CLINICAL ARTICLE - BRAIN TUMORS

Radical resection of nongerminomatous pineal region tumors via the occipital transtentorial approach based on arachnoidal consideration: experience on a series of 143 patients

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Abstract

Backgroud The role of radical resection for nongerminomatous pineal region tumors is still controversial. The purpose of this study was to present the surgical results in a large series and evaluate the feasibility of radical surgical strategy.

Methods We retrospectively reviewed the records of 143 patients with nongerminomatous pineal region tumors surgically treated via an occipital transtentorial approach between 2000 and 2011. The tumor was small (<2 cm) in 14.7 % of patients, medium (2-4 cm) in 52.4 %, and large (>4 cm) in 32.9 %. Results Gross total tumor removal was achieved in 91.6 % of patients, subtotal in 7.0 %, and partial in 1.4 %. Histological diagnosis was nongerminomatous germ cell tumor in 41.3 %, pineal parenchymal tumor in 14.7 %, glial tumors in 28.7 %, and miscellaneous in 15.4 %. The overall complication and mortality rate was 18.2 % and 0.7 %, respectively. Permanent morbidity occurred in 5.6 % of patients, including hemianopsia in 3.5 % and Parinaud syndrome in 2.1 %. Hydrocephalus was resolved in 82.1 % without surgery for the CSF diversion. Sixty-eight patients with malignant tumors underwent radiotherapy; 35 also received adjuvant chemotherapy. One hundred thirty patients were successfully followed up with a mean duration of 43 months. Finally, 86.9 % of the patients achieved a favorable functional outcome (mRS≤

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2), 3.1 % had an mRS score of 3, 1.5 % had an mRS score of 4, and 8.5 % had died (mRS=6).

Conclusions Radical surgery was recommended as the optimal treatment for nongerminomatous pineal region tumors. Favorable results could be achieved by experienced neurosurgeons. Hydrocephalus could be cured by radical tumor removal in the majority of cases. The occipital transtentorial approach was indicated for most pineal region tumors, but surgeon's preference and experience should also be considered. New understanding of the arachnoid membranes of this region may be helpful for tumor resection.

Keywords Arachnoid membrane · Occipital transtentorial approach · Pineal region tumors · Surgery

Introduction

Pineal region tumors present with a spectrum of histological heterogeneous neoplasms from benign to malignant and have a higher incidence in Far Eastern countries (2.1-3.2 %) than in Western countries (0.4-1 %) [1–4]. The diversity of pineal region tumors makes histology-specific therapy mandatory for improving outcome [5–7]. It has been well established that surgical resection plays a small role in the management of germinomas because of their sensitive response to radiotherapy or chemotherapy. However, the optimal treatment strategy for nongerminomatous pineal region tumors remains controversial, especially for malignant tumors or tumors containing mixed components.

With advances in microsurgical technique and postoperative care in recent years, encouraging surgical outcomes of pineal region tumors have been improved dramatically with the mortality reduced to a level under 4 % [5–10]. However, these tumors are still surgically challenging to the neurosurgeon

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because of the complicated adjacent anatomy and deep location. In most publications, special emphasis was given to the anatomical relationship between deep cerebral veins and the tumor [2, 6, 8, 9, 11–18]. The relationship between the tumor removal and arachnoid membranes distributing in this region has not been well described. Besides, the role of radical surgery in the recovery of the accompanying hydrocephalus was also seldom mentioned in previous studies. Therefore, we presented a large consecutive series of 143 patients with nongerminomatous pineal region tumors undergoing radical resection via the occipital transtentorial approach. The surgical results, outcome of hydrocephalus, and follow-up results were described in detail. Our purpose was to evaluate the feasibility of an aggressive surgical strategy for these tumors based on the arachnoid membranes and a specific approach.

Materials and methods

Neither approval of the institutional review board (IRB) nor patient informed consent were required by the ethics committee of our institution (IRB of Nanfang Hospital) for retrospective analyses of patients' records and imaging data.

