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Review Article

An overview of the current surgical options for pineal region tumors

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Abstract

Background: The list of pineal region tumors comprises an extensive array of pathological entities originating within one of the most complex areas of the intracranial cavity. With the exception of germ cell tumors, microsurgical excision is still nowadays the mainstay of management for most pineal region tumors.

Methods: A search of the medical literature was conducted for publications addressing surgical options for management of pineal region tumors.

Results: The infratentorial supracerebellar and the occipital transtentorial approaches are currently the most frequently used approaches for pineal region tumors. Endoscopic tumor biopsy with simultaneous endoscopic third ventriculostomy has emerged as a minimally invasive and highly effective strategy for initial management since it addresses the issue of tissue diagnosis and offers a solution for the associated hydrocephalus frequently encountered in these patients. Endoscope-assisted microsurgery and purely endoscopic excision have been reported in few reports and are likely to be more utilized in the future.

Conclusion: Preoperative planning is very crucial and should most importantly be individualized according to the anatomical features of the lesion and structures encountered during the procedure.

Key Words: Biopsy, endoscopic, infratentorial, pineal, transtentorial, ventriculostomy

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INTRODUCTION

The list of pineal region tumors comprises an extensive array of pathological entities originating within one of the most complex areas of the intracranial cavity.[12,30,32] The intricate arrangement of the anatomical structures therein makes surgical excision of these tumors always a challenging task. Microsurgical excision is still nowadays the mainstay of management for most pineal region tumors.[51] An exception is germ cell tumors in which treatment policies include radiotherapy and chemotherapy after obtaining a biopsy,[51] radiotherapy and chemotherapy without biopsy,[7] and radiation only without biopsy; a treatment strategy that is commonly utilized in Japan where the prevalence of germ cell tumors is known to be much higher than in Western countries.[23] Endoscopic tumor biopsy with simultaneous endoscopic third ventriculostomy (ETV) has emerged as a minimally invasive and highly effective strategy for initial management since it addresses the issue of tissue diagnosis and offers a solution for the associated hydrocephalus frequently encountered in these patients.[15,38,41,43,48,58,69]

In this article we present an overview of the contemporary surgical interventions for management of pineal region tumors. It is to be emphasized that this review is not intended to detail the surgical techniques as much as to elaborate on some aspects of these procedures, which may help selecting an appropriate surgical plan for a given lesion.

OPEN MICROSURGICAL EXCISION

The first successful removal of a pineal tumor was reported by Oppenheim and Krause in 1913 through an infratentorial supracerebellar corridor.[45] Subsequently, a variety of approaches to access lesions of the pineal region were devised and modified over the years. These approaches include the supracerebellar infratentorial,[28] occipital transtentorial,[49] combined supra-infratentorial transsinus,[4,59,72] posterior transcallosal interhemispheric,[13] and transcortical transventricular[66] approaches. The infratentorial supracerebellar and the occipital transtentorial approaches [Figure 1] are currently the most frequently used access ways to excise lesions of the pineal region.[51]

The occipital-transtentorial approach

The occipital-transtentorial approach was first described by Horrax in 1937[21] and later modified by Poppen[49,50] in the 1960s and by Jamieson in 1971.[22] Surgical positions used for the approach include sitting,[47] prone,[57] concorde,[34] and three-quarter prone[3,32] positions. The latter is preferred by some authors owing to the gravity-assisted occipital lobe retraction.[7,32]

Ausman *et al*. described a modified occipital-parietal, transtentorial approach to the pineal region in which surgery was performed with the head placed in the three-quarter prone position or at a 45° angle to the floor. The occipital portion of the skull is elevated and a bone flap is turned more easily than in approaches where the patient is positioned totally horizontally. The approach allowed excellent exposure of the pineal region and access to the midbrain, superior vermis, and third ventricle and also enabled access to the splenium of the corpus callosum and the right lateral ventricle in cases of arteriovenous malformations or thalamic tumors. They utilized the position in 13 cases and found that it combined the advantages of all the previously described operations to the pineal region without the disadvantages. The three-quarter prone approach had a reduced risk of air embolism compared with the seated position despite the slightly tilted up head. Importantly, occipital lobe retraction was greatly reduced as the occipital lobe sufficiently fell away from the operative field and required minimal retraction. No postoperative homonymous hemianopia could be found postoperatively in their cases. Furthermore, they constantly performed the approach through the nondominant side and therefore injury to both the dominant occipital lobe and splenium with total loss of visual and language functions was prevented.[3] Occipital lobe retraction has been linked to postoperative transient or permanent homonymous hemianopia[22,50,61] that is commonly encountered after an occipital transtentorial approach is performed in the semisitting position.[39] The subsequent utilization of the three-quarter prone and park bench positions with gravity-assisted retraction of the occipital lobe was noted to significantly decrease the incidence of postoperative visual field defects.[3,40]

