C2 Segmental Neurofibromas in Patients with Neurofibromatosis Type 1: A Particularly Aggressive Phenotype

Mueez Waqar MBChB, MRes; Calvin Soh; John Ealing; Susan Huson; Gareth Evans; Konstantina Karabatsou; Joshi George
Manchester Academic Health Sciences Centre, The University of Manchester, Manchester, United Kingdom

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Introduction

- Segmental spinal neurofibromas have a reported prevalence of up to 40% in NF1 patients [1].
- Reported rates vary by institution due to differences in criteria for spinal imaging.
- Neurofibromas affecting the C2 level have been commonly described in patients with and without NF1 [2,3].
- The clinical course and imaging characteristics of neurofibromas occuring at the C2 level are unexplored.

Aim

- To present clinical and radiological outcomes of C2 neurofibromas in patients referred to a supra-regional NF1 centre.

Methods

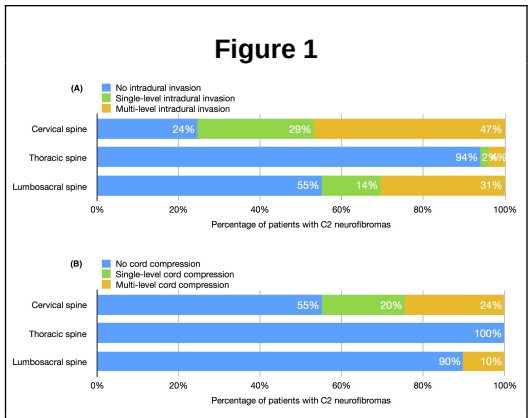
- Review of referrals to a national NF1 referral centre in the United Kingdom (2009-2016).
- Inclusion criteria: (1) diagnosis of NF1; (2) at least one C2 root neurofibroma; (3) magnetic resonance imaging of the C-spine or whole spine.
- Odds and odds ratios were used for group comparisons. Multivariate logistic regression analysis was used to identify factors associated with need for surgery.

Results

54 patients with at least one C2 neurofibroma were included in this study, representing 43% of patients with spinal neurofibromas. The median age was 32.5 (range 15-61) years. There was a slight male excess (M:F, 33:21).

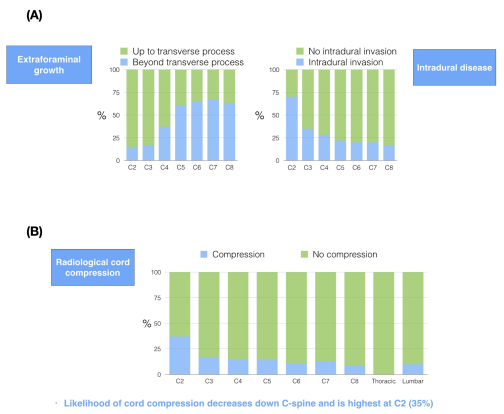
Most patients (67%) had =1 neurofibroma affecting every spine region. Only 3 (6%) had isolated neurofibromas at the C2 level.

Figure 1



Spine segmental neurofibroma patterns in patients with C2 neurofibromas. A - Intradural invasion patterns. These were similar between the cervical and lumbar spine. Intradural invasion was rare in the thoracic spine. B - Cord compression patterns. In general, radiological cord compression was rare outside of the cervical spine.

Figure 2



C2 versus other cervical level neurofibromas. There was a reciprocal relationship between intradural invasion and extra-foraminal growth down the cervical spine (Fig 2A). Compared to other cervical spine neurofibromas, C2 neurofibromas had higher rates of intraspinal extension (OR=6.20, 95% CI 3.85-9.97; p<0.001), intradural invasion (Fig 2B; OR=3.20, 95% CI 2.08-4.92; p<0.001) and cord compression (Fig 2B; OR=2.26, 95% CI 1.35-3.79; p=0.002). C2 neurofibromas had lower rates of extraforaminal growth beyond the transverse process (Figure 2B; OR=0.09, 95% CI 0.05-0.16; p<0.001).

Factors associated with surgery included myelopathy (p = 0.03) but not radiological cord compression (p > 0.99).

Conclusions

C2 neurofibromas are particularly aggressive tumours due to preferential intraspinal growth.

Learning Objectives

By the conclusion of this session, participants should be able to:

- Appreciate the burden of spinal disease in a series of patients with NF1
- Appreciate neurofibroma growth patterns in the spine, with particular emphasis on the cervical spine
- Appreciate C2 neurofibromas characteristics that make them particularly aggressive compared to other segmental neurofibromas
- Appreciate that radiological cord compression is not always an indication for surgery

References

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