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Surgical options for pineal region tumors:

Perspectives on: Tseng K-Y, Ma H-I, Liu W-H, Tang C-T. Endoscopic supracerebellar infratentorial retropineal approach for tumor resection-A technical report. World Neurosurgery 2011

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Pineal region tumors encompass a diverse array of histologies that include germinomas, malignant germ cell tumors and teratomas, pineal parenchymal tumors, gliomas, and meningiomas, among others. Apart from teratomas, which have a characteristic appearance, the histological identity of a pineal region tumor can be difficult to predict based on imaging alone. In the past, patients with pineal tumors that were thought to potentially be germinomas were often given a “test dose” of irradiation, and if a complete response was achieved, they were treated empirically with further irradiation, reserving biopsy for non-responding lesions. Because tumor histologies other than germinomas may respond initially to irradiation, but warrant different management strategies, the use of empiric irradiation is no longer recommended. At present, appropriate diagnostic evaluation incorporates a combination of blood and cerebrospinal fluid (CSF) marker analysis for α -fetoprotein (AFP), indicative of yolk sac components (endodermal sinus tumor, teratoma with endodermal sinus elements, and occasionally embryonal carcinoma) and β -human chorionic gonadotropin (HCG), which is commonly expressed by tumors with trophoblastic components, such as choriocarcinomas, malignant teratomas and embryonal carcinomas (6). Expression of high levels of either AFP or HCG is diagnostic of a non-germinomatous germ cell tumor (NGGCT) and histological confirmation is not mandated. Low-level expression of HCG (less than 50 to 100 mIU/L) may be observed in germinomas with syncytiotrophoblastic cells or NGGCTs and, thus, has less specificity in distinguishing these entities, and the decision regarding whether such patients can be treated initially as germinomas is more controversial. In contrast to the “secreting” tumors, histological examination of a biopsy sample is warranted for those cases lacking significant marker elevation (4–6). In addition to marker analysis, staging by CSF cytology and magnetic resonance imaging of the spine is also an important component of the diagnostic evaluation

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for both germ cell tumors and pineal parenchymal tumors, in view of their risk for dissemination within the neuraxis (6).

The role of surgery in the management of pineal region tumors is influenced by the histological diagnosis and tumor growth characteristics. Tumors of the pineal region typically present with obstructive hydrocephalus from occlusion of the aqueduct, which often requires CSF diversion as an initial management step. Historically, this was commonly accomplished by ventriculoperitoneal shunt insertion, although in recent years endoscopic third ventriculostomy has emerged as a favored option for CSF diversion for patients without significant tumor involvement of the third ventricular floor.

A second element in surgical management relates to establishing a tissue diagnosis in cases without marker elevation. Tissue acquisition can be accomplished by stereotactic or endoscopic techniques, although because of the risk of hemorrhage associated with these approaches, some neurosurgeons prefer to perform an open biopsy (8). This intervention may be combined with surgical cytoreduction, which may have therapeutic benefit in selected situations. One such scenario involves those cases where the tumor has a histology that is likely to respond poorly to chemotherapy or irradiation, such as a benign teratoma, in which surgical removal is required as definitive therapy.

Because germinomas respond rapidly to radiation and chemotherapy and carry a greater than 90% ten-year survival with adjuvant therapy, there is usually no rationale for surgical cytoreduction (7). The situation is more controversial for NGGCTs, which may incorporate treatment-resistant teratoma as well as germinomatous and non-germinomatous components. The management strategies that have been used in recent years for these two subgroups of tumors have focused on reducing the morbidity of treatment and long-term sequelae for patients with germinomas while maintaining excellent survival rates, and enhancing long-term disease control in those with NGGCTs (1,4,6). For the latter group, most recent cooperative group protocols for children have favored proceeding with adjuvant therapy after diagnosis, in an effort to eradicate the malignant components of the tumor, with the understanding that in some instances, a teratomatous component will remain and require “second-look” surgical intervention to biopsy and resect this residual tissue (4,6,10). It is unclear whether this staged approach to surgery improves survival, although it may spare patients with treatment-responsive NGGCTs the risks of surgical intervention. However, this point remains controversial. The role of surgical resection for pineal parenchymal tumors is equally controversial (3), although there is a suggestion from several institutional series that disease control may be improved after more extensive resections, particularly for benign lesions (2).

For those patients who are appropriate candidates for tumor resection, the options for surgical approach are influenced by the location and extent of the tumor, and the preferences and experience of the surgeon. Supracerebellar infratentorial, suboccipital transtentorial, and interhemispheric approaches may each be appropriate, depending upon whether the tumor extends mainly above or below the vein of Galen and the degree to which the lesion grows lateral to the midline and anteriorly into the third ventricle. A challenge with each of these

approaches is the depth of the operative field and the need to work within narrow corridors surrounded by vital structures, such as the deep cerebral veins and brainstem.