The sitting position, in contrast, continuously keeps a clear surgical field, minimizes engorgement of the venous structures making them less likely to be injured during dissection and provides a gentle gravitational retraction of the tumor from the veins after opening the arachnoid.

The occipital transtentorial approach is performed through an occipital craniotomy that is carried out across superior sagittal sinus and torcula. The dura is open in C-shaped fashion[9] or as a pair of triangular leaves based on the superior sagittal and transverse sinuses.[32]

In the literature, the surgical corridor invariably described is between the occipital lobe and falx cerebri[32,57] due to absence of large veins entering the superior sagittal sinus for a distance of 4-5 cm proximal to the torcula, or directly medial to the posterior part of the occipital lobe.[54] However, we found that additional gentle elevation of the occipital lobe off the tentorial surface is always possible and safe; a finding that can be explained by the anatomy of the venous drainage of the inferior surface of the occipital lobe, which is drained by the occipitobasal vein that courses anterolaterally toward the preoccipital notch and frequently joins the posterior temporobasal vein before emptying into the lateral tentorial sinus.[54] The lateral tentorial sinuses usually drain into the transverse-sigmoid junction and anterior two-thirds of the transverse sinus on each side.[35] The occipital lobe is therefore anchored to the underlying tentorial surface at a relatively anterior and lateral point, leaving the posteromedial tentorial surface of the occipital lobe unattached. In our hands, occipital lobe elevation allows a wider angle for maneuverability in the lateral to medial direction and thus enables a better visualization of tumor parts extending contralaterally past the midline and overcomes the difficulty to reach parts of lesions extending to the opposite side, a feature that has been considered by some authors[57] to be one of the disadvantages of the approach.

The occipital transtentorial approach provides the widest view of both the supra- and infratentorial compartments and is preferred in exposing tumors with inferior extension into the cerebellomesencephalic cistern; a blind corner during the infratentorial supracerebellar approach, tumors with significant lateral or supratentorial extent, and in cases with low-lying torcula Herophili [Figure 2].[32] Splitting the tentorium offers a panoramic supra- and infratentorial view of the tumor, the surrounding deep venous structures and the collicular plate of the midbrain.[5,7,57] Additional incision or excision of small parts of the splenium of the corpus callosum has been described by some authors,[3,72] a step that we never felt necessary to gain access to the lesion.

The superior displacement of the deep venous system by the tumor is often considered an advantage of the infratentorial supracerebellar approach over the occipital transtentorial approach since in the former the tumor excision proceeds without the need to manipulate and cross the components of the galenic venous system [Figure 1].[5,7,57,63] One of the potential disadvantages of the occipital transtentorial approach is the oblique trajectory, which may at times be disorienting for surgeons not familiar with the approach.[32] The infratentorial supracerebellar approach, on the contrary, has the advantage of a strictly midline trajectory through which disorientation is less likely.[5]

The infratentorial supracerebellar approach

The infratentorial supracerebellar approach is a midline approach with a direct view of the tumor via an infero-superior corridor through which dissection proceeds without transgressing the Galenic system located superior to the tumor [Figures 1 and 2].[5,7,29,57,63,64] The sitting position is usually preferred for an infratentorial supracerebellar approach. The main advantages are gravity-assisted cerebellar retraction away from the tentorium and easier dissection of adherent veins off the tumor surface.[31,32] An additional advantage is decreased pooling of blood and cerebrospinal fluid (CSF) in the surgical field.[32] Avoidance of systemic complications associated with the sitting position like air embolism and intraoperative hypotension has made some surgeons prefer the Concorde position for the infratentorial supracerebellar approach,[26] as it combines elements of both the prone and sitting positions and allows access to the pineal region that is as good and straight as that provided by the sitting position with a reduced possibility of air embolism.[27]

