Intraventricular meningioma in childhood. Case report.

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Abstract
Central nervous system (CNS) tumors are the first location of solid tumors in childhood, with predominance of the embryonic lineage and relative absence of gliomas. Meningiomas are usually benign tumors of the CNS, arising from the arachnoid cell layer. They are a rarity in childhood, but when they do appear in this age group, the intraventricular location is more frequent than in adults and the usual location is in the trigone of the lateral ventricles and more rarely in the III and IV ventricles. These are slow-growing tumors; moreover, as all intraventricular tumors, they present an asymptomatic growth period, because tumor grows displacing the cerebrospinal fluid (CSF) from the ventricular cavity, so it does not affect the brain tissue. For this reason, they may attain a large volume before becoming symptomatic.

Key Words
Intracranial meningioma, intraventricular tumor.

INTRODUCTION

Tumors of the Central Nervous System (CNS) are the first location of solid tumors in childhood. Two incidence peaks of these tumors are described. The first, or pediatric presentation, occurs around the first decade of life and causes 2.2 to 2.5 cases per 100,000 children per year, with a very discrete predominance in males (1.1:1.0); the second is much wider, ranging from the third to the fourth decade of life, with its peak after 60 years. In the first peak, CNS neoplasms of embryonic lineage predominate with relative absence of gliomas. This continues until early adolescence; the incidence of CNS tumors typical of adults increasing thereafter. Intracranial meningiomas are very rare in childhood and, in general, the intraventricular location is uncommon; but when it does occur at pediatric age, it exceeds the percentage for the same location in adults.

CASE REPORT

An 11 year-old patient, otherwise healthy, presented with a history of headache of approximately 1 month duration before admission, predominantly in the morning, which became more intense in the last weeks. It was associated with emesis on three occasions and transient unconsciousness on one occasion, as well as blurred vision and later, onset of double vision. At the time of admission, neurological examination showed an alert patient, conscious, with clear and coherent language without limb motor deficit. On examination of the cranial nerve pairs, unilateral convergent strabismus of the left eye and diplopia due to paresis of the left sixth cranial nerve pair were detected. Ophthalmic fundus examination showed incipient papilledema. The rest of the neurological physical examination was negative.

Complementary blood, biochemistry, chest radiograph and abdominal ultrasound studies were all normal. Considering the findings of the neurological physical examination, cranium tomography is decided, in which a tumor mass was observed in the left ventricular trigone projection with dilation of the ventricular temporal horn on the same side.

For better definition and differential diagnosis from the radiological point of view, we decided to perform a magnetic resonance study (MRI) study, which showed an isointense mass on both T1 and T2. It was well-defined, localized in the left lateral ventricle at the level of the trigone and temporal horn, causing dilation of the latter. The density was homogeneous and size, approximately 4 x 3 cm. Intravenous contrast administration showed intense contrast uptake by the tumor, which largely corresponds to a meningioma type tumor (Figure 1).

The treatment of this patient in our setting involves transcortical surgical resection by a conventional surgical technique. Therefore, left parieto-occipital transcortical approach is planned and performed to minimize handling of important functional areas. Macroscopically, complete removal of the tumor was achieved (Figure 2), which was classified as transitional meningioma from the histopathological standpoint (Figure 3).

Evolutionarily, there has been no recurrence of the lesion six months after surgery, with favorable clinical and radiologi-

