Pineal Region Tumors – Neurosurgical Review

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REVIEW

SUMMARY

The treatment for the pineal region tumors depends on tumor histology. Nowadays, germinomas can be cured by radiotherapy and chemotherapy without surgical resection but the other pineal region tumors should be primary treated by surgery. Two microsurgical approaches, the infratentorial supracerebellar and the occipital transtentorial, are accepted as the main standard accesses to the pineal region. For benign pineal tumors (pineocytoma, meningioma, mature teratomas, symptomatic pineal cysts, etc.) radical surgical resection can be curative. For malignant tumors radical surgical resection is not an objective. Serum and CSF markers contribute to the diagnosis of pineal parenchymal tumors. b-HCG is mainly positive in choriocarcinomas, embryonal carcinomas and mixed germ cell tumors and AFP is expressed by yolk sac tumors, embryonic carcinomas, immature teratomas and mixed germ cell tumors. b-HCG is usually low in germinomas which are often positive for PLAP on immunohistochemistry. Fifty-one pineal region tumors were surgically treated by senior author (NdT). Only 17 of them were the neoplasms originating from pineal body (pineal tumors). In conclusion it can be stressed that management of pineal tumors requires a multidisciplinary cooperation. With the exception of germinoma where only a biopsy is needed, the role of the surgeons still remains prominent as resection of pineal tumors requires high technical skill and experience as well as precise clinical judgment.

Keywords: neurosurgery, pineal region tumors

1. INTRODUCTION

Pineal region tumors include a variety of neoplasms of different histological origin growing from the pineal gland itself or from structures of the parapineal space. These tumors are rare and account for 0.4 to 1.0 % of intracranial tumors in adults and 3-8 % in children . The most common types are germ cell tumors, pineal parenchymal cell tumors and glial cell tumor. Other pineal region tumors such as meningiomas, PNET, neurocytomas, hemangioblastomas, cavernomas and metastasis are infrequent.

The treatment options for the different pineal region tumors vary according to their histological nature. However, with the exception of germinomas which can be nowadays cured by low-dose radiotherapy and chemotherapy and only require a biopsy for diagnosis, surgery still plays a central role in the management of most of the other pineal region tumors followed or not by adjuvant radiotherapy, chemotherapy or a combination of both. The first successful removal of a pineal tumor was reported in 1913 by Oppenheim and Krause. Krause was the first to describe and successfully use the infratentorial supracerebellar approach in three cases in 1926(1)..In the microsurgical era, Stein further developed and popularized this approach during the 1970's (2). Finally the right suboccipital approach was described by Poppen and further modified by Jamieson in 1971(3)..

The infratentorial supracerebellar and the occipital transtentorial approaches are nowadays accepted as the main standard accesses to the pineal region.

2. MICROSURGICAL ANATOMY

Pineal region tumors lie deep in the center of the cranium and are surrounded by critical anatomical structures that have to be respected at all costs. Therefore, a precise knowledge of the complex anatomy of the pineal region is of paramount importance (4, 5, 6). The pineal gland is located on the midline and forms an appendix of the caudal end of the diencephalons embracing the pineal recess of IIId ventricule. The pineal stem is continuous with the habenular commissure dorsally and the posterior commissure ventrally. The pineal body projects posteriorly in the quadrigeminal cistern where it is flanked by the splenium of the corpus callosum superiorly and lies on the tectal quadrigeminal plate in-between the left and right superior colliculi. The pineal gland is mainly vascularized by the medial and lateral posterior choroidal arteries. The medial posterior choroidal arteries are branches of the posterior cerebral artery and in addition to the pineal body they supply the superior and inferior colliculi, and the choroidal plexus of the third ventricule. These arteries are displaced laterally by pineal tumors in the cistern and rostrally in the posterior part of the third ventricule together with internal cerebral veins. The posterior lateral choroidal artery supplies the pulvinar and is generally displaced laterally by pineal tumors. Other important arterial landmarks are the superior cerebellar arteries that can be displaced inferiorly by pineal tumors and the medial occipital artery branching from the posterior cerebral artery and giving the calcarine artery. During surgical approaches to the pineal gland, the major anatomical obstacle is the Galenic venous system. The vein of Galen has several tributaries: the superior vermian vein and the precentral cerebral vein run in the midline and into the dorsocaudal part of the great vein. The internal cerebral veins and the pineal veins join ventrally. In pineal tumors, the posterior portion of the internal cerebral veins is always elevated rostrally, and the veins are occasionally separated from each other. On the lateral aspect of the great vein, the medial occipital veins, the third segment of the basal veins of Rosenthal, and the posterior mesencephalic veins join. The pineal veins are the draining veins of pineal tumors and drain into either the posterior portion of the internal cerebral veins or the vein of Galen. At this point pineal tumors are thightly adherent to the internal cerebral vein and/or the vein of Galen. An injury to the basal veins or the internal cerebral veins will yield major complications. And a transection of a major medial occipital vein may cause homonymous hemianopsia or visual seizures.