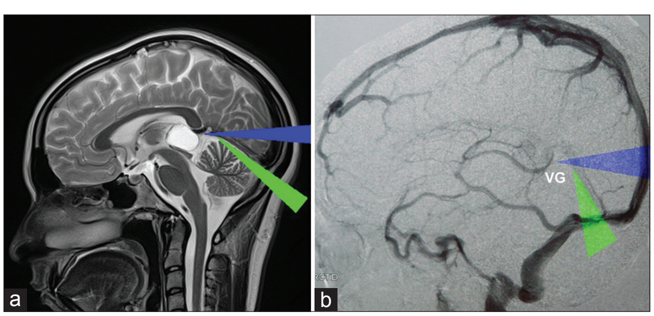

Figure 1: Corridors used during occipital transtentorial (Blue) and infratentorial supracerebellar (Green) approaches (a), and their relation to the Galenic venous system (b). VG: Vein of Galen

As the corridor between the cerebellum and inferior surface of the tentorium is developed, the precentral cerebellar vein and the superior vermian vein are identified and in most cases cut before the tumor is dissected. Rarely, the tectal veins or the superior and inferior quadrigeminal veins are very well developed hindering the approach to the tumor and requiring an oblique trajectory between them and the basal vein of Rosenthal.[57] Dividing the precentral cerebellar vein almost always takes place without sequlae.[10,63,68] Thrombosis of the basal veins of Rosenthal and internal cerebral veins with consequent fatal hemorrhagic infarction has, however, been reported after coagulating and dividing the precentral cerebellar vein at point near the confluence of the basal veins of Rosenthal.[24] The vein should be coagulated as far as possible from the confluence of the basal veins of Rosenthal to avoid progression of venous thrombosis to the collateral venous circulation.[27] The hemispheric bridging veins should be preserved.[46] Only thin bridging veins should be divided while saving the thick ones. In case some of the thick bridging veins strongly obstruct the surgical corridor, only median bridging veins should be sacrificed and paramedian veins are dissected for several millimeters from the cerebellar surface to gain adequate room between the tentorium and the cerebellum.[27]

The approach offers easy orientation,[7,20] but a narrow angle to the lateral and caudal side of the surgical field[27] and a restriction of the operative field in patients with a steep inclination of the tentorium.[20,27] Technical

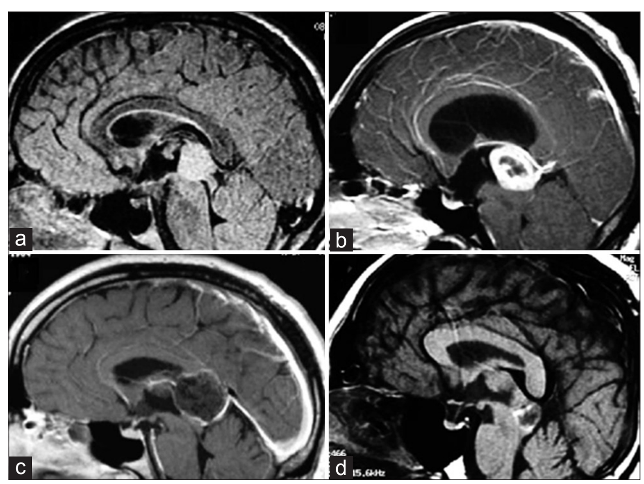

Figure 2: Selection of surgical approach to pineal region tumors. (a) Germinoma with dorsal displacement of internal cerebral veins. (b) Pineocytoma with dorsal displacement of internal cerebral veins. The two lesions are best approached via the infratentorial-supracerebellar or occipital-transtentorial corridor. (c) Epidermoid tumor with ventral displacement of the internal cerebral veins. This lesion is best approached via the posterior-interhemispheric corridor. (d) Tectal glioma with extension into the cerebellomesencephalic fissure. The occipital-transtentorial approach provides the best trajectory to the inferior pole of the tumor. (From Lozier and Bruce, 2003,[32] with permission)

nuances to maximize the exposure of the approach have been suggested. These include extending the craniotomy to the transverse-sigmoid junction on both sides with routine opening of the cisterna magna [Figure 3],[44] and applying stay sutures through the inferior leaf of the tentorium anterior to the transverse sinus with tension applied to the sutures by pulling in a cephalad direction, which enables a more direct operative trajectory with optimized illumination deeper in the field, improved working angles, and minimized need for retractors. It is important to confirm adequate blood flow within the dural sinuses using a microdoppler probe.[53]

