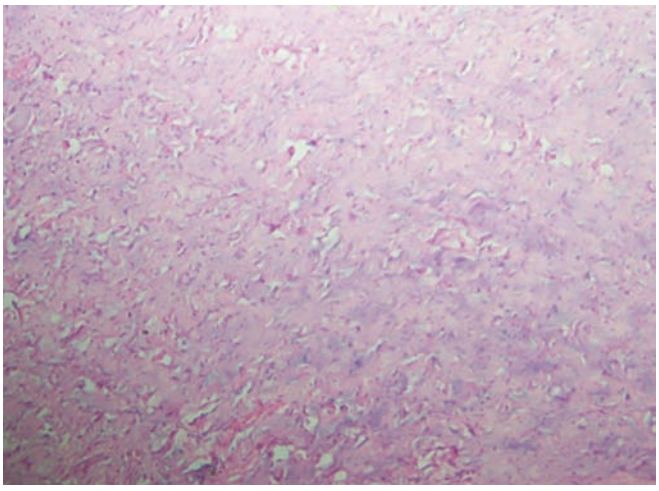

Fig. 2: Low power view showing admixtures of schwann cells and ganglion cells with myxoid changes seen

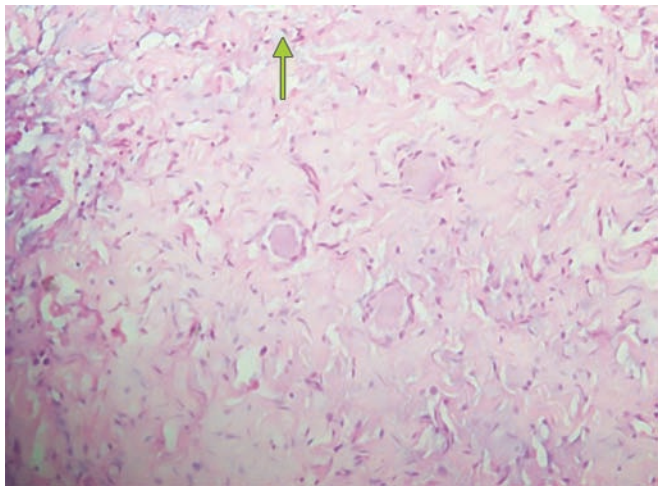

Fig. 3: Shows 3 ganglion cells seen in high power view

The postoperative course was uneventful. The patient gradually recovered in neurological functions (Fig. 6). The gait of the patient recovered with no requirements of any assistance for walking. Motor functions of all the limbs were gradually improved. Postoperative MRI after 10 days was done to see the completeness of tumor. It showed the total removal of tumors bilaterally. We planned to follow up after 6 months with MRI.

DISCUSSION

Ganglioneuroma is a rare tumor of the cervical region which originates from neural crest tissue of the sympathetic nervous system.[13] Ganglioneuroma can be found as multiple and sometimes is associated with neurofibromatosis type 1, but the genetic association is still unclear.[3] The dumbbell growth pattern of this tumor is due to extension into the spinal canal through the

