



Intracranial Meningiomas in Children: Ten-Year Experience

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Meningiomas are central nervous system neoplasms derived from arachnoid cap cells. They are the second most common brain tumors after gliomas, but are rare in children. Furthermore, meningiomas exhibit different behavior in this age group. From 1997-2007, 7 children with brain meningiomas were treated at the Department of Neurosurgery, Brazilian National Cancer Institute, Rio de Janeiro, Brazil. They represented 2.7% of all brain meningiomas, and 2.1% of all brain tumors, in children followed during this period at our institution. There were 4 boys, and the mean age at diagnosis was 7.3 years. Headaches, seizures, and motor deficits were the most frequent signs at presentation. All patients underwent surgery, and total resection was achieved in 6 of 7 patients. Most lesions were World Health Organization class I meningiomas. There were no deaths, and patients were asymptomatic or demonstrated mild motor or sensory signs at follow-up. In conclusion, meningiomas are rare in the pediatric population. Most of the lesions are low-grade, and the prognosis in this setting is good. © 2008 by Elsevier Inc. All rights reserved.

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Introduction

Meningiomas are central nervous system neoplasms derived from arachnoid cap cells of the arachnoid villi [1]. They are the second most common brain tumors after gliomas, constituting 20% of all intracranial tumors [2]. The annual incidence is 2.3-3.1 per 100,000, but many lesions are asymptomatic. Most of them (90%) are benign, slow-grow-

ing tumors that commonly arise between ages 40-70 years. There is a female-to-male predominance of 2:1 in adults [3].

Meningiomas are relatively uncommon in childhood, representing 1-2% of all intracranial tumors in children [4]. Previous studies indicated that childhood meningiomas have different locations and behavior compared with those of adults. This retrospective study sought to describe the characteristics of pediatric patients presenting with meningioma at our institution from 1997-2007.

Methods

We reviewed the clinical charts of all patients with a diagnosis of intracranial meningioma admitted at the National Institute of Cancer, Rio de Janeiro, Brazil, from January 1997 to July 2007. Patients were included if they were 12 years old or younger at onset of signs, and if their diagnosis had been confirmed by brain histologic analysis. Brain magnetic resonance imaging was available in all cases.

To determine the percentage of brain meningiomas among all children with brain neoplasms, we also reviewed the medical charts of all patients 12 years old or younger who underwent brain-tumor surgery during the same period at our institution.

Results

Among 257 patients with a diagnosis of brain meningioma between 1997 and 2007, 7 were children (2.7%). During the same period, 325 children underwent surgery for brain tumors. Meningiomas represented 2.1% (7/325) of brain tumors in these children.

Four patients were boys (57%), and the mean age at onset of signs was 7.3 years (range, 1.5-12 years). The commonest sign at presentation was headache (85.7%), followed by seizures (57.1%) and motor deficits (28.5%). Although lesions in some patients achieved high volume, signs of increased intracranial pressure were absent in all children. None of the patients had clinical signs or a family history

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Table 1. Clinical and pathologic characteristics of children with meningioma

Sex	Age at Onset (yr)	Duration of Signs Before Diagnosis	First Clinical Manifestation	Location	Histologic Diagnosis
Female	4	2 months	Simple partial (motor) seizures, headache, motor deficit	Right cerebral hemisphere	Fibrous
Male	10	6 months	Headache	Right sphenoid wing	Meningothelial
Female	4	2 weeks	Simple partial (motor) seizures, headache, somnolence	Left cerebral hemisphere	Papillary
Female	8	1 week	Simple partial (motor) seizures, headache	Left frontoparietal lobe	Meningothelial
Male	1.5	2 weeks	Generalized tonic-conic seizure, headache, vomiting, motor deficit	Left temporal lobe	Meningothelial
Male	12	2 years	Headache	Left lateral ventricle	Transitional
Male	12	1 year	"Lump in the head"	Left occipital lobe	Meningothelial

of neurofibromatosis type II or of any other hereditary neurologic disease.

The locations of tumors are described in Table 1. The lesion was totally resected in 6 patients, and partially in one. In one patient, because of the large size of the lesion, total resection was achieved only after 3 surgical procedures (Fig 1).

In 6 of 7 patients, histologic analysis revealed a World Health Organization class I meningioma. No progression was observed in this group after a mean follow-up of 3.6 years (range, 0.5-9 years). One patient manifested a World Health Organization class III meningioma, and 10 months after the first surgery, magnetic resonance imaging of the brain revealed a recidive of the lesion. She underwent surgery, followed by cranial irradiation (54 Gy over 30 fractions). Three patients were left with mild motor sequelae, and one exhibited a sensory deficit on the right side. No deaths were recorded.

Discussion

Meningiomas originate from arachnoidal cap cells, which can be intracranial, intraspinal, or ectopic. They are the most common nonglial primary intracranial tumors, and consti-

tute 10-20% of primary central nervous system tumors [2]. Although meningiomas are common in middle-aged individuals, they are rarely evident in children. In our study, meningiomas were diagnosed in only a small fraction of all children who underwent brain tumor surgery. These data are in accordance with previous studies that reported a prevalence of 0.4-4.6% of all pediatric brain tumors [5]. The mean age at onset in our patients (7.3 years) was slightly lower than previously reported (10.1 years), but other studies usually included patients up to age 16 years, whereas ours was limited to the pediatric population.