Endoscopic management

Simultaneous endoscopic third ventriculostomy and tumor biopsy

The introduction of intraventricular neuroendoscopy represents a technically significant shift in the management of pineal region tumors.[38] Simultaneous ETV and biopsy for pineal region tumors was first described by Ellenbogen and Moores in 1997,[15] and has become the most favorable initial diagnostic and therapeutic methodology in many patients with tumors of the pineal region since they commonly present with noncommunicating hydrocephalus [Figures 4-6].[37,38] The procedure has the advantage of combining endoscopic biopsy with ETV to treat hydrocephalus and obtain CSF for tumor markers and cytology. Furthermore, obtaining tissue samples takes place under direct vision with inspection of the surrounding structures to identify malignant dissemination not visible on preoperative MRI.[2] The procedure has proven to be safe with high diagnostic yield reaching up to 100%.[17,18,41,43,48,69]

A single burr hole may be used and is placed 2-3 cm anterior to the standard Kocher's point so as to allow simultaneous tumor biopsy and ETV.[41,55,70] Alternatively, two burr holes are made and the ETV tumor biopsy are performed through separate burr holes [Figure 7].[11,48] A rigid endoscope is more commonly used.[2,11,25,55,56,67] Rigid endoscopes appear to improve the diagnostic yield

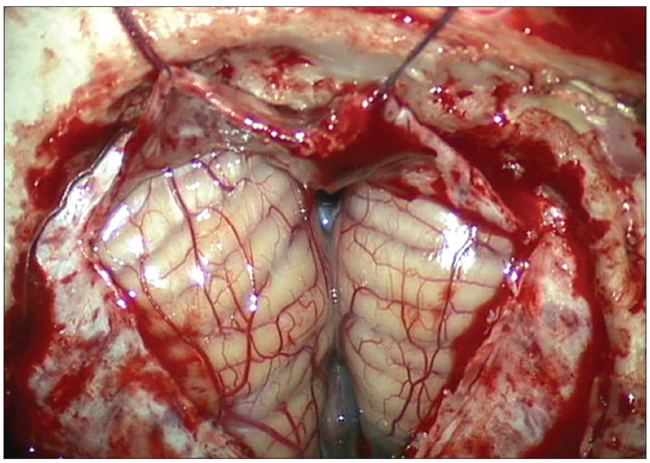

Figure 3: Extending the suboccipital craniotomy laterally to transverse–sigmoid junction on both sides with inferior extension down to the cisterna magna and subsequent CSF drainage to maximize the corridor of the infratentorial supracerebellar approach. (From Oliveira J, *et al*., 2013[44])

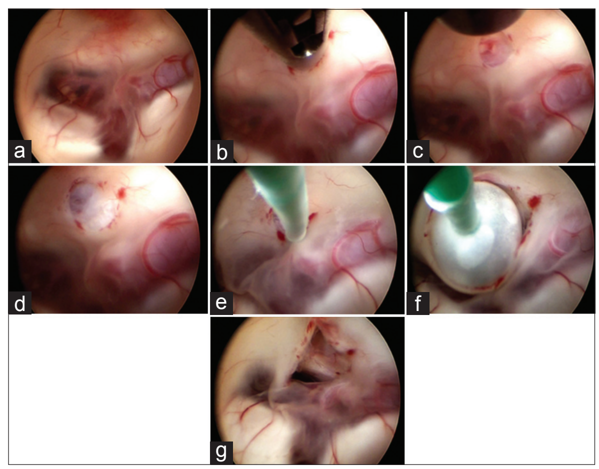

Figure 4: Serial images (a-g) during ETV in a patient with pineal region tumor. Note the very narrow prepontine distance and initial opening of the floor of the third ventricle against the dorsum sellae. Fogarty balloon catheter is then inserted through the initial puncture and then inflated to enlarge the stoma