Patient characteristics

Between 2000 and 2011, 143 patients with nongerminomatous pineal region tumors underwent surgical resection using an occipital transtentorial approach at the Department of Neurosurgery, Nanfang Hospital, Guangzhou, China. Of these patients, there were 97 males and 46 females, and the mean age was 18.0 years (4–67 years). A total of 76 (53.1 %) tumors occurred in children, and 67 (46.9 %) occurred in adulthood. The most common presenting symptoms and signs were obstructive hydrocephalus and Parinaud's syndrome, which were noted in 106 (74.1 %) and 57 (39.9 %) patients, respectively. Other major clinical manifestations included cerebellar syndrome in 23 (16.1 %), hemisyndromes in 12 (8.4 %), hearing impairment in 9 (6.3 %), cognitive/memory disturbances in 7 (4.9 %), and precocious puberty in 2 (1.4 %) cases.

Tumor markers

All patients underwent serum analysis for human β -human choriogonadotropin (β -HCG) and α -fetoprotein (AFP). The cerebrospinal fluid (CSF) was obtained in 118 patients for analysis of β -HCG and AFP through external ventricular drain (EVD), endoscopic third ventriculostomy (ETV), or lumbar puncture. Elevated levels of CSF tumor markers were observed in 18 (30.5 %) of 59 patients with nongerminomatous germ cell tumors (NGGCTs).

Neuroradiological studies

All patients underwent MRI routinely for evaluation of the tumor intensity, size and extension, and relationship between the tumor and deep cerebral veins. CT scans were also performed in all patients to detect calcification, tumor density, or intratumoral hemorrhage for the differential diagnosis. The tumor size was determined by the maximal diameter on MRI, and the tumors were thus classified into 3 groups: small (<2 cm) in 21 (14.7 %) patients, medium (2–4 cm) in 75 (52.4 %), and large (>4 cm) in 47 (32.9 %). The mean tumor size was 38.3 mm (13–67 mm).

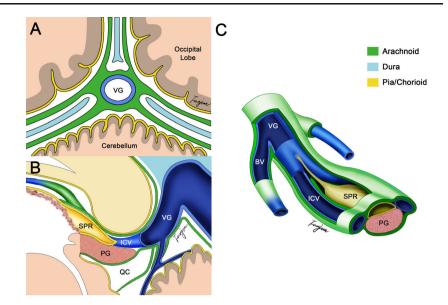
Preoperative intervention

Permanent CSF diversion procedures, including shunt placement and endoscopic third ventriculostomy (ETV), were performed prior to the open surgery in 11 and 2 patients, respectively. Fifteen patients underwent external ventricular drainage (EVD) because of rapid deterioration. Twenty-four patients had received direct radiotherapy or chemotherapy 6 to 35 months before admission without histological diagnosis.

Surgical techniques

The patients were placed in the prone position or the threequarters prone position. In the prone position, the head is slightly rotated with the approach side down to facilitate gravity-dependent retraction of the occipital lobe from the falcotentorial region. For most patients with no or mild hydrocephalus, brain relaxation could be easily achieved by the egress of CSF from the perimesencephalic cisterns. Otherwise, when brain relaxation was still unsatisfactory or the patient presented with severe hydrocephalus, an intraoperative puncture of the occipital horn was always necessary. The puncture point was usually located 6-7 cm above the transverse sinus and 3 cm lateral to the midline, and the catheter was then passed toward the mid-pupillary line. The procedure should be performed before the opening of the dura in order to avoid occipital contusion caused by high intracranial pressure. When an EVD was placed before surgery, it was clapped 8 h before surgery to allow ventricular dilation.

Because of the density of the important deep venous system in the pineal region, it is mandatory to remove tumors through the narrow space between these veins. Based on our anatomical studies on the arachnoid envelope over the pineal region (Fig. 1) [19–21], different surgical techniques were used to facilitate the procedure and reduce the risk of venous injury. For those tumors located within the arachnoid envelope, the envelope should be sequentially dissected prior to tumor removal to minimize the retraction or injury of the deep veins (Fig. 2 and Video 1). For tumors arising outside the envelope, however, the arachnoid envelope should be Fig. 1 Schematic drawings of the arachnoid envelope over the pineal region. a Coronal view; b sagittal view; c 3D anterolateral view. BV, Basal vein; ICV, internal cerebral vein; PG, pineal gland; QC, quadrigeminal cistern; SPR, suprapineal recess; VG, vein of Galen



preserved as intact as possible to provide a protective boundary for deep veins during tumor removal (Fig. 3).

