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CONTEMPORARY NEUROSURGERY

Posterior Surgical Approach to the Third Ventricle and Pineal Area

Henry Brem, M.D.,* and Bennett Stein, M.D.†

Tumors arising in the posterior third ventricle and pineal region lie in the central portion of the brain and are difficult to expose. The involvement of the deep venous system of the brain adds to the hazard of the operation. In most instances, however, the veins tend to be displaced dorsally. The arteries associated with the tumor are generally of small caliber and of relatively minor significance. These include the posterior choroidal arteries, branches of the quadrigeminal arteries, and the posterior cerebral arteries.

Pineal region tumors are rare, representing about 1% of cerebral gliomas. In Japan where the lesion is more common, the incidence is approximately 10% of gliomas. Approximately 25% of tumors occurring in the pineal region are benign and encapsulated and thus amenable to surgical resection. Tumors that can be resected include simple nonneoplastic cysts, ependymal cysts, dermoids, teratomas, meningiomas, ependymomas, and some low-grade astrocytomas. Furthermore, the infiltrative tumors in this region are heterogenous and behave with different degrees of aggressiveness. Histological typing is useful in selecting appropriate therapy for patients with these tumors. In addition to the presently available therapies of shunting and radiation, several chemotherapeutic protocols are under development for specific tumor types. Microneurosurgical techniques including the operating microscope, microinstrumentation, the Cavitron, and laser have permitted pineal tumor operations with minimal morbidity and mortality.

Anatomic Considerations

Pineal region tumors lie deep within the cranium and tend to grow spherically. The important anatomical relationships are shown in Figure 1. The tumor intrudes into the posterior third ventricle or aqueduct by direct pressure or growth of the lesion into the quadrigeminal region. The internal cerebral veins, great vein of Galen, and pontomesencephalic veins, i.e., the deep venous system, lie dorsal and lateral to the tumor. These veins tend to be displaced by tumor growth and rarely are totally enveloped by the tumor. They form an important and dense network overlying the tumor and must not be injured or occluded during attempts at tumor resection. The tumor extends into the posterior fossa and grows in relationship to the anterior lobe of the cerebellum, displacing but not enveloping the precentral cerebellar vein posteriorly.

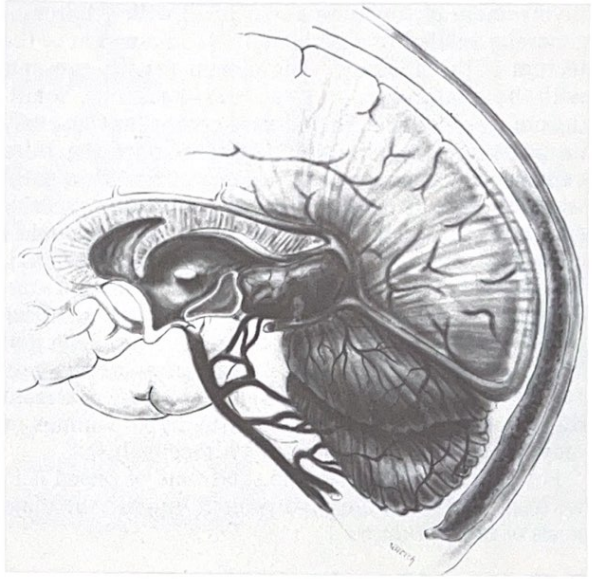

Figure 1. Illustration of a pineal region tumor, showing the anatomical relationships to the third ventricle and internal cerebral venous system, midbrain, and cerebellum.

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Categories:
Tumor • Techniques

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Pineal tumors • Pineal tumor surgery • Posterior third ventricular tumors • Surgical positioning • Pineal tumors, anatomic and pathologic aspects

In many instances, it may be impossible to determine the exact locus of origin of these tumors, which may arise from pineal or posterior third ventricular structures. Although the pineal gland normally contains a mixture of cells, it is highly probable that the gliomas arise from the quadrigeminal region or posterior thalamic regions, whereas meningiomas presumably arise from the tela choroidea. Pineal tumors generally obtain their blood supply from the lateral and medial posterior choroidal arteries and from branches of the quadrigeminal, posterior cerebral and, on occasion, superior cerebellar arteries. In most instances, the pineal tumors are not highly vascular; thus, no major feeding arteries can be shown by arteriography.

Clinical Considerations

These tumors produce symptoms by two methods: first, by production of raised intracranial pressure (ICP) with hydrocephalus secondary to obstruction of the ventricular system, usually in the region of the aqueduct or posterior third ventricle; and second, by direct involvement of contiguous structures with syndromes generally related to destruction or compression of the tectum of the midbrain. The patient usually presents with the classic signs of raised ICP—headache, vomiting and papilledema. In addition, cerebellar signs may be prominent either due to distention of the third ventricle with pressure on the cerebellar outflow pathways or due to direct involvement of the superior cerebellar peduncle. The typical syndrome (Parinaud's syndrome) of involvement of the tectum of the midbrain consists of failure of upward gaze, lack of convergence, and failure of the near reflex. In addition, pupils may be irregular and unequal. Precocious puberty and diabetes insipidus occur in approximately 10% of the cases each. These symptoms may represent direct seeding of the tumor into the hypothalamus or may reflect a severe degree of hydrocephalus.

For practical purposes, the syndrome of raised ICP, with signs of midbrain involvement, implies the diagnosis of pineal tumor.