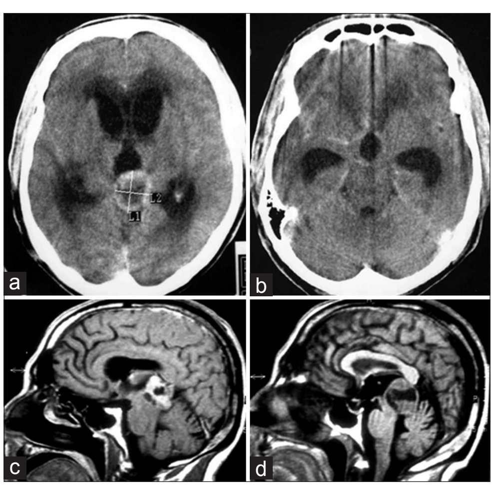

Figure 5: Preoperative CT scans of a Pinealoma (a and b). Post-ETV T1-weighted MR images (c and d)

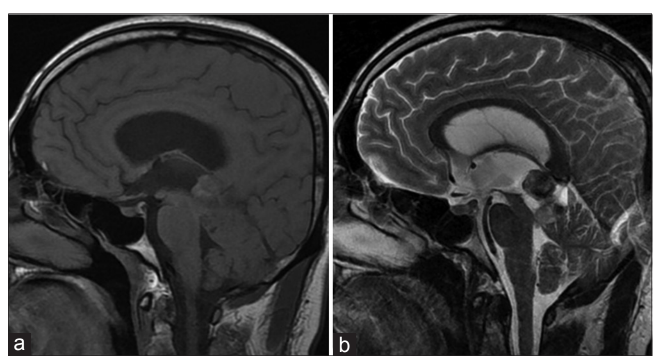

Figure 6: Midsagittal MRI images in a case with mixed germ cell tumor, noncontrast T1-weighted (a) and T2-weighted (b) after ETV. Note the opened floor of the third ventricle and the CSF flow void in through the stoma

as they permit the passage of larger-diameter biopsy forceps and consequently larger sample sizes. In addition, the higher image quality with better visualization allows various regions of the exposed tumor surface to be biopsied.[36] Using flexible endoscopes has been described to overcome the issue of suboptimal trajectories when a single entry site is used.[16-18,69] Compared with rigid endoscopes, the disadvantages of flexible endoscopes include smaller working channels and biopsy forceps, higher potential for disorientation, decreased image definition, and incompatibility with some navigational aids have limited their widespread appeal.[40] Utilization of both flexible and rigid endoscopes to perform the two parts of the procedure has also been described and is based on the rationale of making benefit from the advantages of each instrument.[48,69]

In their experience using rigid endoscopes, Morgenstern and Souweidane point out that the surgical approach should be individualized in each case according to the ventricular size, the relative position of the tumor, the dimensions of the massa intermedia, and the surgical goal [Figure 8]. If a single burr hole is used, it should be located midway between the optimal entry sites for either separate procedure. This strategy is best employed in cases characterized by a tumor that presents anterior to the massa intermedia, a small massa intermedia, a large degree of ventriculomegaly, or when the surgical goal is biopsy without consideration for total removal. A 30° angled lens is recommended to enhance the view off a linear axis. Alternatively, two sites of entry (one for the tumor biopsy and one for the ETV) is best suited in cases with tumors recessed behind the massa intermedia, a large massa intermedia, a moderate or minimal degree of ventriculomegaly, or when the tumor is amenable to total removal (2 cm or less).[40]

Although taking tumor biopsy before the third ventriculostomy has been suggested to minimize dissemination of the tumor cells,[66] most authors advocate taking biopsy after the ETV.[1,14,41,42] Performing the ETV before tumor biopsy prevents visual obscuration of the landmarks of the third ventricular floor by bleeding associated with tumor biopsy, which may risk abandoning the ETV.[36] The procedure starts with CSF collection for cytology and tumor markers, followed by third ventriculostomy and finally biopsy of the tumor is performed.[2]

For visualization of the tumor after ETV is completed, the 30° angled lens is rotated to achieve a posterior direction of view in cases where a single entry site is used. If a separate anterior entry for endoscopic biopsy is used, a 0° lens is used to visualize the posterior third ventricle. An eccentric tumor should be approached using a contralateral entry. Upon visualizing the tumor, cupped biopsy forceps are used to obtain tumor tissue samples from areas on the surface that most likely represent pathological tissue, are relatively avascular, and need the least torque.[40] Although some authors have described initial surface coagulation of the tumor before biopsy,[11] coagulation on the tumor surface should be avoided before sampling to minimize artifacts from cautery that may interfere with histopathological interpretation.[40] Varying degrees of hemorrhage invariably occur with cupped biopsy forceps, the majority of which will be controlled with continued irrigation, balloon tamponade, or electrocautery.[40]