En-bloc resection was recommended in cases of small tumors. For large and tough tumors, however, intratumoral decompression or piecemeal resection was necessary to avoid venous retraction. The internal cerebral veins were frequently displaced superiorly by the tumor and hidden under the splenium of the corpus callosum. In such cases, a cotton sheet was inserted into the velum interpositum cistern to push the tumor downward to enter the surgical field. This technique was especially helpful when the anterior part of the tumor projected into the posterior third ventricle. Tumors slightly extending to infratentorial or contralateral space could be well exposed by retraction of the tentorial edge or falx. Otherwise, a tentorial or falx incision was necessary to gain additional exposure. At the end of tumor removal, the suprapineal recess should be opened if still kept intact, in an attempt to relieve the hydrocephalus without a second procedure.

Results

Extent of tumor resection

The extent of tumor resection was evaluated by both intraoperative findings and immediate MRI review within 48 h after

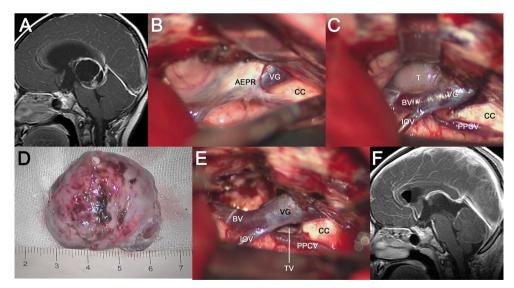


Fig. 2 A case of immature teratoma. **a** Preoperative sagittal MRI; **b-e** intraoperative photographs. In **b**, the tumor was located within the arachnoid envelope and remained invisible before dissection of the envelope; in **c**, the tumor was exposed after sequential dissection of the envelope; in **d**, the tumor was removed en bloc after dissection; in **e**, the vein of Galen

and its tributaries were well preserved without injury after tumor removal. **f** Immediate MRI after surgery demonstrating total tumor removal. AEPR, arachnoid envelope over the pineal region; BV, basal vein; CC, corpus callosum; IOV, internal occipital vein; PPCV, posterior pericallosal vein; T, tumor; TV; third ventricle; VG, vein of Galen surgery (Fig. 4). However, in one patient who died early postoperatively, the extent of tumor removal could only be estimated by the intraoperative findings. Accordingly, gross total resection (GTR) was defined as 100 % macroscopic removal of the tumor or no visible tumor remaining, subtotal resection (STR) was defined as>80 % but<100 % resection, and partial resection (PR) was defined as no more than 80 % resection. GTR was achieved in 131 patients (91.6 %); 10 patients (7.0 %) had STR, and 2 (1.4 %) underwent PR.

Pathohistology

All 143 tumors were histologically verified. The distribution of histological subtypes is noted in Table 1. Nearly half of the tumors (59 cases, 41.3 %) were NGGCTs, in which teratomas (29 cases, 20.3 %) and mixed GCTs (19 cases, 13.3 %) were most common subtypes. The histological diagnosis was a pineal parenchymal tumor in 21 patients (14.7 %), neuroepithelial tumors in 41 (28.7 %), and miscellaneous in 22 (15.4 %).

Outcome of hydrocephalus

Of the 106 patients presented with hydrocephalus preoperatively, permanent recovery was achieved in 87 (82.1 %) patients after radical tumor removal; 8 (7.5 %) patients remained unchanged, and 11 (10.4 %) recovered temporarily but had a recurrence within 1 to 3 months, possibly due to the reocclusion of the opening of the posterior third ventricle after surgery. Finally, 19 patients needed a second CSF diversion surgery, including ETV in 16 and shunting in 3. Of the 11 patients who had undergone shunt placement before tumor removal, 4 (36.4 %) required shunt revision or ETV due to shunt malfunction. (Fig. 5).

