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Research Article

CAN BILATERAL SYMMETRIC DUMBBELL SHAPED C2 NERVE SHEATH TUMORS BE CONSIDERED AS CRITERIA OF NF-1?

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ABSTRACT

Ganglioneuroma is rare benign tumors of autonomic nerve fibers arising from neural crest cells. We report a case of 40-year-old male, with features of NF -I, presented us with complaints of gradually progressive quadriparesis for one year and bladder dysfunction. His imaging (MRI cervical spine) was suggestive of bilateral symmetric dumbbell shaped tumor at C2, and multiple similar small lesions at other cervical level. Tumor resection was done and histopathology report was consistent with ganglioneuroma. Out of the 14 cases reported in literature till date, association with NF-1 and multiplicity of lesion was seen. In the post MRI advent era, these features can be considered as one of the minor criteria for NF1.

INTRODUCTION

Ganglioneuroma is rare, differentiated, benign, and slow growing tumor arising from the neural crest tissue of the sympathetic nervous system, and is histologically composed of mature Schwann cells and ganglion cells with fibrous stroma. Their most common location is retroperitoneum and posterior mediastinum. The incidence in cervical region and being bilaterally symmetric is further extremely rare. [1] Only 4 cases of bilateral symmetric dumbbell shaped tumor at C2 causing severe cord compression are reported till date. [2] We report first case report from India with bilateral symmetric C2 tumor.

Case Report

A 40 years old male, with no known comorbid illness was admitted with history of gradually progressive, ascending weakness in lower limb more than upper limb associated with numbness in all limbs for last one year. He also had diffuse swelling in right thigh for 2 years. He had initially difficulty in walking and now is bedridden for last few days. He also had urinary retention for last one month for which he was catheterized at other hospital and referred to our center for further management. No family history of neurofibromatosis - 1.

On examination, the he was conscious, alert and oriented. Cranial nerve examination revealed No abnormality. He had spastic quadriparesis with Nurick grade 5 (completely bedridden). Neurological examination revealed bilateral wasting of muscles with increased tone. Power as per MRC grading was 2/5 in Right Upper limb and 0/5 in right lower limb; left upper limb 3/5 and lower limb 2/5 with bilateral grip weakness. He had graded sensory loss below C4 dermatome. Deep tendon reflexes were exaggerated, planters showed positive Babinski sign. Physical examination revealed multiple small cutaneous neurofibromas over back and abdomen and large diffuse swelling at right mid-thigh consistent with large plexiform neurofibroma. He had no other NF I feature. Magnetic resonance imaging (MRI) revealed dumbbell shaped bilateral symmetric T2 hyperintense, T1 isointense with minimal post contrast enhancing lesion at C1- C2 with some extraforaminal extension, and causing severe cord compression ventrally. Similar small lesion bilaterally at C6-7, C7-T1, and T1-2 levels without cord compression were also noted. [Figure 1]. He underwent C1 posterior arch and C2 superior part of lamina excision with complete bilateral tumor excision. Tumors were bilateral symmetric, large, well encapsulated, extradural, firm with severe cord compression. Bilateral C2 posterior roots were sacrificed and tumor resected.

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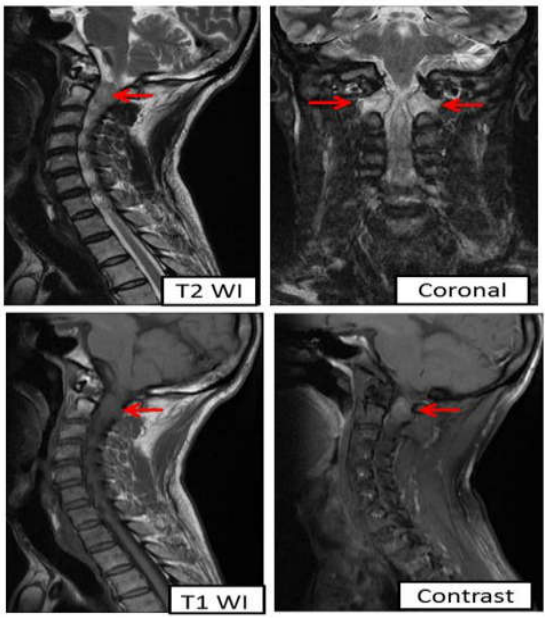

Figure 1 Magnetic Resonance Imaging (MRI) showing C1-C2 nerve sheath tumor.

Other lesions left untouched. As the bilateral C2 lesion alone was causing severe cord compression, we have operated on the symptomatic lesion only and total resection of the bilateral lesion was achieved. Histopathological examination (HPE) of tumor was suggestive of ganglioneuroma. [Figure 2].