Neuronavigational guidance enables preoperative planning of optimal entry sites and trajectories with a precise and real-time control of endoscope advancement during the procedure minimizing brain injury.[25,58] Stereotactic guidance has a special importance in cases without hydrocephalus to improve accuracy and minimize brain trauma.[62]

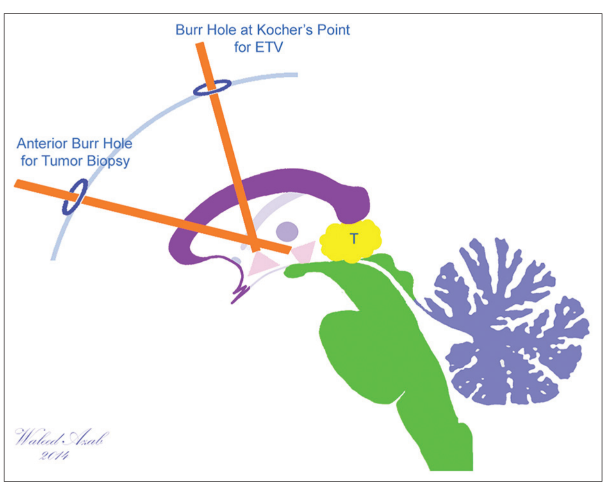

Figure 7: The standard burr holes used for ETV and for tumor biopsy

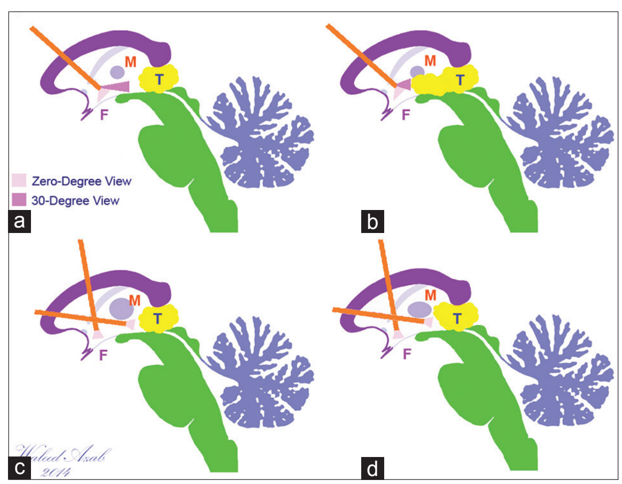

Figure 8: One and two burr hole strategies using a rigid endoscope for ETV and biopsy. Note that when one burr hole is used, the site of the single burr hole is chosen between the two standard burr holes. Small massa intermedia (a), Tumor presenting anterior to massa intermedia (b), Large massa intermedia (c), and Tumor recessed behind massa intermedia (d)

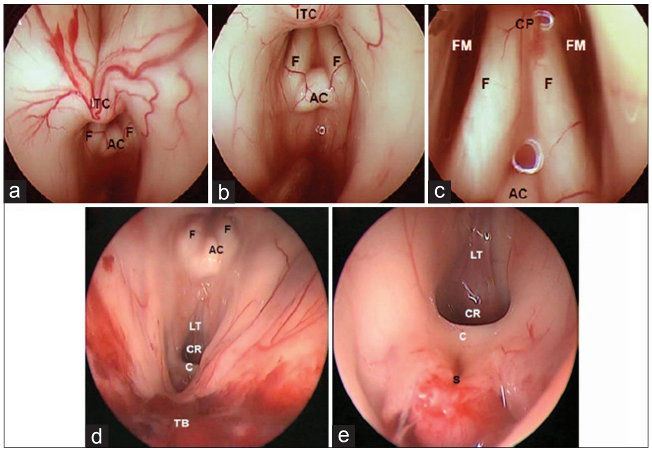

Figure 9: Intraoperative endoscopic views (a-e) during endoscope-assisted infratentorial supracerebellar approach. AC: Anterior commissure, C: Chiasm, CP: Choroid plexus, CR: Chiasmatic recess, F: Fornix, FM: Foramen Monro, ITC: Interthalamic commissure (collapsed), S: Stoma, TB: Tumor bed, LT: Lamina terminalis. (From Gu *et al*., 2013.[19] Pharma Professional Services)