Several features distinguish meningiomas in children and adults. We did not observe a clear sex predominance in our population, but meningiomas in children are more frequent in boys [6], whereas there is a 2:1 female predominance in adults [3]. It is known that a large proportion of meningiomas express hormonal receptors for several hormones, especially progesterone, that may modulate tumor growth [7]. This may explain the differences observed before puberty.

Meningiomas in children commonly arise in atypical locations, and may achieve larger volumes than in adults [8]. They have a predilection for the lateral ventricles, posterior

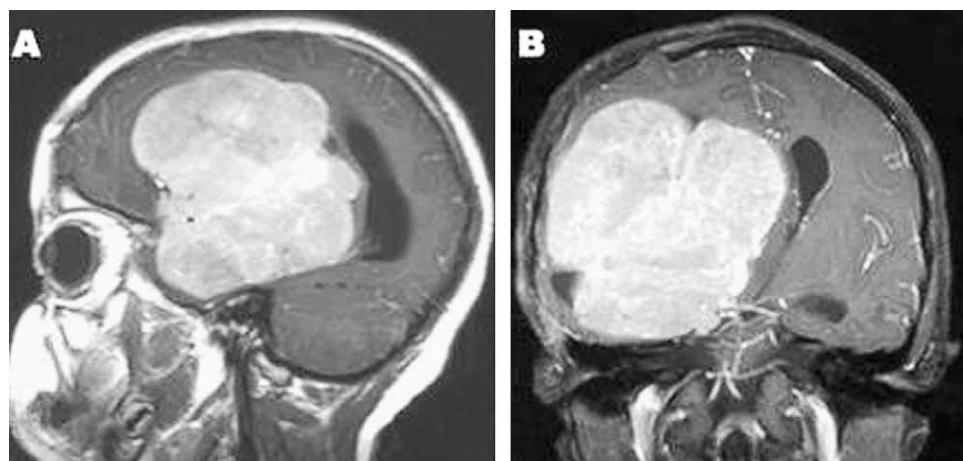


Figure 1. Magnetic resonance T_1 -weighted sequence ($TR/TE = 4400/111$ ms) reveals large, lobulated hyperintense lesion, suggestive of meningioma in right cerebral hemisphere. There is midline shift and compression of right lateral ventricle. (A) Sagittal view. (B) Coronal view.

fossa, and brain parenchyma without dural attachment, which may be explained by the presence of leptomeningeal elements inside the parenchyma or arachnoid cells in the choroid plexus. They also exhibit a cystic component more frequently than in adults. In all but one of our patients, lesions were found in typical adult locations.

There is a clear association between pediatric meningiomas and type 2 neurofibromatosis [9]. The *NF2* tumor-suppressor gene, located at the chromosome 22q12.1 region, encodes for the Merlin/Schwannomin protein. In a recent study, *NF2* gene deletion was observed in about 72% of cases, with corresponding absent or minimal Merlin protein expression according to immunohistochemistry [10]. Other known associations include previous radiation therapy, Klinefelter syndrome, and type I diabetes mellitus [9]. Our patients had not undergone genetic analysis, but there was neither a family history nor the development of clinical signs of any of these conditions during follow-up.

As observed in other series, the commonest signs at presentation were headache, probably related to intracranial hypertension, because the lesions can achieve larger volumes before diagnosis, followed by seizures and motor deficits [6].

The World Health Organization classification divides meningiomas into three grades: I, benign; II, atypical; and III, anaplastic/malignant. Each grade is predictive of growth, metastatic, and recurrence potential, as well as prognosis [7]. Early reports indicated a higher propensity for malignant degeneration in children [11], but there is recent evidence that the frequency is similar in adults [6]. The subtype more often observed in children is meningotheelial (grade I), which was also most frequently found in our series.

Although a conservative approach is acceptable in asymptomatic patients with indolent lesions, the main treatment for meningiomas is total surgical resection [9]. Radiation therapy is reserved for nonresectable lesions, or high-grade or recurrent tumors. Recurrence does not decrease regardless of when radiation therapy is administered, whether at first resection or on recurrence [12]. Moreover, because of the potential risk of radiation-induced lesions in an immature nervous system, radiation therapy should be avoided in very young children. Only one patient in this study underwent radiation therapy because of a recurrence of the lesion <2 years after the first resection. The

role of chemotherapy for pediatric meningiomas has not been well-established until now.

In conclusion, intracranial meningiomas are rare in the pediatric population. Most lesions were World Health Organization class I meningiomas. Total resection is associated with a better prognosis, and should always be attempted. Radiation therapy is reserved for recidives or high-grade lesions. Although our follow-up was too short for a definitive conclusion, the neurologic prognosis seems to be good in this population.

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