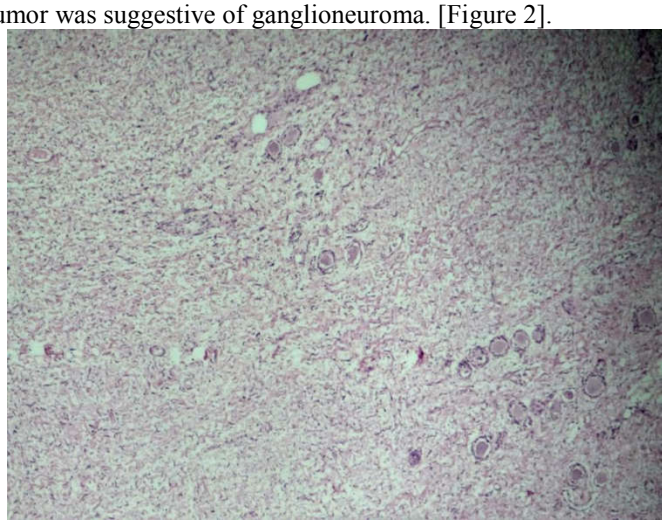

Figure 2 Histopathological examination (HPE) suggestive of ganglioneuroma.

Genetic evaluation was not done due to financial constraints. We have followed up the patient till 6 months and there was no further neurological deterioration.

DISCUSSION

NF, I have been associated with nerve sheath tumors in many case reports. Total 14 cases of cervical nerve sheath tumors were reported till date. [Table 1] [2-15] Only four cases are reported with bilateral symmetric lesion at cervical region. [2,7,8,12] Our case report is addition to this list.

Bilateral symmetric lesion at cervical region was initially reported by Ugarriza *et al.* in 53-year-old male from Spain in 2001. [7] Later three cases were added from Japan in 15, 35 and 72 year males. [2,8,12] Multiplicity of lesions and association with NF-1 was noted by Kyoshima *et al.* and Miyakoshi N, *et al.* In the presentcase, we had also seen multiplicity of nerve sheath tumor and features of NF-1.

NF-1 is autosomal dominant disease and with prevalence been estimated to be about 1/3500 in the USA and the United Kingdom. [16] Since pre-MRI era NIH criteria for NF1 (National Institute of Health, Consensus Development Conference held in Bethesda, Maryland, July 1987) are used for diagnosis. [Table 2] [17] There is a need to update these criteria and include few lately identified features in post MRI era, to be included with NIH criteria of NF-1for the diagnosis.

Including the present case, seven of the total 15 cervical nerve sheath tumors with HPE of ganglioneuromas, the NIH criteria for NF1 were also noted and seven cases also had multiplicity of the lesion. So, this clinical criterion can be also considered as one of the minor criteria of NF-1.

CONCLUSION

Bilateral symmetric cervical nerve sheath tumors at craniovertebral junction are extremely rare. Many such cases have features of NF-1 and multiple nerve sheath tumors. After the advent of MRI, there are no modifications done to the age-old criterion of NF-1. The bilateral symmetric neve sheath tumor with multiplicity of tumors can be considered as one of the minor criteria for NF1.

Table 1 List of cervical ganglioneuroma published till date.

S. No.	Authors	Year reported	From	Age/sex	Site of Origin	Multiplicity	NF-1
1	Shepherd and Sutton *et al.*	1958	England	35/M	Unilateral C2-C7	+	+
2	Sinclair and Yang *et al.*	1961	USA	44/F	Unilateral C2-C5	+	+
3	Strang and Nordenstam *et al.*	1962	Sweden	63/F	Unilateral C2-C4	-	-
4	Maggi *et al.*	1995	Italy	1.5/F	Unilateral C2-C6	-	-
5	Ugarriza *et al.*	2001	Spain	53/M	Bilateral C2	+	-
6	Kyoshima *et al.*	2004	Japan	35 /M	Bilateral C2 and C3	+	+
7	Radulovi *et al.*	2005	Serbia	39/M	Unilateral C5	-	-
8	Bhand *et al.*	2005	Pakistan	22/F	Unilateral C3	-	-
9	Tei *et al.*	2007	Japan	51/F	Unilateral C1	-	-
10	Miyakoshi N,*et al.*	2010	Japan	15/M	Bilateral C2 unilateral C4	+	+
11	Ji Zhang *et al.*	2011	China	24/F	Unilateral C6-7	-	-
12	Son DW *et al.*	2013	Korea	13/M	Unilateral C2-C3	+	+
13	Hioki A, *et al.*	2014	Japan	72/M	Bilateral C1/C2	-	-
14	Kharosekar *et al.*	2015	India	32/M	Unilateral C1	-	+
15	Case 1	2016	India	40/M	Bilateral C2	+	+

Table 2 NIH (National Institute of Health, Consensus Development Conference held in Bethesda, Maryland, July 1987) criteria for NF1.

Clinical diagnosis based on presence of two of the following:
1. Six or more café-au-lait macules over 5 mm in diameter in prepubertal individuals and over 15mm in greatest diameter in postpubertal individuals.
2. Two or more neurofibromas of any type or one plexiform neurofibroma.
3. Freckling in the axillary or inguinal regions.
4. Two or more Lisch nodules (iris hamartomas).
5. Optic glioma.
6. A distinctive osseous lesion such as sphenoid dysplasia or thinning of long bone cortex, with or without pseudarthrosis.
7. First-degree relative (parent, sibling, or offspring) with NF-1 by the above criteria.

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