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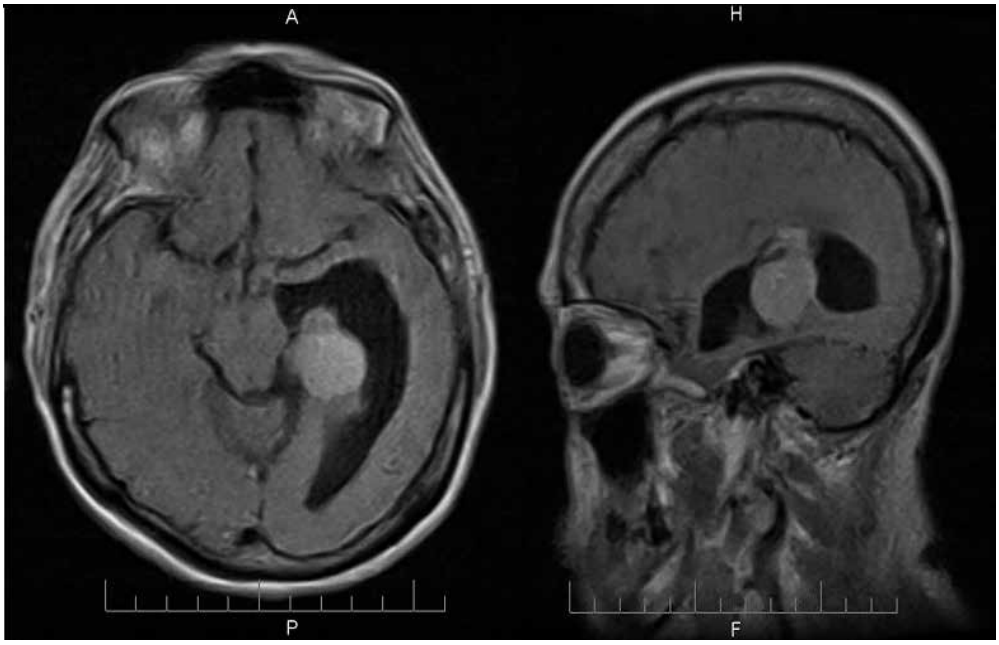

Figure 1. Magnetic resonance images showing the left intraventricular tumor

cal outcome (Figure 4), a progressively decreasing strabismus remaining as sequel.

DISCUSSION

Meningiomas are usually benign tumors that originate from meningothelial cells of the arachnoid covering, which can be found anywhere in the meninges. Intracranial meningiomas are rare in childhood, accounting for approximately 0.5 to 5.0 % of pediatric tumors; intraventricular meningiomas representing between 1.5 to 2.0 % of all intracranial meningiomas (1-4). In adults, their diagnosis is slightly more common in women than in men (1.0:1.4) and around the fifth decade of life, constituting between 13 and 27% of primary intracranial tumors.

Most meningiomas are low grade benign lesions and associated with symptoms of gradual intracranial pressure increase: headaches and convulsions, as the most frequent and others that depend on tumor size and location. They are rarely malignant lesions. Hence, the World Health Organization (WHO) provides the classification to predict clinical features, with a three-tiered system based on clinicopathologic correlation (5-6), in which Grade I are benign meningiomas; Grade II, atypical meningiomas and Grade III, malignant meningiomas.

The most common locations of meningiomas are: cerebral falx and parasagittal (25%), in the convexity (20%), sphenoid wings (20%), suprasellar region in the tubercle of the sella turcica (10%), olfactory groove (10%), cerebellopontine angle, tentorium, foramen magnum (posterior fossa 10%), intraspinal and other locations (3%) (7). Intraventricular meningiomas constitute from 0.5 to 2.0 % of all intracranial meningiomas, 12% of all intraventricular tumors and 20% of all neoplasms of the lateral ventricles. These can be located in the third and fourth ventricles (8) and, when found in the lateral ventricles, are usually located in the trigone. Meningiomas located here are believed to originate from arachnoid cells in the stroma of the choroid plexus (9).

The slow growth of these lesions, due to their biological nature, as well as intraventricular location, favor late onset of clinical manifestations. These clinical signs are usually secondary to increased intracranial pressure; focal neurological deficits and seizures are also frequently reported in the literature (10). Headaches are often intermittent, diffuse and ipsilateral at onset; but once intracranial hypertension is established, these headaches become continuous and frequently associated with vomiting. Ocular symptoms are subjectively expressed as decreased visual acuity in 50% of cases and papilledema is detected in fundus examination in 2 out of 3 cases.

Language disorders occur in approximately 40% of patients with tumors affecting the dominant hemisphere. Nonspecific psychiatric disorders may also occur, such as: depression, disorientation and behavioral disorders.