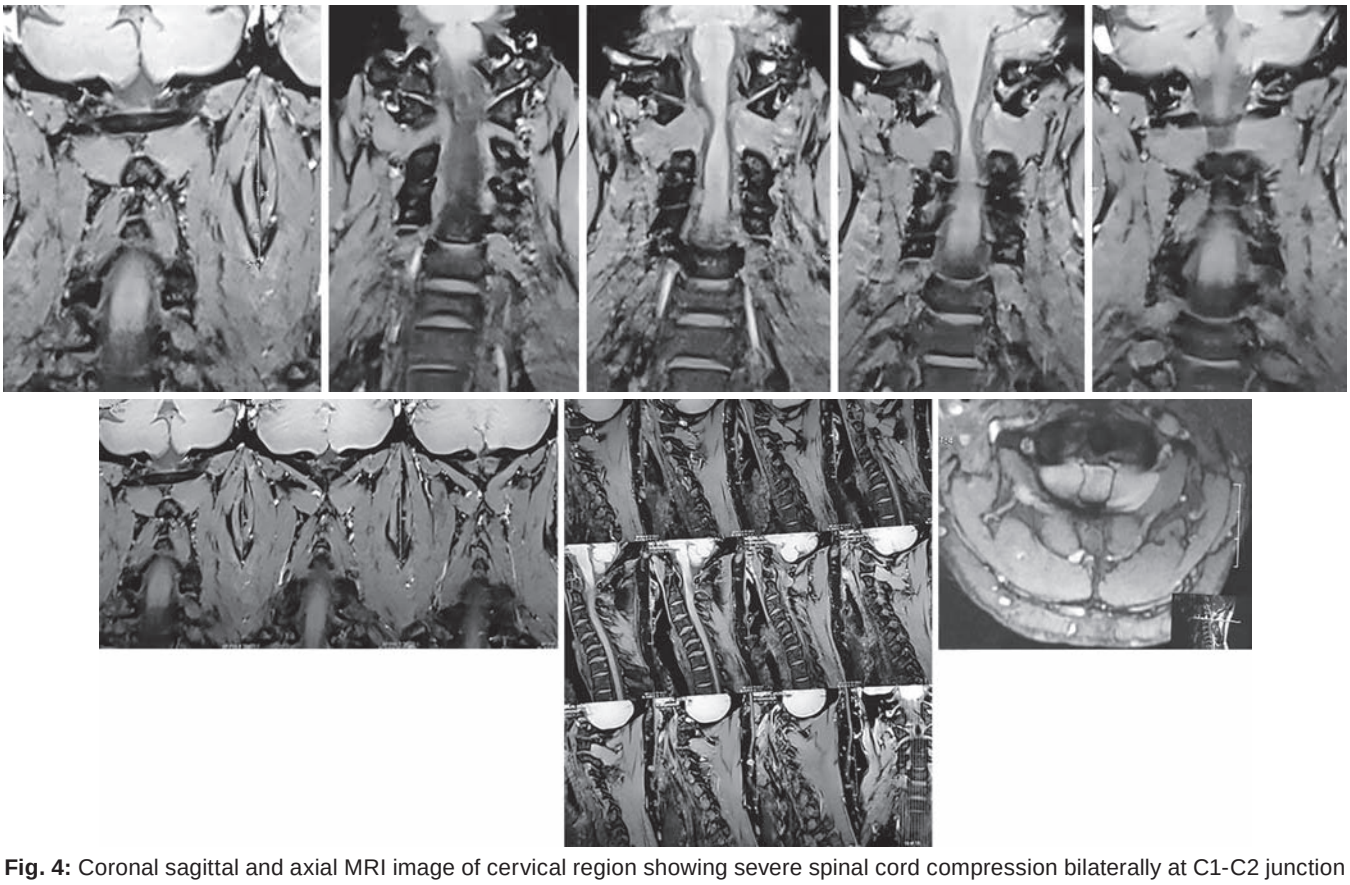

Fig. 4: Coronal sagittal and axial MRI image of cervical region showing severe spinal cord compression bilaterally at C1-C2 junction (preoperative image)

