Pineal region tumor treatment

- Transient cortical blindness following occipital lobe retraction in a pineal region meningioma resection
- A Rare Presentation of Ruptured Pineal Region Teratoma with Postoperative Aseptic Meningitis
- Melatonin Prevents Tumor Growth: The Role of Genes Controlling the Circadian Clock, the Cell Cycle, and Angiogenesis
- Pituitary and pineal gland metastases from pulmonary neuroendocrine carcinoma: A case report and literature review
- Treatment of a Large Hemorrhagic Midbrain Cavernoma Within the Silvian Aqueduct in a Five-Year-Old Girl-A Case Report
- Machine learning decision support model construction for craniotomy approach of pineal region tumors based on MRI images
- Stroke-like migraine attack after radiation therapy (SMART) syndrome in a pediatric patient with a ventriculoperitoneal shunt: illustrative case
- Lu-177 DOTATATE Therapy in a Treatment Naive Metastatic NET Pineal Gland With Drop Metastasis to Spinal Cord: A Great Leap Forward in Oncology

---

The optimal management strategy for pineal region tumors has yet to be determined.

“Test dose” radiation

“Test dose” radiation: The practice of this is declining. The logic behind the test dose was that if a pineal region tumor enhanced uniformly and had the classic appearance of a pineal germinoma on MRI, a test dose of 5 Gy was given, and if the tumor shrank then the diagnosis of germinoma was virtually certain and XRT was continued without surgery.

This would needlessly expose a patient with benign or radioresistant tumors to the test dose of XRT. “Test dose XRT” should especially be avoided in tumors suspected of being teratomas or epidermoid cyst on MRI, and the response may be misleading in the relatively common situation of tumors with mixed cell types. This practice is that giving way to the paradigm of surgery to ascertain histology in almost all cases (e.g. via endoscopic surgery) because of the harmful effects of XRT and because 36–50% of pineal tumors are benign or radioresistant.

Management suggestions

1. perform MRI of cervical, thoracic, and lumbar spine to assess for drop mets

2. send for GCT markers (β-hCG, AFP, PLAP). Somewhat helpful, but not adequate for diagnosis: if negative for GCT markers, it may be a pineal cell tumor, or it may be a GCT without markers, see Tumor markers; if positive, it can still be a mixed cell-type tumor:

a) serum

b) CSF(if able to safely obtain; LP is contraindicated with large intracranial mass and/or obstructive hydrocephalus; CSF may be obtained from EVD if placed)

3. obtain histology in most cases. Most often this involves a biopsy, which should be generous (to avoid missing other histologies in mixed cell tumors)

a) if hydrocephalus:transventricular biopsy

b) if no hydrocephalus:

● open biopsy or

● stereotactic biopsy or

● Computer-assisted cisternal endoscopic approach (CACE): Employs a supracerebellar infratentorial approach that permits visualization of neurovascular structures and avoids traversing brain parenchyma [1].

4. based on markers and histology:

a) germinoma:XRT+chemo

b) all the tumors:one option is resection followed by adjuvant therapy(usually not very helpful)

Hydrocephalus: Patients presenting acutely due to hydrocephalus may be best treated with external ventricular drainage (EVD). This permits control over the amount of CSF drained, prevents peritoneal seeding with tumor (a rare event), and may avoid having a permanent shunt placed in the significant number of patients who will not need one after tumor removal (although ≈ 90% of patients with a pineal GCT require a shunt). Ventricular access, via EVD or Frazier burr hole, in the post-op period, is important in the event of acute hydrocephalus.

The limited access and anatomical complexity have generated a spectrum of anatomical approaches and raised the interest for neuroendoscopic approaches. Equally complex is the spectrum of treatment modalities such as microsurgery as the main option but stereotactic radiosurgery as an alternative or adjuvant to surgery for selected cases, radiation as for germinoma and or combinatorial chemotherapy, which may need to precede any other ablative technique as constituents.