Differential diagnosis of intraventricular meningiomas should mainly be done with other types of tumors such as choroid plexus papillomas, low-grade astrocytomas, ependymomas, oligodendrogliomas, metastasis and lymphoma, among others.

Today, surgical treatment of meningiomas of the trigone remains a challenge, even with the new neurosurgical technologies available (11), which aim towards complete tumor resection. There are different approaches using conventional surgical techniques, including the posterior transcallosal, posterior transparietooccipital and transtemporal through the middle temporal gyrus, the latter two being transcortical. Application of endoscopic techniques allows exeresis of intraventricular lesions with reduced morbidity; however, these techniques may be limited in the case of large lesions with firm consistency, as is the case of meningiomas (3,11) and depending on the scientific development of the institu-

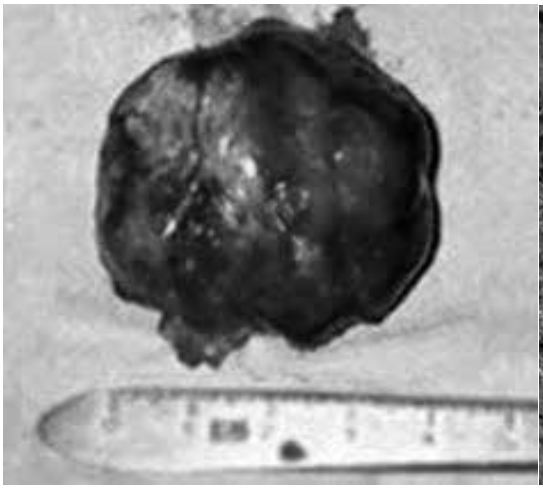

Figure 2. Excised tumor mass

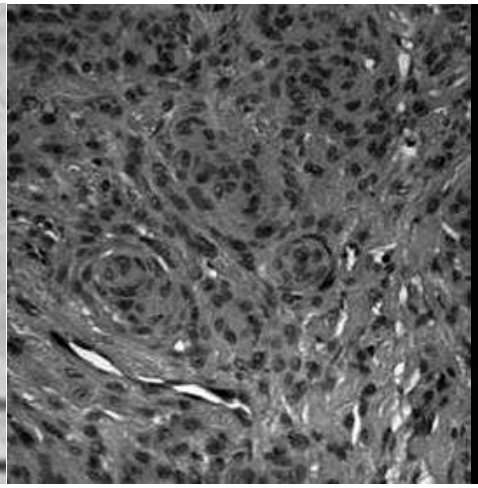

Figure 3. Histological pattern of the lesion removed.

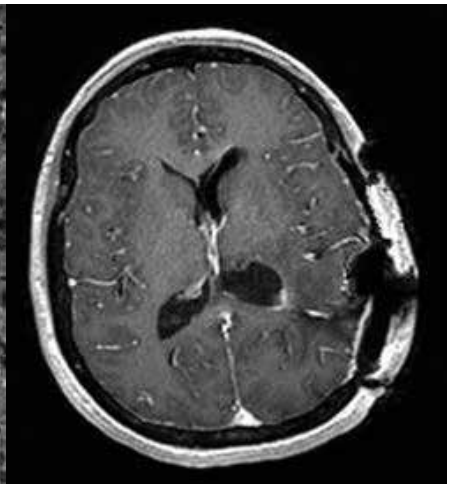

Figure 4. Postoperative CT image with contrast, 6 months after operation

tion where the surgery is performed.
In our particular case, the transcortical parieto-occipital route was used, due to the location of the lesion in the dominant hemisphere, with the aim of reducing transoperative morbidity of functional sites, as the language area, achieving complete resection of the lesion that had histopathological classification corresponding to benign meningioma, WHO group I.

CONCLUSIONS

Meningioma type tumors are rare in childhood, but can be diagnosed, preferably in their intraventricular location. The conventional surgical approach is transcortical, an option to be considered for their total elimination and cure in places with poor technological resources, without major sequelae for the patient in relation to the magnitude of the procedure.

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