It should, however, be emphasized that pineal region tumors in general and germ cell tumors in particular commonly display heterogeneity and mixed cell populations within the same tumor. This diversity makes it difficult for neuropathologists to appreciate the subtleties of histologic diagnosis when only small specimens are examined. The ability to obtain larger amounts of tissue and perform more extensive tissue sampling offered by open resection is a clear advantage over endoscopic biopsy.[7]

Endoscope-assisted microsurgery

In one cadaveric study, the endoscope-assisted infratentorial supracerebellar approach to the third ventricle was found to offer an unsurpassed view into the third ventricle from a posterior perspective.[8] Gu *et al.* successfully applied the technique in seven patients with pineal region tumors and concluded that the value of endoscope was to complement the microscope in the detection and removal of residual tumor and to remove blood clots from the tumor bed and aditus of the aqueduct, which may decrease the rate of postoperative obstructive hydrocephalus [Figure 9].[19] Broggi *et al.* used an endoscope-assisted interhemispheric transtentorial retrosplenial approach for excision of 15 pineocytomas. The endoscope enabled detection of residual tumor located either behind the vein of Galen or attached to the undersurface of the corpus callosum in six cases and guided the residual tumor resection.[6]

Purely endoscopic infratentorial supracerebellar approach to the pineal region has also been described [Figure 10].[60,65] Uschold *et al.* reported using the endoscope for excision of pineal cysts and selected solid tumors of the pineal region. Tumors with high vascularity, infiltrative nature or those with significant extent across the midline bilaterally or above the tentorium were considered unsuitable candidates that should rather be excised via open microsurgical procedures. Endoscopic infratentorial supracerebellar approach is performed in the sitting position via a paramedian corridor to avoid obstruction by the vermis. The intracranial entry point is either a 1.5- to 2.5-cm burr hole or a microcraniectomy placed at the inferior margin of the transverse sinus and 1-2 cm lateral to the torcula. After opening the dura the endoscope is inserted and microsurgical instruments are used for microdissection under endoscopic vision. Zero, 30, and 45°C

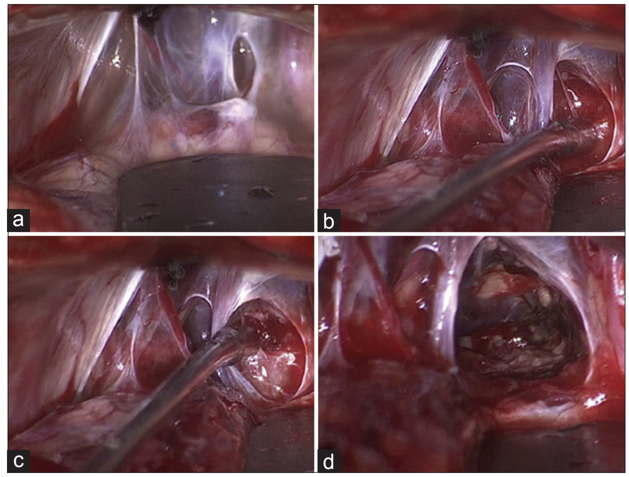

Figure 10: Intraoperative views using the VITOM® system (Karl Storz GmbH & Co., Tuttlingen, Germany) to resect a pineal region tumor: (a) initial exposure of prepineal arachnoid; (b) initial exposure of tumor; (c) initial resection; (d) end of resection with residual tumor attached to brainstem. (From Mamelak *et al*., 2012,[33] with permission)

Stereotactic biopsy

Stereotactic biopsy is inherently associated with a limited amount of tissue obtained leading to difficulties of histopathological diagnosis. The high frequency of mixed tumors in this region further contributes to diagnostic inaccuracies of the procedure.[17] With the Galenic venous system present in the close proximity of the pineal region, preoperative stereotactic angiography has been suggested as a prerequisite for a safe stereotactic biopsy of pineal region lesions.[52] A recent review of all major series reporting stereotactic biopsy for pineal region lesions revealed a mean diagnostic yield of 94%, with a morbidity of 1.3% and a mortality of 8.1%.[71] The procedure is not commonly used nowadays and has been largely replaced by endoscopic biopsy.

CONCLUSION

Surgical management of pineal region tumors comprises various microsurgical and endoscopic options. Preoperative planning is very crucial and should most importantly be individualized according to the anatomical features of the lesion and structures encountered during the procedure.

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