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Clinical Study

Third ventricular meningiomas

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ABSTRACT

We report 13 patients with third ventricular meningiomas (TVM) and discuss the clinical, radiological, pathological and surgical features, as well as follow-up of these tumors. TVM are rare intracranial tumors, and because of this, there are few reports in the literature. Of 11,600 intracranial meningiomas that were surgically treated and pathologically confirmed at Beijing Tian Tan Hospital over a period of 10 years (2003–2013), 13 TVM were selected for a retrospective review. We recorded the clinical, radiological, pathological, and surgical data and statistically analyzed the preoperative, postoperative and 6 month postoperative Karnofsky performance scale (KPS) scores. TVM represented 0.11% of intracranial meningiomas. Radiologically, TVM were divided into three groups: anterior (n = 3), posterior (n = 3), and entire third ventricle (n = 7). Three patients (23.1%) were misdiagnosed preoperatively. Total removal was achieved in 61.5% (8/13) of patients, and subtotal resection was achieved in 38.5% (5/13). Pathologically, the tumors were World Health Organization (WHO) Grade I in 11 patients (84.6%) and WHO Grade II in two (15.6%). There were no statistically significant differences in the preoperative, postoperative, or 6 month postoperative KPS scores ($F = 0.814$; $p = 0.401$). TVM without dural attachments are rare neoplasms that should be differentiated from choroid plexus papilloma, craniopharyngioma, and pineocytoma. Surgery is the optimal treatment and may result in a favorable prognosis, and understanding of the radiological subtype can help with the choice of surgical approach.

1. Introduction

Intraventricular meningiomas are rare tumors, most of which are located in the lateral ventricle. They lack dural attachments and account for 0.5–5% of all intracranial meningiomas [1,2]. Third ventricular meningiomas (TVM) are extremely uncommon, representing only 0.1–0.18% of intracranial meningiomas [2–4]. An extensive review of the literature in 2003 found only 83 reported patients with TVM [5], and, to our knowledge, only seven further patients have been reported since 2003 in the English language literature. Therefore, a total of only 90 TVM have been reported to date. Most of these are from case reports or are sporadic cases in series of intraventricular meningiomas.

Due to their deep location and proximity to crucial neurovascular structures, surgery for TVM is challenging. The morbidity and mortality rates were relatively high in early surgical reports. In 1978, Avman et al. [3] summarized 14 surgical TVM patients from the literature. Seven of these patients died after surgery, and five survived with severe neurological deficits. However, with advances in microsurgical techniques, perioperative equipment and postoperative intensive care, this situation has greatly improved. In 2014, Karki et al. [6] summarized nine patients with TVM. In their series, two patients died after surgery, but all others had good outcomes.

Due to the rarity of the disease and the surgical challenges, further research is still needed to illuminate this rare entity. We retrospectively reviewed all TVM surgically treated and pathologically confirmed at Beijing Tian Tan Hospital over 10 years, from 2003–2013. Thirteen patients were identified, and the clinical, radiological, and pathological features, as well as surgical approaches and outcomes, were evaluated. To our knowledge, this is the largest single institution series of TVM.

2. Materials and methods

A review of medical records identified 11,600 patients who underwent surgical treatment of pathologically confirmed intracranial meningiomas at Beijing Tian Tan Hospital during the study period. TVM were selected for analysis through a retrospective review of clinical, radiological, surgical, pathological and

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follow-up records. Prior written and informed consent was obtained from every patient, and the study was approved by the Ethics Review Board of Beijing Tian Tan Hospital. Clinical information was collected, including age and sex, symptoms and signs, the duration from symptom onset to admission, and preoperative diagnosis. The radiologic data included density on CT scans, intensity of T1- and T2-weighted enhancement on MRI, tumor location, tumor size, tumor shape, and the presence of hydrocephalus (based on MRI). The tumors were divided into three types according to location: anterior, posterior and entire third ventricle (using the massa intermedia as a boundary). Tumor size was defined as the maximum diameter on MRI. The surgical findings included the surgical approach, tumor color, tumor texture, tumor border, tumor blood supply, extent of tumor removal, blood loss during the operation, surgical time, surgical outcomes, and surgical complications. The extent of tumor removal was classified as total, subtotal or partial, based on the surgical records and postoperative MRI. The pathological type was determined according to the 2007 World Health Organization (WHO) classification of tumors of the central nervous system [7]. The follow-up data regarding complications, recurrences, treatment after discharge, and patient deaths were collected. Karnofsky performance scale (KPS) scores were also evaluated preoperatively, postoperatively and at 6 months after surgery.

Statistical analyses were performed using SPSS software (version 17.0; IBM Corporation, Armonk, NY, USA). Statistical significance was defined as $p < 0.05$.

