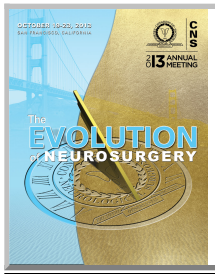

Spinal Cord Compression from Spinal Root Neurofibromas in Neurofibromatosis type 1 (NF1)

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Introduction

Neurofibromas are characteristic of NF1, growing along cutaneous and subcutaneous nerves. Although mainly asymptomatic, spinal neurofibromas are present in at least 40% of NF1 patients. Only few small series are available, with limited information, about the management of spinal cord compression from spinal neurofibromas. We here review our experience in the management of these tumors, and their particular characteristics.

Methods

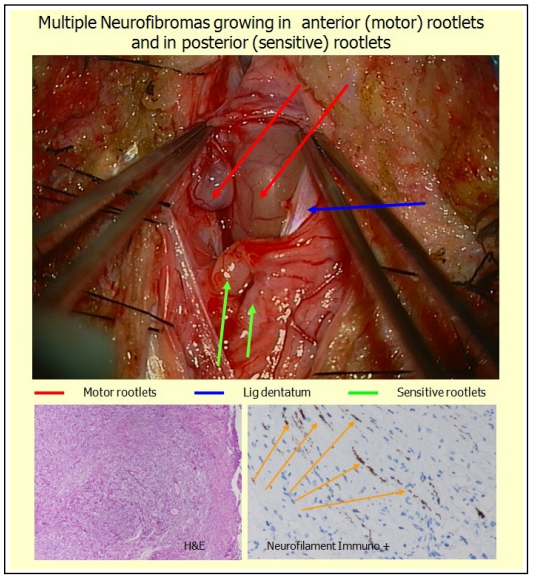
We performed a retrospective clinical reports review in a series of NF1 patients, followed and treated in a neurosurgical NF referral center. Demographic, clinical, anatomical, and pre and postoperative neurological status (McCormick´s classification) result data were analyzed.

Table I

Patient	Gender	Age	Clinic	Level	N Tumors	Preop	Postop	Follow-up
1	M	51	MONOPARESIS	C1-2 right Ant+Post	1	II	I	11 y
2	M	32	TETRAPARESIS	C5 left Ant root	3	III	I	10 y
3 (I)	M	42	MONOPARESIS	T11-12 left Ant+Post	2	III	II	9 y
3 (II)		46	TETRAPARESIS	C5-6-7 bilat Ant+Post	5	IV	IV	6 y
4 (I)	M	29	MONOPARESIS	T12 right Ant+Post	1	II	I	6 y
4 (II)		31	PARAPARESIS	C7-8 bilat Post roots	4	II	I	4 y
5	M	16	TETRAPARESIS	C2-C5 bilat Ant-Post	18	III	I	17m
6	M	42	TRIPARESIS	C2-6 right + C4-6 left Ant+Post	14	III	II	16 m
7	F	43	PARAPARESIS	C1-2+C5-6-7 left Post	6	II	I	13 m
8	M	34	TETRAPARESIS	C4, C7, C8 bilat Post	7	III	II – III* (infection)	8 m

Results

Ten surgical procedures were performed in a series of 260 patients in 11 years (2001-2012) (Table I). Two procedures at distant levels in two patients were evaluated as separated events. Most patients were male. Age ranged from 16 to 51 years. Clinical presentation was tri-quadriparesis in 5, paraparesis in 2, and monoparesis in 3 cases. The involved level was cervical in 8, and thoraco-lumbar in 2. Tumors grew from the anterior (7) and/or posterior (9) roots (Image). During the surgical procedure, intraoperative neurophysiological monitoring was performed (IONM: SSEP, MEP, Root identification) in the last 7 cases. Tumors were resected in all cases, totally in sensitive roots and subtotally in motor eloquent levels, sparing rootlets with small tumor nodules. Posterior vertebral elements were sectioned at the approach and reimplanted with titanium plates when C2 to C7 segments were treated. Pathological analysis in all cases confirmed Plexiform Neurofibromas, encasing neural fibers within the tumor tissue (Image 1). Most surgical procedures were followed by one clinical level upgrade, with full recovery in some patients (Table II). In one patient, with a longstanding severe quadriparesis, the deficit persists. No tumor recurred or progressed at 12m – 12y follow-up. No kyphotic deformity was observed in the follow-up.

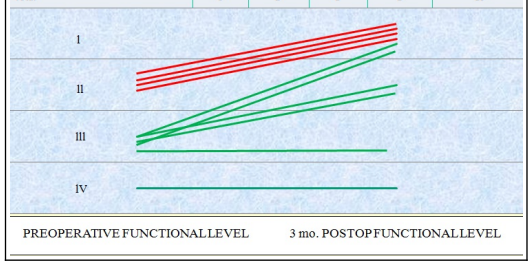


Conclusions

Spinal cord compression from spinal root Neurofibromas is an infrequently reported complication in NF1 (1, 2, 4, 6, 7). Although lesions are more frequently cervical, caudal spinal cord is also affected. As opposed to Schwannomas, Neurofibromas grow encasing neural fibers, and at motor as at sensitive roots. Subtotal resection should be performed in motor roots in order to spare motor function. Osteoplastic laminotomies can be performed for cervical segments instead of fusions, in order to avoid postoperative kyphosis (3,5). Resection of symptomatic neurofibromas is followed by very good results, except in patients with severe previous neurological deficit. Although asymptomatic spinal root neurofibromas should not be treated, surgery under IONM should be considered and planned as the initial symptoms appear for the best functional result.

Table II

Functional Pre-Op.	Funcional Post-Op. McCormick´s Scale				
	I	II	III	IV	Total
I No / Mild focal deficit					-
II Independent ambulation	4				4
III Ambulation with cane	2	2	1		5
IV Wheelchair dependent				1	1
Total	6	2	1	1	10



Learning Objectives

Identifying the characteristics and adequate timing for surgical treatment of NF1 spinal root neurofibromas.

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