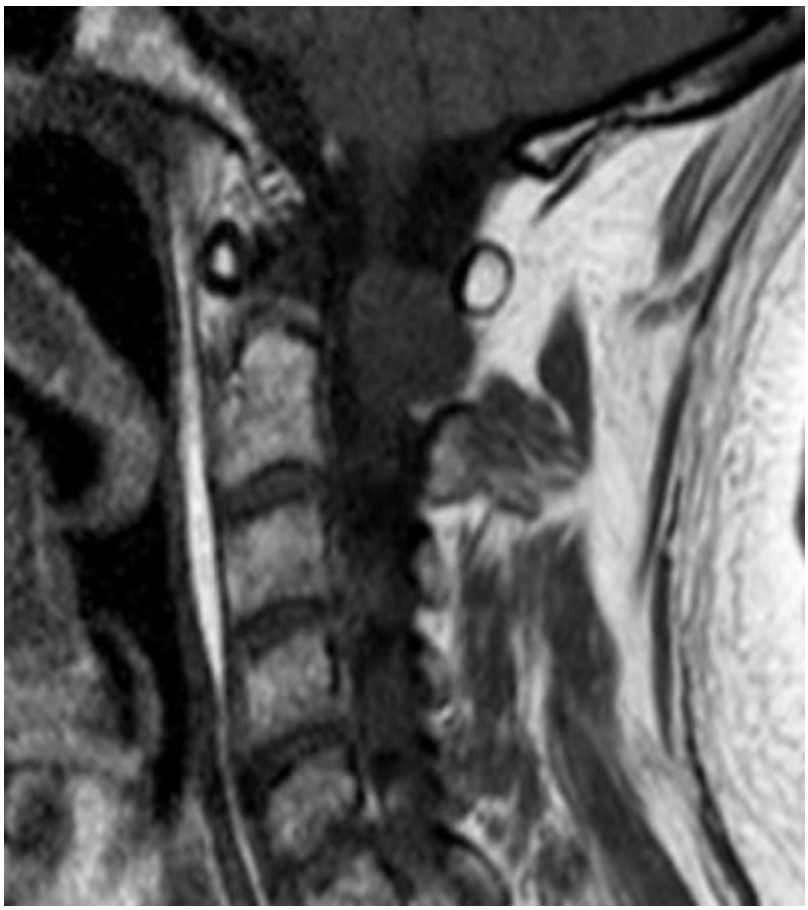
Cervical bilateral spinal neurofibroma

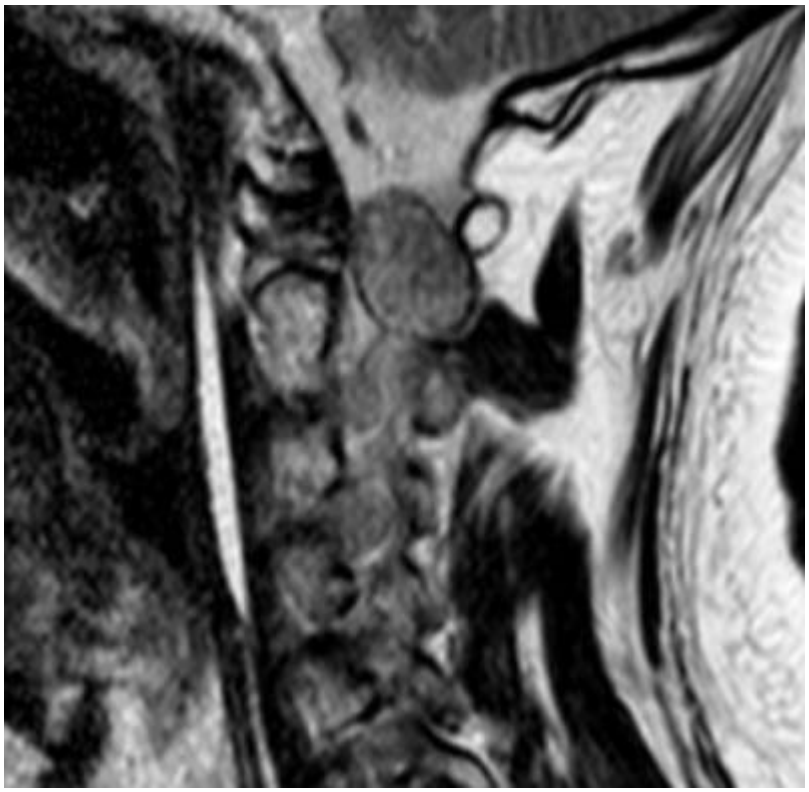
Although rare, upper cervical mirror-image spinal neurofibromas have been reported in the medical literature, and their surgical management has been addressed in several reports; however, little has been mentioned or is known regarding upper cervical or craniocervical stability following resection of these tumors.