Adjuvant therapy

Sixty-eight patients with malignant tumors (including 37 NGGCTs, 7 pineal parenchymal tumors, 21 neuroepithelial tumors, and 3 miscellaneous tumors) underwent postoperative radiotherapy; 35 of these patients also received adjuvant chemotherapy according to standard protocols [22–24].

Complications

Overall postoperative complications occurred in 26 (18.2 %) of 143 patients. The most life-threatening complication was intracranial hemorrhage, which was noted in five patients (3.5 %). All these patients required an urgent reoperation to remove the hematoma. However, one patient died within 3 days because his parent refused the reoperation. Homonymous hemianopsia was the most common complication associated with surgery and occurred in 23 patients

(16.1 %). However, the condition began to improve in the early postoperative period in most patients, and a permanent visual field defect was observed in only five patients (3.5 %). All five patients had hemianopsia immediately after surgery and MRI showed T2 hyperintense areas in the medial occipital lobe, suggesting damage to the visual cortex attributed to the excessive brain retraction. It should be mentioned that most of these permanent defects occurred in our early period. With increased experience, permanent hemianopsia only occurred in 1 of the last 81 cases (1.2 %, 2006-2011). New cases of Parinaud syndrome occurred in nine patients (6.3 %), three (2.1 %) of whom developed permanent disturbances. Other complications were mostly transient and reversible, including mild hemiparesis in three patients, bacterial meningitis in three, mental dysfunction in two, and subdural hygroma in two (Table 2). Overall permanent morbidity was noted in eight patients (5.6 %), most commonly associated with large and malignant tumors.

Follow-up and prognosis

One hundred thirty (90.9 %) patients were successfully followed up by way of outpatient visit, telephone, or e-mail. We were unable to get information on the remaining 13 (9.1 %) cases after discharge from the clinic. The mean duration of follow-up was 43 months (range, 6–125 months). The final mortality and functional outcomes were determined according to the modified Rankin Scale (mRS) score at the end of follow-up. Finally, 113 patients (86.9 %) achieved a favorable functional outcome (mRS \leq 2), 4 (3.1 %) had an mRS score of 3, 2 (1.5 %) had an mRS score of 4, and 11 (8.5 %) had died (mRS=6) (Table 3). The 11 patients included those with pineoblastoma (*n*=2), NGGCT (*n*=5), and astrocytoma (*n*=4). All patients with benign or low-grade lesions are still alive.

Discussion

Role of radical surgery

The primary role of radical surgery is to obtain a sufficient volume of tumor tissue for accurate histological diagnosis of pineal region tumors, which has been regarded as an important step in guiding histology-specific therapy [5–7]. Some authors chose stereotactic or endoscopic biopsy as the initial surgical decision and mentioned minimal invasiveness and low risk associated with both procedures [25–30]. Endoscopic procedures have even demonstrated the potential to relieve hydrocephalus. However, the disadvantages of both methods lie mainly in the limited tissue sampling and insufficient ability to control bleeding. Kanamori et al. [31] performed endoscopic biopsy in 18 patients with pineal region tumors and found

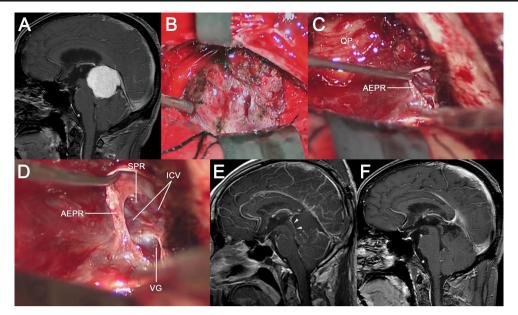


Fig. 3 A case of meningioma. a Preoperative sagittal MRI; b-d intraoperative photographs. In b, the tumor was situated outside the arachnoid envelope; in c, the arachnoid envelope was preserved intact at the end of tumor removal, providing a protective boundary for deep veins; in d, a detailed observation of the surgical field under higher magnification showed the content of the arachnoid envelope. e Immediate MRI after