A standard treatment strategy for these tumors has not yet been established. Various treatments from surgery or radiotherapy and chemotherapy alone or in combination have been applied.

Empiric treatment of pineal region tumours with fractionated radiation therapy is no longer warranted. Image-guided stereotactic technology provides a safe method to accurately diagnose and effectively treat selected pineal region masses. After definitive histologic diagnosis is established, proper treatment may be instituted. Options for treatment include stereotactic radiosurgery for selected tumours and vascular malformations, microsurgical resection of benign tumours or fractionated external beam radiation therapy for malignant germ cell and glial tumours. Stereotactic surgery should be the first option in the diagnosis and therapy of pineal region masses [2].

The therapeutic modalities used for tumors of the pineal region in Western countries differ from those in Japan, mainly because of the different patient populations. An extensive survey was conducted to

delineate the racial differences in Japan and in Korea in the epidemiology and recent therapeutic modalities for this tumor group. Among the members of International Society for Pediatric Neurosurgery (ISPN), 15 from Japan and 5 from Korea reported their recent findings in 118 (1-25 years of age, mean 7.38-year period) and 125 (1-12 years of age, mean 6.69-year period) histologically verified cases, respectively. The patient populations in the two countries were found to be almost identical, with an extremely high incidence of germ cell tumors representing 71.2% (in Japan) and 80.0% (in Korea) of all pineal region tumors and neuroectodermal tumors representing only 15.2% and 16.8%, respectively. The most common type of pineal region tumor was germinoma (46.6% in Japan and 47.2% in Korea). The majority of tumors were radio- and/or chemosensitive, and adjuvant therapy rather than extensive surgery played the major role in the treatment in both countries. Radical resection of the tumor was recommended as the initial procedure by only 22.2% of neurosurgeons in Japan and 16.6% in Korea. Biopsy was recommended by 38.9% and 50.0%, and radiation therapy by 38.9% and 37.5%, respectively. A minimally invasive procedure, by either a neuroendoscopic (33.3% of biopsies in Japan) or a stereotactic approach (33.3% of biopsies in Korea), was considered to be appropriate as the initial procedure. The study disclosed the almost identical epidemiology of this brain tumor in Japan and in Korea and clarified the consequent therapeutic modalities. Oi et al. emphasize that minimally invasive tissue diagnosis with or without tumor debulking should be considered as the initial step for the treatment planning of the pineal region tumor, followed by the most commonly indicated major procedures, including radiation therapy, chemotherapy and/or radical resection with various methodologies [3)].

Endoscopic Third Ventriculostomy and Tumor Biopsy for Pineal region tumor

Endoscopic Third Ventriculostomy and Tumor Biopsy for Pineal region tumor.

Pineal region tumor surgery

see Pineal region tumor surgery.

Stereotactic surgery

Stereotactic surgical procedures: May be used to ascertaining diagnosis (biopsy), or to treat symptomatic pineal region cysts.

Caution is advised since the pineal region has numerous vessels (vein of Galen, basal veins of Rosenthal, internal cerebral veins, posterior medial choroidal artery) which may be displaced from their normal position. The complication rate of stereotactic biopsy is ≈ 1.3% mortality, ≈ 7% morbidity, and 1 case of seeding in 370 patients, and the diagnostic rate is ≈ 94%.

A shortcoming of stereotactic biopsy is that it may fail to disclose the histologic heterogeneity of some tumors.

Two main stereotactic trajectories: 1) anterolateral (low frontal) approach below the internal cerebral veins, and 2) posterolateral trans-parieto-occipital.

One study found that the trajectory correlated with complications, and they recommended the anterolateral approach. However, the correlation of trajectory and complications was not born out in another study, and they found that the complication rate was higher in firm tumors (pineocytomas, teratomas, and astrocytomas) and they recommend an open approach when the tumor appears difficult to penetrate on the first attempt at biopsy.