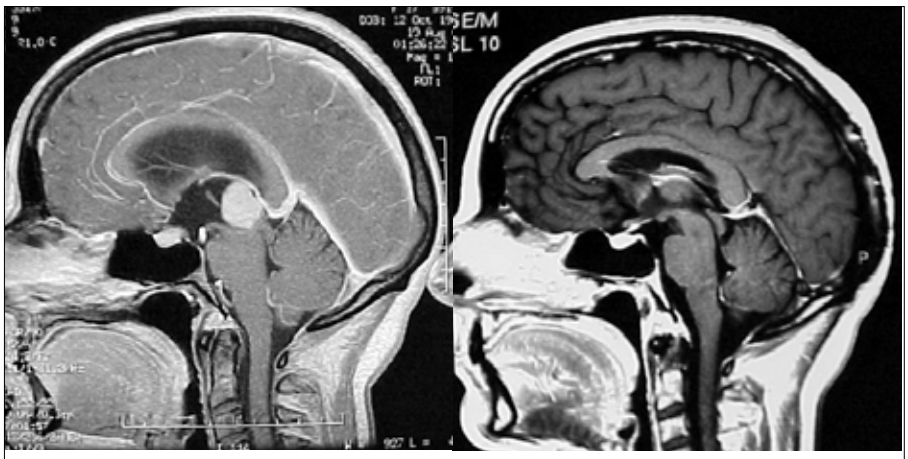
3. RATIONALE

For benign pineal tumors (pineocytoma, meningioma, neurocytomas, mature teratomas, hemangioblastomas, cavernous hemangiomas, gangliogliomas, and symptomatic pineal cysts) total surgical resection is a primary goal as surgery alone can be curative (7, 6).. For malignant tumors surgery is only a part of the treatment which will consist of adjuvant therapies and therefore radical surgical resection is not an objective (6, 8)..In all cases focus should be given to reduce post treatment morbidity.

4. DECISION MAKING

a) Diagnosis

Clinical presentation: Symptomatic



1a. preoperative MRI 1b. postoperative MRI

FIGURE 1A and 1B. Sagittal MRI section (T1+gadolinium) of a pineal lesion removed by NdT through occipital transtentorial approach. Note the steep angle of the straight sinus and the position of the lesion anteriorly to the quadrigeminal plate making a supracerebellar infratentorial approach difficult.

hydrocephalus or occulomotor signs are generally the first clinical manifestation of pineal region tumors. Hydrocephalus is triventricular by compression of the aqueduct of Sylvius and can be acute or chronic. Symptoms include headaches, gait problems and occulomotor signs such as Parinaud syndrome. In slow growing tumors, chronic hydrocephalus may develop and cause dementia. Occulomotor signs can also occur through direct compression of the superior colliculi or the posterior commisure (6)..