that endoscopic biopsy may be useful for distinguishing GCTs from other lesions, but not for determining the subtypes of GCTs in patients with normal and moderately elevated levels of tumor markers. In contrast, open surgery allows for obtaining a maximal tissue sample and provides wide exposure to lower the risk of bleeding, especially in large and highly vascular tumors. Furthermore, for those patients with nongerminomatous tumors, a second open surgery was

surgery demonstrating total tumor resection and favorable preservation of the quadrigeminal plate (arrowhead). e A 2-year follow-up MRI showing no evidence of tumor recurrence. AEPR, arachnoid envelope; ICV, internal cerebral vein; QP, quadrigeminal plate; SPR, suprapineal recess; VG, vein of Galen

usually inevitably required, and the accumulated risk associated with two surgical procedures was obviously higher than a single open surgery.

A second role of radical surgery is to provide maximal removal of tumor volume. It has been clearly defined that aggressive surgical resection is the optimal treatment for benign or low-grade pineal region tumors involving meningiomas, mature teratomas, pineocytomas, and epidermoids

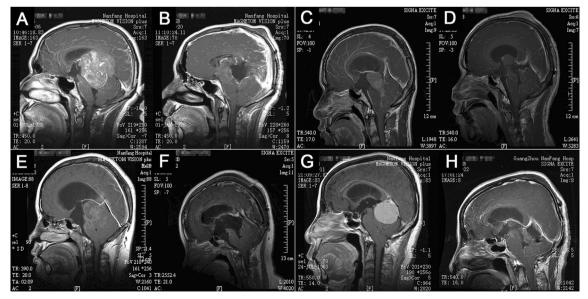


Fig. 4 Postoperative immediate sagittal MRI showing total tumor removal compared with preoperative MRI in several cases with pineal region tumors. **a** and **b** Mixed germ cell tumor in a 13-year-old male; **c**

and **d** pineoblastoma in a 22-year-old male; **e** and **f** oligodendroglioma in a 16-year-old female; **g** and **h** meningioma in a 56-year-old male

Table 1 The distribution of histological subtypes in 143 patients with nongerminomatous pineal region tumor

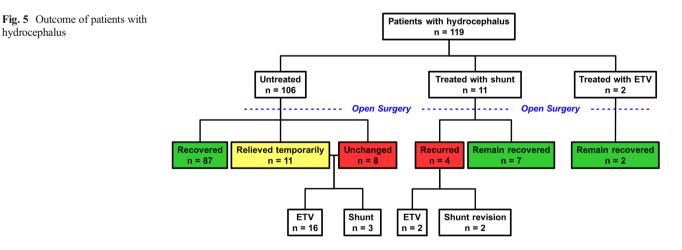
Pathology	Number
Nongerminomatous germ cell tumor	59
Mature teratoma	12
Immature teratoma	15
Malignant teratoma	2
Mixed germ cell tumor	19
Choriocarcinoma	6
Embryonal carcinoma	3
Yolk sac tumor	2
Pineal parenchymal tumor	21
Pineocytoma	12
Intermediate differentiation	3
Pineoblastoma	6
Neuroepithelial tumor	41
Pilocytic astrocytoma	6
Fibrillary astrocytoma	5
Oligodendroglioma	9
Anaplastic astrocytoma	4
Glioblastoma	5
Ependymoma	2
Anaplastic ependymoma	2
Subependymoma	1
Ganglioglioma	1
Dysembryoplastic neuroepithelial tumor	2
Central neurocytoma	2
CNS primitive neuroectodermal tumor	2
Miscellaneous	22
Meningioma	16
Papillary tumor	2
Epidermoid cyst	2
Lipoma	1
Metastatic tumor	1

Table 2 Postoperative complications

n (%)
26 (18.2 %)/8(5.6 %) ^a
1 (0.7 %)
23 (16.1 %)
18 (12.6 %)
5 (3.5 %)
9 (6.3 %)
6 (4.2 %)
3 (2.1 %)
5 (3.5 %)
3 (2.1 %)
3 (2.1 %)
2 (1.4 %)
2 (1.4 %)

^a The numbers in individual rows will not add up to the total number of complications because more than one complication may occur in a single patient