Bartolomei and Crockard described four cases of large mirror-image C-2 neurofibromas resected in two stages via the posterolateral approach. One patient presented with acute neurological deterioration after a biopsy sample had been obtained, whereas the other three presented with gradual onset of lower-extremity weakness over several months. The time interval between the first and second decompressive surgery ranged from 10 days to 12 weeks. There were no surgery-related complications, and all patients recovered motor function in their extremities. During a follow-up period of 16 to 36 months, there was no clinical or radiological evidence of upper cervical spine instability. Although the series is too small to draw any definitive conclusions, in the authors' experience the posterolateral approach provides a direct route for the successful surgical treatment of bilateral craniocervical nerve root tumors without destabilizing the upper cervical segments [1)].

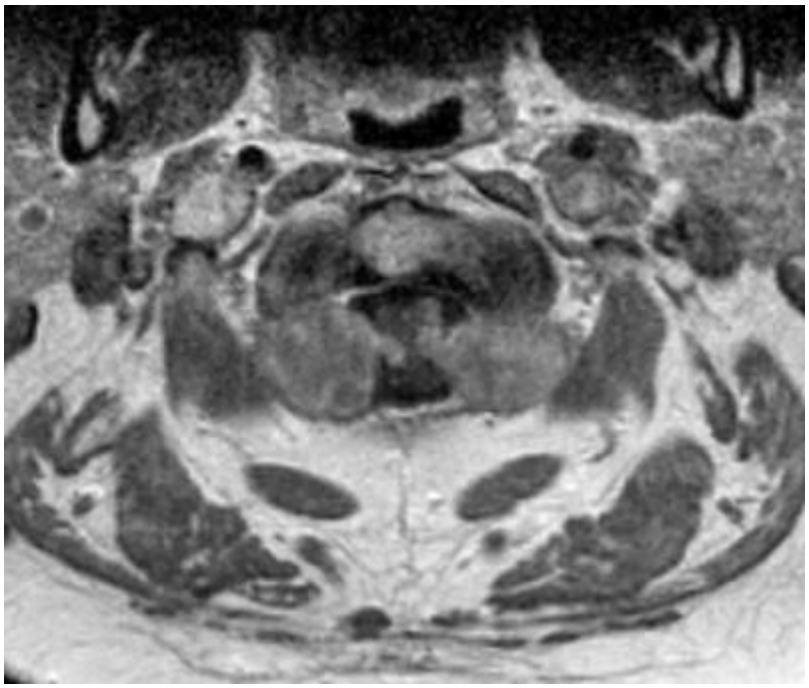
MRI



T1: hypointense



T2: hyperintense



Case reports

2017

Finet and Raftopoulos report the case of a patient with bilateral C2 spinal neurofibromas presenting with a myelopathy of the upper cervical spine with no spinal cord compression on standard magnetic

resonance imaging. The spinal cord compression occurred between the cervical neurofibromas only during head rotation.

A patient with bilateral neurofibromas of the C2 nerve roots showed a progressive neurological deterioration with an intramedullary MRI hypersignal without visible compression. Only an additional MRI carried-out with the head in rotation demonstrated the tumoral dynamic compression. A review of the literature showed that only two similar cases had previously been reported. The largest C2 tumor was completely removed uneventfully.