Tseng et al. (9) report their adaptation of endoscopy to the supracerebellar approach. Because their approach, as outlined, involved making a lengthy skin incision and two sizable cranial and dural openings, which in total, were comparable to those used in a standard posterior fossa craniotomy and supracerebellar exposure, the principal contribution of the report is to highlight the feasibility of using an endoscope, rather than a microscope, to provide visualization of the pineal region needed to accomplish resection of a small tumor. A purported advantage of the approach was the ability to avoid dividing the midline occipital sinus in order to minimize the risk of air embolism, although it is not clear that the occipital sinus is a principal contributor to this problem. Nonetheless, this report adds to the growing literature that is expanding the boundaries of endoscopic applications for challenging, deep-seated intracranial lesions. The authors correctly note that the approach has a number of limitations, and allude to some of these in the paper's discussion, although it is unclear in how many other cases they have attempted this technique and the results they have achieved. They acknowledge that the endoscopic corridor may not be sufficient to allow removal of larger, firm tumors that do not deliver into the operative field after internal debulking, and that this approach can pose significant challenges in the event of serious bleeding, which can result from malignant tumors in this region as well as the surrounding venous structures that can be inadvertently injured during tumor removal.

Although the case illustrated by Tseng et al. (9) seemed ideally suited to consideration of an endoscopic approach as a proof of feasibility, their observations regarding a relatively small, centrally located lesion cannot be generalized to the management of larger, more extensive, highly vascularized tumors, based on this report alone. However, it is likely that additional applications of this approach as an investigational strategy, potentially in a more minimally invasive context, will be reported in the coming years as endoscopic instrumentation continues to improve. Appropriate case selection, focusing on lesions that may potentially benefit from surgical debulking, will be essential to such applications, keeping in mind that, independent of surgery, a combination of radiotherapy and chemotherapy continues to be an important component of the management of patients with germinoma, NGGCT, and malignant pineal parenchymal tumors.

REFERENCES

1. Alapetite C, Brisse H, Patte C, Raquin MA, Gaboriaud G, Carrie C, Habrand JL, Thiesse P, Cuilliere JC, Bernier V, Ben-Hassel M, Frappaz D, Baranzelli MC, Bouffet E. Pattern of relapse and outcome of non-metastatic germinoma patients treated with chemotherapy and limited field radiation: the SFOP experience. *Neuro-Oncology*. 2010; 12:1318–1325. [PubMed: 20716594]
2. Clark AJ, Sughrue ME, Ivan ME, Aranda D, Rutkowski MJ, Kane AJ, Chang S, Parsa AT. Factors influencing overall survival rates for patients with pineocytoma. *J Neuro-Oncol*. 2010; 100:255–260.
3. Jakacki RI, Zeltzer PM, Boyett JM, Albright AL, Allen JC, Geyer JR, Rorke LB, Stanley P, Stevens KR, Wisoff J, et al. Survival and prognostic factors following radiation and/or chemotherapy for primitive neuroectodermal tumors of the pineal region in infants and children: A report of the Childrens Cancer Group. *J Clin Oncol*. 1995; 13:1377–1383. [PubMed: 7751882]

4. Jubran RF, Finlay J. Central nervous system germ cell tumors: Controversies in diagnosis and treatment. *Oncology*. 2005; 19:705–711. [PubMed: 15971448]
5. Matsutani M, Sano K, Takakura K, Fujimaki T, Nakamura O, Funata N, Seto T. Primary intracranial germ cell tumors: A clinical analysis of 153 histologically verified cases. *J Neurosurg*. 1997; 86:446–455. [PubMed: 9046301]
6. Packer RJ, Cohen BH, Coney K. Intracranial germ cell tumors. *The Oncologist*. 2000; 5:312–320. [PubMed: 10964999]
7. Sawamura Y, de Tribolet N, Ishii N, Abe H. Management of primary intracranial germinomas: diagnostic surgery or radical resection? *J Neurosurg*. 1997; 87:262–266. [PubMed: 9254091]
8. Tomita, T. Pineal region tumors. In: Albright, AL.; Pollack, IF.; Adelson, PD., editors. *Principles and Practice of Pediatric Neurosurgery*. New York: Thieme; 2008. p. 585-605.
9. Tseng K-Y, Ma H-I, Liu W-H, Tang C-T. Endoscopic supracerebellar infratentorial retropineal approach for tumor resection-A technical report. *World Neurosurgery*. 2011
10. Weiner HL, Lichtenbaum RA, Wisoff JH, Snow RB, Souweidane MM, Bruce JN, Finlay JL. Delayed surgical resection of central nervous system germ cell tumors. *Neurosurgery*. 2002; 50:727–734. [PubMed: 11904022]