Radiology: the radiological exam of choice is MRI which will reveal the tumor and its relations to adjacent anatomical structures. Particular attention has to be given to T1+gadolinium sequences, high resolution T2 squences for surrounding vessels (flow void) and cranial nerves, phlebo-MRI sequences for assessing the 3D anatomy of the deep venous system and its relation with the tumor. A CT is also useful to detect intratumoral calcifications or hemorrhage.

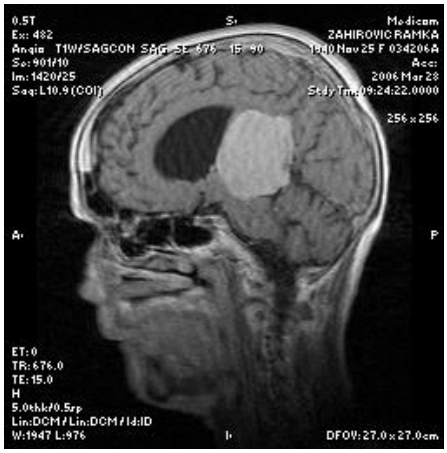


FIGURE 3. Giant pineal region meningioma removed by co-author (KD) using the combined Sekhar's approach

Biopsy: Histological diagnosis is obtained either by stereotactic or endoscopic transventricular biopsy or directly during open surgery. For large pineal tumors a stereotactic biopsy is a safe initial procedure to obtain diagnosis. For tumors extending into the posterior part of the third ventricule, endoscopic transventricular biopsy allows access to tumor tissue as well as third ventriculostomy to treat hydrocephalus (6)..

b) Indications for surgery and microsurgical approaches

If a newly diagnosed pineal mass is accessible by stereotactic or endoscopic biopsy and the cranial MRI is compatible with a germinoma, a biopsy should first be done in order to avoid an unnecessary craniotomy in that case. If the radiological examination is compatible with an asymptomatic benign pineal cyst and the serum and CSF markers are negative, the patient can be followed up without treatment. The treatment of other pineal tumors requires surgery but the choice of radical or conservative resection will depend on the diagnosis of the pre-surgical biopsy or the intraoperative frozen section. Benign tumors such as mature teratomas, pineocytomas or meningiomas require radical surgical resection when feasible without compromising surrounding neurovascular structures. More aggressive tumors, such as malignant teratomas, pinealoblastomas, embryonal carcinomas, choroicarcinomas and yolk sac tumors require a combination of surgery, radiation therapy and

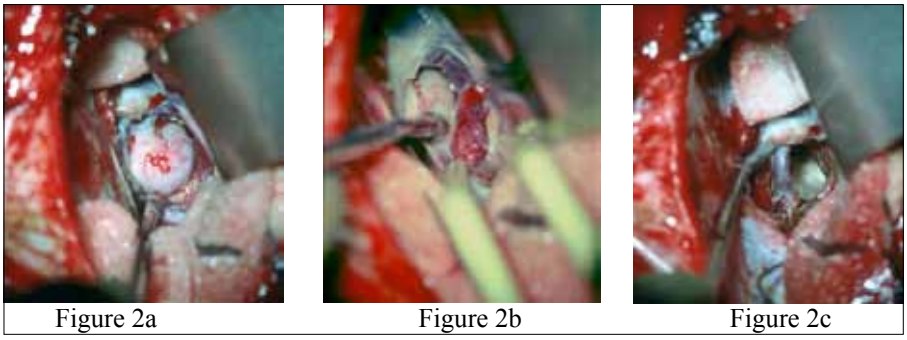


Figure 2a Figure 2b Figure 2c

FIGURE 2A, 2B and 2C. Surgical presentation of a right occipital transtentorial approach include: a) After dissecting the arachnoid, the tumor comes into full view. b) Resection of the tumor with dissection of the capsule from surrounding structures. c) Final view of the operative field after resection of the tumor, under the corpus callosum the posterior part of the third ventricule is visible. The great vein of Galen and the left basal vein are also visible.