Dynamic compression of the spinal cord in patients with bilateral C2 nerve root tumors must be routinely investigated even if the standard MRI shows no compression [2)].

Bigder et al. present a 21-year-old male diagnosed with NF1 in infancy and followed for multiple mirror-image neurofibromas involving the entire spine. He was asymptomatic until age 14 when he developed neck pain and progressive tetraplegia with magnetic resonance imaging showing severe cord compression secondary to bilateral C2 neurofibromas. Emergent cervical decompression was performed at C1-C3 along with debulking of bilateral neurofibromas. Postoperatively he regained full strength with no signs of myelopathy several years postoperatively. This case demonstrates a dramatic neuroimaging finding and emphasizes the potential for significant neurologic deterioration in previously asymptomatic NF1 patients, highlighting the need for long-term follow-up [3)].

Pandey et al. reported an interesting case of bilateral symmetrical cervical neurofibroma with multiple spinal neurofibromas appearing as mirror image on CT, associated with non familial NF-1 as a rare presentation in a 25-year-old adult male [4)].

2014

A 37-year-old man affected by NF1 referred to the department for progressive weakness of both lower extremities and gait disturbance. Radiological imaging showed bilateral dumbbell-shaped C2 spinal neurofibromas. After its resection, at the 1-month follow-up evaluation, the patient reported headache and nausea. A CT brain scan showed a postoperative cervical pseudomeningocele and an increase in the ventricular sizes, resulting in hydrocephalus. Results A ventriculoperitoneal shunting was performed using a programmable valve opening pressure set to 120 mmH20. After surgery, the patient's neurological status markedly improved.

Hydrocephalus must be considered a possible complication of cervical spine tumor resection [5)].

2012

Ozawa et al. describe 2 patients with C-2 nerve root tumors in whom the lesions were located bilaterally in the lateral portions of the C1-2 interlaminar space and compressed the spinal cord when the atlantoaxial joint was rotated. The patients were adult men with neurofibromatosis. Each presented with clumsiness of both hands and motor weakness of the extremities accompanied by spastic gait. Magnetic resonance imaging of the cervical spine performed with the neck in the neutral position showed tumors at the bilateral lateral portion of the C1-2 interlaminar space without direct

compression of the spinal cord. The spinal cord exhibited an I-shaped deformity at the same level as the tumors in one case and a trapezoidal deformity at the same level as the tumors in the other case. Computed tomography myelography and MRI on rotation of the cervical spine revealed bilateral intracanal protrusion of the tumors compressing the spinal cord from the lateral side. The tumors were successfully excised and occipitocervical fusion was performed. The tumors were pushed out into the spinal canal from the bilateral lateral portion of the interlaminar spaces due to rotation of the atlantoaxial joint. This was caused by a combination of posteromedial displacement of the lateral mass on the rotational side of the atlas and narrowing of the lateral portion of the interlaminar space on the contralateral side due to the coupling motion of the lateral bending and extension of the atlas. The spinal cord underwent compression from both lateral sides in a one-way rotation. Without sustained spinal cord compression, intermittent long-term dynamic spinal cord compression from both lateral sides should induce a pathognomonic spinal cord deformity and the onset of paralysis. To the authors' knowledge, there have been no reports of the present conditions-that is, the bilateral protrusion of tumors from the bilateral lateral portion of the C1-2 interlaminar spaces into the spinal canal due to atlantoaxial rotation [6)].

2010

A case with bilateral and symmetric dumbbell ganglioneuromas of the cervical spine as part of multiple ganglioneuromas of the spine in a patient with neurofibromatosis type 1 (NF-1).

A 15-year-old boy with NF-1 presented with a 6-month history of progressive tetraparesis. Magnetic resonance imaging showed voluminous bilateral and symmetric dumbbell masses at the C1-C2 level severely compressing the spinal cord. The spinal cord was also indented by a dumbbell mass at the left C3-C4 level. A systemic imaging survey of the patient showed numerous asymptomatic foraminal and extraforaminal tumors at all neuroforamina of the spine.