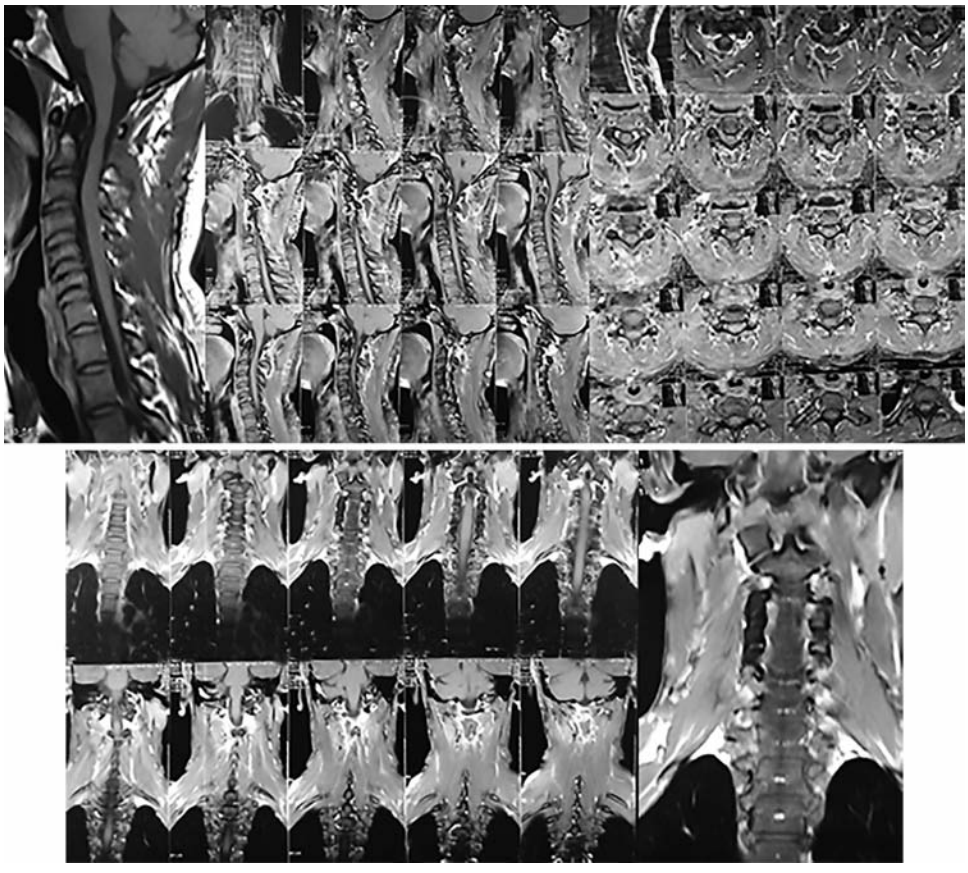

Fig. 5: Sagittal, axial and coronal MRI image with contrast showing complete removal of tumor bilaterally (postoperative image)

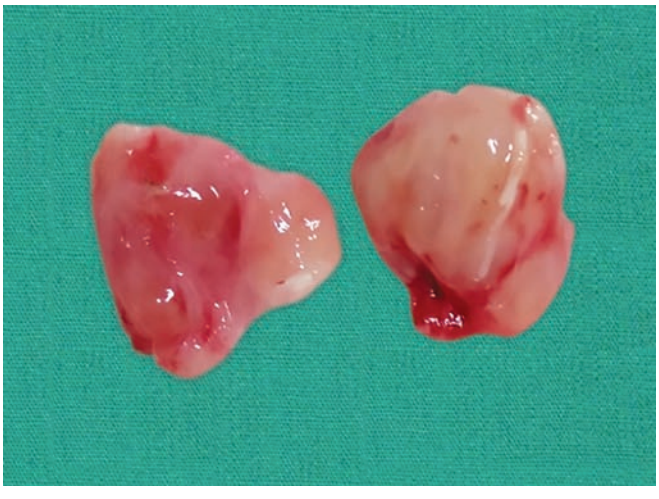

Fig. 6: Showing bilateral ganglioneuroma removed completely

intervertebral foramen. The pattern of dumbbell shape depends on whether the tumor originates from within or outside the spinal canal or within intervertebral foramen. Extradural extension of this tumor is more common than an intradural.[3] Classification system for dumbbell cervical tumor as proposed by Toyama has been given below in Fig. 7.[9] According to this classification, our tumors belong to Type II. Table 1 shows summary of the previously reported case of cervical gangliomeuromas.[2]

Summary of the previously reported case of Cervical Ganglioneuroma [2]

Bilateral symmetric dumbbell tumor of ganglioneuroma in cervical region is very uncommon, and its association with neurofibromatosis type 1 is still more uncommon and rare.[15] Till now only four cases of bilateral dumbbell cervical ganglioneuroma have been described. Ours case is the 5 th case report in a row (Table 2). But only three

Fig. 7: Image after 7th postoperative day (with consent) patient can stand and walk without support

cases has been described of bilateral cervical dumbbell ganglioneuroma associated with neurofibromatosis Type 1, and our are the 4th case report adding to them (Table 3). Ugarriza et al. reported the first case.[12] which was about bilateral and symmetric C1-C2 dumbbell ganglioneuroma producing severe spinal cord compression in 53 years old man.[16] Kyoshima et al.[3] described the second case in 35 years old man about symmetric dumbbell ganglioneuroma of bilateral C2 and C3 root with an intradural extension associated with von Recklinghausen disease.[3] Miyakoshi et al.[2] in his 3rd case described bilateral and symmetric C1-C2 dumbbell ganglioneuroma associated with neurofibromatosis Type 1 causing severe spinal cord compression.[2] Son DW et al.[8] reported 4th case in 13 years boy which was about ventrally located cervical dumbbell ganglioneuroma producing spinal cord compression.[8] The fifth case was reported by Hioki et al.[6] about cervical symmetric dumbbell ganglioneuroma causing severe paresis in 72 years old man.[6] The present case is the 6th case in a row (Table 2).

The age range of patients were 13-36 (Table 3). The mean range was 24.75 years. The present case was the eldest case among them. All patients were male.Among five cases (Table 2) four of them have an association with neurofibromatosis Type 1.