Even if the many different pineal tumor types may have a preferential appearance on cranial imaging, no such characteristics are specific for one or another tumor type and do not preclude obtaining tissue for histological examination. An exception are benign pineal cysts which have a homogenous cyst content with a thin enhancing rim and have no or only mild mass effect on surrounding structures. Except for pineal region meningiomas or falcotentorial notch meningioma extending in the pineal region, angiography is usually not necessary(6)..

Markers: Serum and CSF markers contribute to the diagosis of pineal parenchymal tumors and assessment of their malignancy. BHCG and α-foetoprotein are found in germ cell tumors. β–HCG is mainly positive in choriocarcinomas, embryonal carcinomas and mixed germ cell tumors and AFP is expressed by yolk sac tumors (high levels), embryonic carcinomas, immature teratomas and mixed germ cell tumors. β-HCG is usually low in germinomas which are often positive for PLAP on immunohistochemistry (8)..

chemotherapy. In any case the prime goal of surgery should be avoiding surgical morbidity even at the cost of a less radical surgical resection. The choice of approach is a matter of evaluating the anatomical relation of the tumor with the surrounding structures. A steep angle of the straight sinus makes the infratentorial supracerebellar approach difficult as an extensive retraction of the cerebellum is required to visualize and reach the pineal area. Moreover, in that case the lateral exposure of the surgical field is restricted and renders the resection of larger tumors more complicated. Evaluating the relationship of the tumor with the quadrigeminal plate is also important. For smaller midline tumors located in the posterior part of the third ventricle and displacing the quadrigeminal plate and the tegmentum of the midbrain caudally, the infratentorial supracerebellar approach is favored as it allows simple, direct and symmetrical exposure of the walls of the third ventricle and internal cerebral veins on both sides. In the case the tumor lies more caudally and extends in the upper portion of the aqueduct of sylvius, lying therefore cranially of the tectum, the infratentorial approach is inappropriate as the quadrigeminal plate obstructs the surgical exposure. Finally, the occipital transtentorial approach is preferred as well in big tumors with lateral extension in the pulvinar thalami as it gives a better lateral exposure of the walls of the third ventricle (6)..

The giant tumors of the pineal region can be removed by the combined occipital , transtentorial, supracerellar transsinus approach described by Sekhar(10) (Figure 3).

a) Pineal gland tumors

Germinoma	Excluded
Mature teratoma	3
Immature teratoma	4
Embryonal carcinoma	2
Pineocytoma	3
Intermediate differentiation	1
Pineoblastoma	2
Yolk sac tumor	2
NUMBER	17
Radical resection	11
Subtotal resection	2
Biopsy	4 (2 embryonal carcinoms and 2 yolk sac tumors)

c) Complications

Type of complications	Number of complication	Reason of complications
Hemianopsia	1	Occipital lobe retraction
Visual seizures	1	Occipital lobe retraction
Metamorphopsia	1	Venous infarction
Parinaud syndrome (permanent)	1	quadrigeminal plate manipulation
IV CN palsy	2	Nerv traction
Air embolism	0	0

b) Pineal region tumors without pineal gland tumors

cavernous angiomas	4
PNETs	2
Ependymomas	4
Astrocytomas grade II	5
Hemangioblastomas	4
Gangliogliomas	2
Meningiomas	9
plexus papillomas	2
Neurocytoma	1
Neurenteric cyst	1
NUMBER	34
Radical resection	29
Subtotal resection	5 (2 Astrocytomas and 3 meningiomas)

5. RESULTS

The fifty-one pineal region tumors were surgically treated by senior author (NdT). Only 17 of them were the neoplasms originating from pineal body (pineal tumors)

6. CONCLUSION

Contemporary management of pineal tumors requires a multidisciplinary cooperation where surgery represents only one aspect of the treatment plan. However, with the exception of germinoma where only a biopsy is needed, the role of the surgeons still remains prominent as resection of pineal tumors requires high technical skill and experience as well as precise clinical judgment. The infratentorial supracerebellar approach and the occipital transtentorial approach when used appropriately allow access to nearly every type of pineal neoplasms.

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