The result was found to be surgical decompression of the spinal cord by subtotal resections of bilateral tumors at the C1-C2 level and unilateral tumor at the left C3-C4 level alleviated patient symptoms. Histopathological diagnosis was ganglioneuroma for all resected tumors.

Multiple ganglioneuromas, particularly bilateral and symmetric dumbbell tumors, are extremely rare but could be associated with NF-1 [7)].

2004

An extremely rare case in an adult with von Recklinghausen's disease who had bilateral, symmetric and multiple dumbbell ganglioneuromas with intradural extension, and also multiple bilateral ganglioneuromas at the neck. The intradural ganglioneuromas were suspected to have originated from the posterior root ganglions of the bilateral C2 and C3 nerves and to have extended ventrally to the spinal cord involving not only sensory but also motor rootlets; the ganglioneuroma of the neck was suspected to have originated from the cervical nerve itself [8)].

1)
Bartolomei JC, Crockard HA. Bilateral posterolateral approach to mirror-image C-2 neurofibromas. Report of four cases. J Neurosurg. 2001 Apr;94(2 Suppl):292-8. PubMed PMID: 11302634.
2)
Finet P, Raftopoulos C. Myelopathy secondary to a spinal cord compression induced by C2 nerve

tumors during head rotation. Neurochirurgie. 2017 May;63(2):107-108. doi: 10.1016/j.neuchi.2016.12.003. Epub 2017 May 11. PubMed PMID: 28502567.
3)
Bigder M, Szelemej P, Berrington N. Extensive Mirror-Image Neurofibromas of Entire Spine Resulting in Spastic Tetraplegia. World Neurosurg. 2017 Dec;108:985.e7-985.e8. doi: 10.1016/j.wneu.2017.08.139. Epub 2017 Sep 1. PubMed PMID: 28867317.
4)
Pandey S, Singh K, Sharma V, Khan MT, Ghosh A, Santhosh D. Bilateral Mirror Image Cervical Neurofibroma in an Adult with Neurofibromatosis Type 1. Malays J Med Sci. 2017 Mar;24(1):117-120. doi: 10.21315/mjms2017.24.1.13. Epub 2017 Feb 24. PubMed PMID: 28381935; PubMed Central PMCID: PMC5346010.
5)
Montemurro N, Cocciaro A, Meola A, Lutzemberger L, Vannozzi R. Hydrocephalus following bilateral dumbbell-shaped c2 spinal neurofibromas resection and postoperative cervical pseudomeningocele in a patient with neurofibromatosis type 1: a case report. Evid Based Spine Care J. 2014 Oct;5(2):136-8. doi: 10.1055/s-0034-1387805. PubMed PMID: 25364327; PubMed Central PMCID: PMC4212698.
6)
Ozawa H, Kusakabe T, Aizawa T, Nakamura T, Ishii Y, Itoi E. Tumors at the lateral portion of the C1-2 interlaminar space compressing the spinal cord by rotation of the atlantoaxial joint: new aspects of spinal cord compression. J Neurosurg Spine. 2012 Dec;17(6):552-5. doi: 10.3171/2012.9.SPINE12562. Epub 2012 Sep 28. PubMed PMID: 23020210.
7)
Miyakoshi N, Hongo M, Kasukawa Y, Misawa A, Shimada Y. Bilateral and symmetric C1-C2 dumbbell ganglioneuromas associated with neurofibromatosis type 1 causing severe spinal cord compression. Spine J. 2010 Apr;10(4):e11-5. doi: 10.1016/j.spinee.2010.01.023. PubMed PMID: 20362242.
8)
Kyoshima K, Sakai K, Kanaji M, Oikawa S, Kobayashi S, Sato A, Nakayama J. Symmetric dumbbell ganglioneuromas of bilateral C2 and C3 roots with intradural extension associated with von Recklinghausen's disease: case report. Surg Neurol. 2004 May;61(5):468-73; discussion 473. Review. PubMed PMID: 15120227.