[8, 32–34]. For malignant tumors, however, no consensus has been reached on the extent of resection. With the exception of pure germinomas, malignant pineal region tumors frequently show poor response to either radiotherapy or chemotherapy. Under the circumstance, radical surgery can eliminate the tumor mass effect, reduce the tumor burden prior to adjuvant therapy, or remove the tumor components resistant to adjuvant therapy. Numerous reports have associated extensive resection of the malignant tumors with an improved response to adjuvant therapy and increased survival rate, although most studies are retrospective [35-38]. Another benefit of radical resection is the considerable recovery rate of hydrocephalus without a second procedure, which was noted in 82.1 % patients of this series. The risk of postoperative hemorrhage into the residual tumor bed may also be reduced by radical surgery. Most importantly, all the benefits should be established on the basis of acceptable mortality and morbidity.



hydrocephalus

The radical surgical strategy was further justified by the low rates of mortality and permanent morbidity (0.7 % and 5.6 %, respectively) achieved in this series.

Selection of surgical approach

Various surgical approaches were adopted in the treatment of pineal region tumors [2, 6–9, 16, 17, 39, 40]. The supracerebellar infratentorial approach is the most widely used approach, followed by an occipital transtentorial and interhemispheric approach. The results of recent major surgical series using different approaches are listed in Table 4 [1, 2, 6–9, 15–17, 41–45]. The overall GTR and mortality rates of surgery were comparable between different approaches. However, neither the transient nor permanent morbidity could be compared between series because of the insufficiently provided data in nearly half of the series and the lack of unified standards of morbidity. Besides, the tumor size was not presented in most studies. Therefore, it was difficult to conclude which approach is the optimal one.

As we know, each approach has its advantages and disadvantages. The supracerebellar infratentorial approach, placing the patient in the sitting position, allows a relaxed natural surgical corridor to the pineal region without cerebellar retraction, reduces the risk of bleeding, and provides a clean surgical field because of the low venous pressure and outflow of fluid. Other benefits include facilitating dissection of the tumor from the deep venous system and avoiding cortical injury during the approach. The main limitation of the supracerebellar infratentorial approach is the risk of air embolism, although it rarely occurs. We have also used this approach in some cases. However, we found that the patient's neck had to be extremely flexed in a sitting position, especially when a steep tentorial apex existed [14]. In these cases, the surgeon had to lean forward with the arms outstretched to overcome the long working distance, which may cause discomfort and rapid fatigue, as mentioned by Reid and Clark [40]. In comparison, the occipital transtentorial approach allows the surgeon more comfort and easy access to the pineal region. This is one of the main reasons why we preferred the occipital transtentorial approach in dealing with most pineal region tumors.

On the other hand, the supracerebellar approach is usually considered to be indicated for relatively small tumors without lateral expansion and primarily below the deep cerebral veins [9, 13, 40]. Besides, a very steep tentorium or extension of the tumor above the deep veins is a relative contraindication to the supracerebellar approach [13, 39]. In contrast, the occipital transtentorial approach is more appropriate for larger tumors because it can provide an excellent view of the tectum and tentorial notch [13]. It should also be noted that medium- and large-size tumors accounted for nearly 90 % of all cases in our series. The goal of radical tumor resection may not be achieved in some cases when using the supracerebellar

approach. This is another reason why we choose the occipital transtentorial approach as the main method to remove pineal region tumors (in approximately 90 % of all cases). Based on our experience, this approach was indicated for most of the pineal region tumors.

The major limitation of the occipital transtentorial approach is the relatively difficult manipulation between complex deep cerebral veins during tumor removal, especially for large tumors. However, based on our increased experience and new understanding of arachnoid membranes in the pineal region, this is not such a big problem. Another drawback of the transtentorial approach is postoperative hemianopsia due to intervention of the medial occipital lobe. The hemianopsia, although mostly transient, developed into a permanent defect in 3.5 % of patients in this series. A possible reason for this complication was excessive retraction of the occipital lobe or vein injury when managing large tumors in our early period (2000-2005). With increasing experience, the permanent hemianopsia was reduced to a low level of 1.2 % in the last 81 cases (2006-2011). Based on our experience, the risk of permanent morbidity can be reduced by several methods: (1) do not use brain retractors as far as possible when managing small or medium tumors; (2) if a retractor must be used, place it superiorly to avoid direct contact with the visual cortex around the calcarine fissure; (3) use retractors more to hold the brain out of the way rather than aggressively retract.

