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Management of spinal tumors in neurofibromatosis type 2 patients

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ABSTRACT

Objective: We sought to determine clinical characteristics of NF2 patients with spinal lesions and to define when and like the spinal tumors are a major problem in the treatment of patients with NF2.

Methods: The authors retrospectively reviewed the clinical records, neuroimaging studies, and follow-up data of the 34 patients with neurofibromatosis type 2, who were treated at our institution between 1998 and 2014. 23 patients harbored one or multiple spinal tumors.

Results: Patients with spinal tumors had a lower age at first symptoms of the disease, a higher number of intracranial meningiomas and non-vestibular schwannomas. 11 patients had one or more intramedullary tumors with MRI characteristics of spinal ependymomas. 22 patients had intradural extramedullary tumors. 7 patients presented with symptomatic spinal tumors on admission or developed symptoms during the follow-up. Only two intramedullary and four extramedullary tumors demonstrated growth in the mean radiological follow-up period of over 6 years. It was found that symptomatic both intra- and extramedullary tumors were associated with younger age at the onset of NF2-related symptoms. 2 patients with intramedullary tumors and 12 patients with extramedullary tumors underwent their tumors resection. In case of symptomatic tumors partial recovery was observed in two patients.

Conclusion: It seems that close surveillance with MR imaging is a reasonable option for asymptomatic spinal tumors. Nevertheless, intramedullary tumor removal in non-growing and asymptomatic cases may be an option when ABI implantation is considered. Symptomatic tumors and those of documented growth should be eligible for surgical intervention.

1. Introduction

Neurofibromatosis type 2 is an autosomal dominant syndrome predisposing to multiple benign tumors of the central and peripheral nervous system. The hallmark of this disease is the development of bilateral vestibular schwannomas, which occurs in 90–95% of patients [1–3]. Spinal NF2-associated tumors include schwannomas of spinal nerves, meningiomas and spinal cord ependymomas. Schwannomas of the spinal

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nerve root are frequently multiple and they account for almost 90% of extramedullary spinal tumors [4,5]. Intradural, extramedullary spinal meningiomas are present in about 20% of patients [1–4]. These tumors, however, may not be distinguished radiologically or even at the time of surgery. Ependymomas account for more than 75% of intramedullary spinal cord tumors associated with neurofibromatosis type 2 [4,6–8]. Their imaging evidence is found in 18–53% of patients but they cause clinical symptoms in fewer than 20% [4–6,9]. Intramedullary astrocytomas of the spinal cord and intramedullary schwannomas have been rarely reported in NF2 [4–9]. In contrast to sporadic tumors, the majority of NF2-related spinal tumors are asymptomatic during observation. Although several previous reports concerned clinical and imaging characteristics of spinal tumors little is known about their natural history in these patients. In this study we sought to evaluate probability of growth of various spinal tumor subtypes, identify prognostic factors for symptomatic cases and establish a strategy for the management of these tumors. Moreover we aim to assess the risk and effectiveness of surgical treatment for this tumor types. We compare NF2 patients with spinal tumors to those without in order to identify differences between the two patient subgroups. We assessed clinical characteristics, growth patterns and surgical outcome of spinal tumors in NF2 patients with long-term clinical and imaging follow-up.

2. Material and methods

2.1. Patient population

34 patients with neurofibromatosis type 2, as defined on the basis of the modified National Institute of Health (NIH) Consensus Panel Criteria [10], were surgically treated at our institution between 1998 and 2014. We have retrospectively reviewed the clinical records, neuroimaging studies, and follow-up data of the treated patients. Among 34 patients, in 11 we did not identified any spinal tumors whereas 23 harbored one or multiple spinal tumors. Patients with spinal tumors were divided into those with intramedullary tumors and those with extramedullary tumors or intradural tumors of the cauda equina. Some patients were included in more than one subgroup because they had tumors in both locations. Three symptomatic spinal tumors in 3 patients (1 meningioma in thoracic spine and 2 schwannomas in cauda equina) were resected in their childhood at another institution without sequel and were not included in the surgical treatment and growth rate analysis. We included in the natural growth study patients who harbored intra- or extramedullary spinal tumor and were observed for at least 1 year. Resected tumors were graded according to the WHO 2000 and the WHO 2007 [11] grading scheme.

2.2. Clinical and radiological follow-up

Our treatment philosophy assumed that the optimal strategy for each NF2 patient is to maintain good quality of life for as long as possible with special attention to maintain hearing. NF2 is not a one uniform illness but comprises several subtypes of tumor of the brain, spinal cord and peripheral nerves and as the spontaneous clinical course of the disease varies from patient to patient it may need different treatment strategies. The rule of conduct in every newly diagnosed case of NF2 was the performance of the brain and entire spine MRI diagnostics. Then all patients with spinal tumors had a clinical examination and spinal MRI study performed at least once a year. MRI was aimed at determining the presence of a tumor, defining its nature and in particular evaluating of any tumor growth in subsequent studies.

2.3. Statistical analysis

Data analyses were performed using Statistica version 12.0 (StatSoft) by means of Student's *t*-test and contingency tables using the Pearson Chi-square test or Chi-square test with Yates correction. Significance level was established at $p < 0.05$.

3. Results

3.1. Patient and tumor characteristics

There were 19 women and 15 men in the entire cohort of NF2 patients. Characteristics of the patients and the comparison between NF2 patients with spinal tumors and those without the lesions are given in Table 1. Among the 23 patients with spinal tumors non-vestibular schwannomas were significantly more frequently observed ($p < 0.05$). Besides, patients with spinal tumors had a lower age at first symptoms of the disease and had a higher number of intracranial meningiomas.

Table 1 – NF2 patients' characteristics.

Characteristic	No. of patients (%)		*p*-Value
	Spinal tumor present 23 (67)	Spinal tumor absent 11 (33)	
Female	14 (61)	5 (45)	0.63
Mean age at first symptoms (SD) (years)	19 (10.3)	23 (9)	0.28
Bilateral vestibular schwannoma	20 (87)	11 (100)	0.54
Non-vestibular schwannoma	**9 (39)**	**0 (0)**	**0.045**
Intracranial meningioma	16 (70)	5 (45)	0.33

SD – standard deviation.
Values in bold indicate statistical significance