Surgical decompression is the treatment of choice to relieve the spinal cord compression. If possible complete excision should be done. In case it can't be remove

Table 1: Summary of the previously reported case of cervical gangliomeuromas[2]

Authors	*Age/Sex*	*Site of origin*	*Dumbbell shape*	*Intraspinal extension*	*Multiplicity*	*NF1*
Shephard and Sutton	35 m	Unilateral C2-C7	p	Intradural	Present	Present
Sinclair and Yang	44 f	Unilateral C2-C5	p	Intradural	Present	Present
Stranga and Nordenstam	63 f	Unilateral C2-4	p	Intradural	-	-
Maggi et al.	1.5 f	Unilateral C2-6	p	Extradural	-	-
Ugarriza et al.	53 m	Bilateral C2	p	Extradural	Present	-
Kyoshima et al.	35 m	Bilateral C2 and C3	Present	Intradural	Present	Present
Bhand	22 f	Unilateral C3	P	Extradural	-	-
Radulovi et al.	39 m	Unilateral C5	p	Extradural	-	-
Tei et al.	51 f	Unilateral C1	-	Intradural	-	-
Miyakoshi et al.[2]	15 m	Bilateral C2, unilateral C4	p	Extradural	Present	Present
Bacci et al.[7]	32 f	Bilateral C1-C7	-	Intradural	Present	Present
Radulovi et al.[12]	39 m	Unilateral C4-C5	Present	Intradural	-	-
Ma et al.[5]	4 f	Unilateral C4-C7	-	Extradural	Present	-
Son DW et al.[8]	13 m	Bilateral C3	Present	Intradural	Present	Present
Hioki et al.[6]	72 m	Bilateral C2	Present	Extradural	Present	-
Kharosekar et al.[10]	32 m	Unilateral C1-C2	Present	Intradural	-	Present
Dutta[4]	18 months m	unilateral	-	Extradural	-	-
Lonescu et al.[13]	53 f	Unilateral C5-C6	-	Intradural	-	-
Badri et al.[14]	41 f	Unilateral C5-C6	Present	Intradural	-	-
Helal et al.[15]	10 f	unilateral	-	Extradural	Present	-
Chaurasia et al. (present case)	36 m	Bilateral C1-C2	Present	Extradural	Present	Present

Table 2: Summary of previously published case report with bilateral symmetric ganglioneuroma producing spinal cord compression[2]

Authors /Year	*Age/ Sex*	*Site of origin*	*Dumbbell shape*	*Intraspinal extension*	*Multiplicity*	*Nf1*
Ugarriza et al.[11] 2001	53 m	Bilateral symmetric C2	Present	Extradural	Present	Absent
Kyoshima et al. 2004	35 m	Bilateral C1 and C2 symmetric	Present	Intradural	Present	Present
Miyakoshi et al. 2010	15 m	Bilateral symmetric C2	Present	Extradural	Present	Present
Son DW et al. 2013	13 m	Bilateral symmetric C3 ,ventrally located	Present	Intradural	Present	Present
Hioki et al. 2014	72 m	Bilateral symmetric C2	Present	Extradural	Present	Absent
Chaurasia et al. (present case) 2018	36 m	Bilateral symmetric C2	Present	Extradural	Present	Present

Table 3: Summary of previously published case report with bilateral symmetric ganglioneuroma producing spinal cord compression in association with neurofibromatosis Type1[2]

Authors /Year	*Age/Sex*	*Site of origin*	*Dumbbell shape*	*Intraspinal extension*	*Multiplicity*	*Nf1*
Kyoshima et al. 2004	35 m	Bilateral C1 and C2 symmetric	Present	Intradural	Present	Present
Miyakoshi et al. 2010	15 m	Bilateral symmetric C2	Present	Extraduraal	Present	Present
Son DW et al. 2013	13 m	Bilateral symmetric C3, ventrally located	Present	Intradural	Present	Present
Chaurasia et al. 2018	36 m	Bilateral symmetric C2	Present	Extradural	Present	Present

totally, partial decompression should be done.[2] In our case, spinal cord was severely compressed by tumor bilaterally which was removed totally and the cord was decompressed. The extra spinal canal and for aminal portions were also removed. Although both the tumors were completely removed follow up of the patient may be needed for recurrence of tumor at the same site or new tumor growth may be found at other sites. We didn't advise any post-operative adjuvant therapy.[17]

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