Overall, there is not a specific surgical approach suitable for all pineal region tumors. The optimal approach for a given pineal region tumor depends on the tumor size, anatomical features of the tumor extension, the tumor's relationship to the deep cerebral veins, the intended extent of tumor removal, and the surgeon's experience and preference [5, 7, 39].

Arachnoidal consideration during tumor removal

Numerous anatomical and surgical studies of the pineal region have been reported in the past literature, but the arachnoid membranes of this region as well as their relationships with neighboring structures were seldom described. Based on our

 Table 3
 The mRS score of follow-up patients with different subtypes of pineal region tumors

	mRS score							
	0	1	2	3	4	5	6	
Pathological subtype								
NGGCT	38	4	7	1	1	—	5	
Pineal parenchymal tumor	12	2	2	1		_	2	
Neuroepithelial tumor	20	4	5	2	1	_	4	
Miscellaneous	13	3	3			_		
Total number	83	13	17	4	2	0	11	

Table 4 Results of recent major surgical series for pineal region tumors using different approaches

Authors	Year	Cases	Approach			Mean tumor	GTR	Surgical	Major	Permanent
			ITSC	OTT	Others ^a	size (mm)		mortality	morbidity	morbidity
Luo et al. [16]	1989	64	_	100 %	_	NA	21 %	10 %	NA	NA
Pluchino et al. [43]	1989	25	100 %	_	_	NA	25 %	8 %	NA	NA
Vaquero et al. [45]	1992	29	31 %	13.8 %	55.2 %	NA	NA	10.3 %	NA	NA
Bruce and Stein [8]	1995	160 ^b	86.3 %	13.7 %	_	NA	45 %	4 %	> 34 %	> 10 %
Cho et al. [1]	1998	33	_	100 %	_	NA	54.5 %	0	NA	> 21 %
Pendl [7]	2002	111 ^c	67.6 %	14.4 %	18 %	NA	69 %	3 %	NA	4 %
Tanaka et al. [17]	2003	82	—	100 %	_	NA	61 %	0	NA	12 %
Konovalov et al. [9]	2003	244 ^d	33.8 %	54.2 %	12 %	32	58 %	8.2 %	NA	> 20 %
Hernesniemi et al. [6]	2008	119	93.3 %	6.7 %	_	NA	88 %	0	18 %	4.2 %
Sajko et al. [44]	2009	39	100 %		_	NA	59 %	0	33.3 %	NA
Kodera et al. [15]	2011	26	100 %		_	19	88.5 %	0	7.7 %	0
Jia et al. [2]	2011	150	_	_	100 %	NA	86 %	0	NA	> 10 %
Oliveira et al. [42]	2013	32	100 %		_	NA	70 %	0	NA	0
Mottolese et al. [41]	2014	233	—	100 %		NA	57.9 %	0	NA	> 2 %
Our series		143	—	100 %		38.3	91.6 %	0.7 %	18.2 %	5.6 %

^a Other approaches included transcallosal, subchoroidal, and transventricular ones

^b One hundred sixty operations in 154 patients

^c One hundred eleven operations in 100 patients

^d Two hundred forty-four operations in 201 patients

ITSC infratentorial supracerebellar approach, OTT occipital transtentorial approach, GTR % of patients with gross total resection

previous anatomical studies [19-21], the vein of Galen and its tributaries, as well as the pineal body and the suprapineal recess, were typically wrapped by a thickened arachnoid envelope over the pineal region. So far, no evidence has explained how the arachnoid membrane influences the growth pattern of pineal region tumors for their invasive entities. However, it is conceivable that the arachnoid membrane may play a role in benign or low-grade tumors and even in malignant tumors at their early phase. This presumptive theory was especially useful in our series due to the fact that benign or low-grade tumors account for nearly one half of all cases. As such, understanding of the relationship between the tumor and the arachnoid envelope may be helpful to protect deep veins during tumor removal. For example, it could be useful when deciding whether and how the membranes should be sequentially dissected or carefully reserved prior to tumor resection. We believe that surgical strategies based on arachnoid considerations may at least partially contribute to the favorable results achieved in this series.