Stern and Ross report the results of the stereotactic aspiration of benign pineal region cysts in two patients with symptomatic ventriculomegaly secondary to obstruction of the aqueduct. Stereotactic aspiration produced several cubic centimeters of brown-to-yellow fluid that was negative for a tumor on cytological examination. Ventriculomegaly and symptoms were relieved without complications by the procedure in one patient who remains asymptomatic after 30 months of follow-up. In the second patient, persistent ventriculomegaly demonstrated on computed tomography resulted in the placement of a ventriculoperitoneal shunt; however, this was removed several months later during an episode of appendicitis and the patient remained asymptomatic without the shunt. The symptoms and ventriculomegaly recurred 71 months post aspiration, and the cyst was reaspirated. Ten months later, she remains asymptomatic. Options for managing such patients include open resection, shunting, and stereotactic aspiration [4].

This procedure permitted aspiration of the cyst content, normalization of aqueductal stenosis, and a differential histological diagnosis. Postoperative computed tomographic scan examination showed a stable reduction of the cystic volume and a complete clearance of the hydrocephalus. At present, the two patients are clinically normal and are leading a productive life (follow-up, 42 and 40 mo). It is important that this kind of nonneoplastic lesion be identified, in order to avoid confusion with pineal tumors and possibly inappropriate treatment. Pineal cysts rarely become symptomatic. Up to the present, only 27 symptomatic cases have been described in the literature. Of those, signs of intracystic bleeding were found in 17 of the 21 cases in which the cyst contents were known. Moreover, in our own two cases, numerous hemosiderin-laden macrophages were present in the cystic fluid. We think, therefore, that intracystic bleeding has been the determining factor for the occurrence of symptoms in most of these cases. The authors underline the advantages of this type of stereotactic management, which can provide both diagnosis and treatment and thus avoid the possible risks of surgical excision and/or of radiotherapy [5].

Computer-assisted cisternal endoscopic approach

Computer-assisted cisternal endoscopic approach (CACE): Employs a supracerebellar infratentorial approach that permits visualization of neurovascular structures and avoids traversing brain parenchyma.

Radiotheraphy

Radiation treatment: Germinomas are very sensitive to radiation (and chemotherapy), and are probably best treated with these modalities and followed.

XRT is also utilized post-op for other malignant tumors. For highly malignant tumors or if there is evidence of CSF seeding, craniospinal XRT with a boost to the tumor bed is appropriate.

If possible, XRT is best avoided in the young child. Chemotherapy may be used for age < 3 yrs until the child is older when XRT is better tolerated.

Stereotactic radiosurgery

Stereotactic radiosurgery may be appropriate for treatment of some lesions.

Gamma Knife Radiosurgery for Pineal Region Tumor

Gamma Knife Radiosurgery for Pineal Region Tumor.

References

1)
Youssef AS, Keller JT, van Loveren HR. Novel applica- tion of computer-assisted cisternal endoscopy for the biopsy of pineal region tumors: cadaveric study. Acta Neurochir (Wien). 2007; 149:399–406
2)
Dempsey PK, Kondziolka D, Lunsford LD. Stereotactic diagnosis and treatment of pineal region tumours and vascular malformations. Acta Neurochir (Wien). 1992;116(1):14-22. PubMed PMID: 1615765.
3)
Oi S, Matsuzawa K, Choi JU, Kim DS, Kang JK, Cho BK. Identical characteristics of the patient populations with pineal region tumors in Japan and in Korea and therapeutic modalities. Childs Nerv Syst. 1998 Jan-Feb;14(1-2):36-40. PubMed PMID: 9548339.
4)
Stern JD, Ross DA. Stereotactic management of benign pineal region cysts: report of two cases. Neurosurgery. 1993 Feb;32(2):310-4; discussion 314. PubMed PMID: 8437672.
5)
Musolino A, Cambria S, Rizzo G, Cambria M. Symptomatic cysts of the pineal gland: stereotactic diagnosis and treatment of two cases and review of the literature. Neurosurgery. 1993 Feb;32(2):315-20; discussion 320-1. Review. PubMed PMID: 8437673.