3. Results

3.1. Incidence and clinical features

The incidence of ventricular meningiomas and TVM was 2.1% (240/11,600) and 0.11% (13/11,600) of all meningiomas, respectively. The clinical features of TVM are summarized in Table 1. Of the 13 TVM patients, there were seven males and six females. The age at hospital admission was 31.3 ± 18.3 years (mean ± standard deviation [SD]; range: 7–56). The duration from symptom onset to admission was a median of 12 months (range: 3 months to 5 years). In terms of the major symptoms and signs, eight patients presented with headaches, four with visual deficits, four with somnolence, three with ataxia, and three with limb weakness. Hearing loss, urinary incontinence, memory defect, and hormone abnormalities occurred in two patients each. Upgaze palsy, dysgraphia, amenorrhea, and diabetes insipidus occurred in one patient each.

There were no patients with recurrent tumors or neurofibromatosis, and no patients underwent radiotherapy before hospital admission. Three patients (23.1%) were misdiagnosed preoperatively as having choroid plexus papilloma, craniopharyngioma, or pineocytoma (Fig. 1).

3.2. Radiological features

The radiological features are summarized in Table 1, 2. In terms of tumor location, there were three anterior, three posterior, and seven entire third ventricle type tumors (Fig. 2–4). Three tumors bulged into the bilateral lateral ventricles, two into the unilateral lateral ventricle, and two involved the septum pellucidum. In terms of the tumor shape, six were spherical, three were lobulated, and two were irregular. Dumbbell and cylindrical shapes were observed in one patient each. A total of nine patients had hydrocephalus, and nine had brain edema. The maximal tumor diameter was 5.1 ± 1.8 cm (mean ± SD; range: 2.3–8.2).

CT scans were available for seven patients. All tumors were hyperdense, and three had calcification. MRI examinations were performed for all patients. Seven tumors were isointense on T1- and T2-weighted images. One tumor each was isointense on T1- and isohypointense on T2-weighted images, isointense on T1- and hyper-hypointense on T2-weighted images, isohypointense on T1-weighted and hypointense on T2-weighted images, isointense on T1- and slightly hyperintense on T2-weighted images, slightly hypointense on T1- and isohyperintense on T2-weighted images and hypointense on T1- and T2-weighted images. MRI demonstrated homogenous enhancement in seven tumors, and heterogeneous enhancement in six.

3.3. Surgical findings and pathological types

The surgical findings are summarized in Table 1, 3. Before surgery, one patient underwent placement of a ventriculoperitoneal shunt because of obstructive hydrocephalus. To remove the tumor, a right frontal craniotomy with a transcallosal transforniceal approach was performed in eight patients. A right frontal craniotomy with a transcallosal transventricular transchoroidal approach was performed in two patients, a right frontal craniotomy with a transcortical transventricular transchoroidal approach was performed in two, and an occipital transtentorial approach was used in one. Most of the tumors were gray–white in color (n = 9), three were gray–red, and one was pink. The tumor texture was tenacious in four, a mixture of firm and tenacious in three, soft in three, a mixture of soft and tenacious in one, and a mixture of firm, tenacious and soft in one patient. Seven tumors had a rich blood supply and six had a moderate blood supply. All tumors, except one, were well defined from the surrounding tissues. Total removal was achieved in eight (61.5%), and subtotal resection was achieved in five (38.5%) patients.

The surgical duration was 7 ± 2.3 hours (mean ± SD; range: 4–13). The mean blood loss during surgery was 500 ml (range: 200–2000). Eight patients had a drainage tube inserted, and all tubes were removed 1–3 days postoperatively.

Regarding postoperative complications, electrolyte disturbances were found in six, hormone abnormalities in three, and subdural fluid accumulation in one patient. Diabetes insipidus, mutism, obstructive hydrocephalus, and intracranial infection occurred in two patients each. Two patients with obstructive hydrocephalus underwent a ventriculoperitoneal shunt placement after tumor removal. All complications were resolved before discharge, except in two patients who had hormonal abnormalities, one with diabetes insipidus who needed supplementary medication, and one with an intracranial infection who was discharged before the infection was controlled.

The pathological subtypes are shown in Table 1. There were four fibrous, three lymphoplasmacyte-rich, and three transitional subtypes. Meningothelial, chordoid, and fibrous tumors with brain invasion were observed in one patient each (Fig. 5, 6). WHO Grade I tumors occurred in 11 patients (84.6%), and WHO Grade II in two (15.6%).

3.4. KPS and follow-up

Data regarding the KPS are shown in Table 1. The median preoperative KPS was 80 (range: 70–100), and was unchanged postoperatively (median 80; range: 30–100). The median KPS 6 months after surgery was 90 (range: 0–100). There were no significant differences among the three groups on the test of repeated measures ($F = 0.814$; $p = 0.401$).

All patients were followed up for 65.3 ± 42.9 months (mean ± SD; range: 3–129). Two patients had hormonal abnormalities, and one had diabetes insipidus, which resolved during the