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Multiple faces of children and juvenile meningiomas: A report of single-center experience and review of literature

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ABSTRACT

Objective: Pediatric meningiomas are extremely rare tumors often associated with various medical conditions. This report is an attempt to analyze the clinical behavior, pathological presentation and recommended management of both isolated and neurofibromatosis type 2 (NF2) related pediatric meningiomas in comparison to their adult counterparts.
Methods: The investigated group consisted of three male and six female patients suffering from meningiomas of the central nervous system, who were hospitalized at our department in years 2001–2012.
Results: In our group three children were diagnosed with NF2. Two were released with a suspicion of that disease. Three patients had multiple meningiomas at the first presentation. The most common manifestation were focal neurological deficits (47%). Meningiomas were mostly located at the brain convexity (29%). Histological examination revealed the domination of fibrous type in our group (29%). Apart from surgical management a preoperative embolization of the tumors' vessels was implemented in two children. Another three children underwent postoperative radiotherapy (RTh), one received a chemotherapy (ChTh).
Conclusion: Meningiomas in children differ from their counterparts in adults. Due to common coexistence of pediatric meningiomas and neurofibromatosis type 2, this group of patients should be taken under strict control, because of high risk of tumor recurrence.

1. Introduction

Although meningiomas are uncommon in children and adolescents they are undoubtedly an important and challenging tumors for pediatric neurosurgeons. Not only because they differ from their adult counterparts, but also because they are prone to accompany several medical conditions: neurofibromatosis type 2 (NF2), Rubenstein–Taybi syndrome and Castleman disease [1]. Meningiomas amount to 0.4–4.6% of all primary central nervous tumors (CSN) in pediatric group [2]. These may explain why their biological behavior and therapeutical guidelines still remain uncertain.

This report is an attempt to analyze the clinical behavior, pathological picture and recommended management of pediatric meningiomas in comparison to their adult counterparts and to elucidate the differences between sporadic and NF2-related meningiomas on the basis of own experience and literature.

2. Material and methods

Over a 11-year period (2001–2012) nine patients younger than 18 years were hospitalized at the Department of Neurosurgery at University Children's Hospital, Jagiellonian University Medical College. Altogether there were twenty-four meningiomas in the investigated group. All patients were analyzed retrospectively. The data collected from the hospital records included age, sex, clinical presentation, extent of resection, number of recurrences, associated factors (NF2) and histopathology. Preoperatively, all children were evaluated by neuroimaging (magnetic resonance imaging (MRI)/computed tomography (CT)). The tumors which increased in size after primary subtotal resection and caused new neurological deficits were considered as a regrowth. The tumor recurred if a total excision was documented at the primary surgery. The time interval and correlation to the histological subtype in recurred and regrown tumors were noted. The diagnosis of NF2 was based on the Manchester Clinical Diagnostic Criteria. Gene mutation research was not part of the protocol. Statistical analyses were performed using

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StatSoft version 10.0 and Microsoft Excel version 2010. We used χ^2 test to compare distribution of histological types of meningiomas.

3. Results

3.1. Clinical data

Nine children with meningiomas underwent a surgery in our department in years 2001–2012. We noticed a strong female predominance with six girls and three boys in the study group. Mean age at the first admission to the department was 11.3 years. The youngest child was 5 years old. The mean length of symptoms was 9.5 month ranging from 3 weeks to 3 years. Focal neurological deficits were the most common manifestation of a meningioma in our group (47%), followed by the symptoms of raised intracranial pressure (35%), and epilepsy seizures (18%).

3.2. Associated factors

Three out of nine children had associated NF2. We noted a positive family history in one patient (case 1). Two patients (case 4 and 5) were released home with a suspicion of that disease. Children with NF2 manifested additional tumors: bilateral vestibular schwannomas, schwannomas of peripheral and spinal nerves. Bilateral vestibular schwannoma appeared in all patients long after the first meningioma: after 2 months, 3 years and 5 years respectively (case 1, 2 and 3). In two of them right-sided deafness was diagnosed (case 1 and 3). A plexiform schwannoma of the skin was found in one patient with multiple meningiomas (case 4). None of the children underwent radiation in the past. We noticed neither associated Rubenstein–Taybi syndrome nor Castleman disease. Patients' age, gender, tumor location and histopathological aspects are summarized in Table 1.

3.3. Relapse

Solitary tumors at the first presentation were found in six (out of nine) patients. Three patients had multiple meningiomas when the symptoms began (case 1, 4 and 9), but only one of them had definite NF2. We noted five recurred tumors (case 1, 3 and 8) and one meningioma which regrew (case 6). Time to recurrence was defined as the time from total surgical excision to radiological reappearance of a new tumor.

3.4. Location

Convexity meningiomas were the most common in the series (29%), followed by skull base meningiomas (25%) and spinal canal meningiomas (25%). Parasagittal meningiomas comprised four out of twenty-four meningiomas (17%) and were more common than intraventricular ones (4%).

3.5. Histopathology

The results of histopathological aspects are summarized in Table 2. We observed lower WHO grading in children with NF2 in contrast to the children with sporadic meningiomas. In two patients the suspicion of NF2 came from neuropathologist (D.A.), who observed plexiform schwannoma in the excised skin tumor (case 4) and meningioangiomatosis (MA) (Fig. 1) in the excised brain mass (case 5). Most of meningiomas were benign (WHO grade I), four were grade II tumors and one grade III meningioma (Fig. 2). After comparing our data with those of other authors we noticed dissimilarity of distributions of histopathological types among all the groups ($p = 0.0001$ for our series vs. Kotecha et al.; $p = 0.024$ for

Table 1
Clinical features of 9 patients.

Case no.	Age at first manifestation	Sex	Diagnosis of NF2	Bilateral vestibular schwannoma	Number of meningiomas	Histological grading of meningioma	Location of meningioma(s)	Other tumors observed	Additional treatment
1	15	M	Yes	Yes	4	WHO I, WHO II	Parasaggital, skull base	Bilateral schwannomas of spinal nerves (L2)	Embolization
2	13	F	Yes	Yes	4	WHO I	Thoracic intradural	Schwannoma of intercostal nerve	No
3	13	F	Yes	Yes	7	WHO I	Parasaggital, skull base, intraventricular, thoracic intradural	Suspected lesion in spinal cord	Embolization, RTh
4	17	F	Suspicion of forme fruste	No	2	WHO I	Convexity, skull base	Plexiform schwannoma of the skin	No
5	8	F	Suspicion	No	1	WHO I	Skull base	No	No
6	14	F	No	No	1	WHO III	Convexity	No	ChTh, RTh
7	7	F	No	No	1	WHO II	Convexity	No	No
8	10	M	No	No	2	WHO I, WHO II	Convexity	No	RTh
9	5	M	No	No	2	WHO I	Convexity	No	No