Management of hydrocephalus

Treatment of obstructive hydrocephalus is one of the most important goals for surgical management of pineal region tumors. For a given patient with a nongerminomatous pineal region tumor, the treatment of the accompanying hydrocephalus should be carefully planned during the perioperative period based on several concerns: (1) shunt placement should be avoided whenever possible [5, 29,39]; (2) a single-stage surgical procedure is preferred to a two-stage one except for emergency conditions; (3) the intervention for the hydrocephalus should be designed with the consideration of facilitating the tumor removal and the specific approach selected.

The relationship between tumor removal and the beforehand relief of the hydrocephalus has been demonstrated by several authors. Konovalov et al. [9] found that total tumor removal could be achieved in 72 % of cases among shunt-free patients, whereas the rate was only 39 % in cases of previously shunted patients. They thereby concluded that collapsed ventricles may hamper radical tumor resection because of unsatisfactory exposure. Weshphal, in his comment on the study by Deshmukh et al. [46], also stressed the importance of controlling the ventricular size and advocated an ETV when the hydrocephalus needed to be relieved before the operation. As he stated, the ventricular system will remain dilated for a few days after this procedure and allows for easier manipulation of the tumor and protection of the posterior third ventricular wall. We agreed with this theory but preferred to place an EVD before surgery if needed. Compared with an ETV, an EVD would provide a more flexible ventricular volume by changing the drainage level or controlling the valve of EVD. Moreover, the hydrocephalus was resolved in the majority of cases (82.1 %) after radical tumor removal; thus, a second surgery for CSF diversion could be avoided.

Hydrocephalus is also an important factor hampering tumor exposure during the surgical procedure. For the supracerebellar infratentorial approach, rapid relaxation of the cerebellum can be achieved by opening the cisterna magna. In our experience, for most cases with moderate hydrocephalus when using the transtentorial approach, the occipital lobe can be relaxed by step-by-step dissection of the perimesencephalic cisterns and egression of the CSF. If it was still not sufficient or the patient presented with severe hydrocephalus, we advocated an intraoperative EVD to offer a size-controllable ventricle, which can definitely facilitate rapid brain relaxation during surgery and therefore minimize the risk of hemianopsia inherent to this approach.

Limitations of the study

This retrospective analysis has some inherent biases and drawbacks. Although this study is a large series, its retrospective character and the fact that the analyzed data derived from a single center limit the strength of the conclusions. Furthermore, because of the lack of statistical comparison among several surgical approaches, our selection of surgical approach was mainly established based on the surgeon's preference, personal experience, and existing literature. This series also represents the continuous evolution of indications and treatment strategies for various tumor types and reflects the lack of consensus regarding their management. Besides, the follow-up was still insufficient for some cases. Further prospective randomized controlled trails based on the surgical approaches, treatment strategies, and long-term follow-up will be more helpful to guide clinicians managing these patients.

Conclusions

Radical surgery is recommended as the optimal treatment for nongerminomatous pineal region tumors. Favorable results can be expected with modern microsurgical techniques and experienced neurosurgeons. An occipital transtentorial approach was indicated for most pineal region tumors, but the surgeon's preference and experience with each approach should also be considered in the selection of the surgical approach. Detailed knowledge about the arachnoid membranes spanning the pineal region may be helpful for total tumor removal and protection of the adjacent neurovascular structures. Hydrocephalus caused by tumors may be eliminated by radical tumor removal in the majority of the cases. In cases of unresolved hydrocephalus after surgery, an ETV procedure is preferred to the insertion of a shunt. **Acknowledgments** We are grateful to Xiaoyu Qiu and Yiping Mo (Medical Secretary, Department of Neurosurgery, Nanfang Hospital, Southern Medical University, Guangzhou, China) for their help with the data collection.

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Conflicts of interest None.

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