Pathology of Pineal Region Tumors

Tumors in the pineal region represent a variety of pathological lesions. Rubinstein describes three major categories of pineal region tumors: (1) tumors of the pineal parenchyma including pineocytoma, pineoblastoma, ganglioneuroma, chemodectoma, and glioma; (2) tumors of germ cell origin including germinoma, teratoma, choriocarcinoma, and embryonal carcinoma; and (3) miscellaneous neoplasms of the pineal region including meningioma, pineal cyst, and metastatic tumor.

Tumors arising from pineal parenchymal cells are less common than germinomas and teratomas which constitute over 50% of pineal region tumors. Pineal parenchymal cells are neither neuronal nor neuroglial. Histologically, they are characterized by single and branching cytoplasmic argyrophilic processes with typical club-shaped protrusions surrounding blood vessels. According to Rubinstein, the natural history of these tumors is not known: some behave as malignant neoplasms, and others have a clinical history of several years, suggesting a slow evolution. In the 1982 supplement to the *Fascicle of the Armed Forces Institute of Pathology*, however, Rubinstein describes new advances in pathology which provide further insight into these lesions. He reviews the pathology of 28 pineal parenchymal tumors and separates them into two main categories: pineoblastomas (uniform sheets of immature, poorly differentiated cells) and pineocytomas (lobar architecture resembling the normal pineal gland). Both types may show evidence of further cellular differentiation as demonstrated by newer methods of silver impregnation; immunoperoxidase, demonstrating glial fibrillary acidic protein; and electron microscopy. All of the patients with either pineoblastomas or pineocytomas without differentiation died within 2 years of onset of symptoms. These tumors should, therefore, be treated vigorously, possibly with neuroaxis radiation. By contrast, 10 of 17 pineocytomas showed evidence of astrocytic and neuronal differentiation. These tumors were all observed in adults, invariably remained localized, and often had a prolonged clinical course, indicating the slow growth of the tumor. Neoplasms with this degree of neuronal differentiation may not be radiosensitive; thus surgical excision, in our opinion, is an appropriate form of treatment.

For certain tumors of this region (cysts, teratomas, dermoids, meningiomas, and some astrocytomas), therefore, resection is appropriate; for those lesions that cannot be totally removed, biopsy will aid in determining the appropriate treatment for each individual tumor type.

Radiological Considerations

Plain skull x-rays may be important in identifying an abnormal amount of calcium in the pineal region. Calcium occurring in the pineal at an early age is considered pathological, and a calcified pineal measuring in excess of 5 mm is abnormal at any age. In

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addition, signs of raised ICP, such as suture separation, demineralization of the sella turcica, and distortion of the various cranial fossae, may be present. Definitive diagnosis and identification of the size and configuration of a pineal tumor are based on an evaluation of the ventricular system.

Computed tomography (CT) provides more information than was previously obtained from ventriculography. In addition, some clue as to the intrinsic nature and vascularity of the tumor may also be afforded by CT scanning, with and without contrast. Ganti has recently reviewed the CT features of 49 histologically proven pineal region tumors from the Neurological Institute of New York. On the basis of CT characteristics, he divided the tumors into five groups—germinomas, teratomas, glial tumors, pineal parenchymal cell tumors, and meningiomas. The germinomas were high-density lesions with prominent pineal calcification. CSF seeding had occurred in one third of the patients with germinomas. The *teratomas* showed calcification associated with fat densities. There was no contrast enhancement. Most of the *glial* tumors contained areas of hypodensity which showed enhancement in a nodular or ring pattern. The pineal calcification, when present, appeared to be displaced anteriorly. The *pineal parenchymal cell tumors* appeared isodense to hyperdense before contrast, and most enhanced in a nodular fashion. No pineoblastoma showed calcification. The *meningiomas* were hyperdense before contrast and enhanced in a nodular fashion. Thus CT scanning adds useful information but does not definitively provide a tissue-specific diagnosis.

Arteriography has been the least valuable of all of the specialized radiographic techniques. Most pineal region tumors are not highly vascular, and even those of moderate vascularity may vary in histological type and not necessarily be highly malignant. Arteriography will, however, rule out a vein of Galen vascular malformation as the cause of a pineal region mass.

In rare cases, a rapid neurological deterioration (measured in weeks) combined with CT and arteriographic evidence of a high degree of vascularity with malignant tumor vessels and arteriovenous (AV) shunting within the tumor will indicate with some certainty the diagnosis of a high-grade malignancy and preclude surgery.

The use of biological markers such as α-fetoprotein, human chorionic gonadotropin, and tumor angiogenesis factor have more value in gauging the effectiveness of therapy than in predicting the histological type of tumor.

In spite of contemporary specialized radiographic techniques, it is still impossible, except for those tumors mentioned previously, to establish without question the histological nature of these tumors prior to tissue diagnosis at the time of surgery.

Surgical Considerations

Classic Techniques

Dandy advocated an interhemispheric approach, with section of the posterior portion of the corpus callosum, to gain access to the pineal region. This approach required sacrificing a number of parietal cortical veins and retracting a large area of the parietal hemisphere, usually the right. Furthermore, in the region of the pineal, difficulties in dissecting through or around the deep venous system were encountered, and many of the fatalities appeared to arise from deep-vein thrombosis and edema of the diencephalic region. Van Wagenen proposed an approach through the right lateral ventricle, which is almost always dilated when tumors compromise or obstruct the aqueductal region. Exposure of the tumor still involved dissection of the deep venous system, since the approach was primarily dorsal to the tumor. In addition, Van Wagenen's approach had the disadvantage of a transcortical incision and possible hemispheric collapse when the ventricle was opened. Poppen and Horrax advocated an approach beneath the occipital lobe, in which bridging veins were sacrificed, a large portion of the hemisphere was retracted, and the free edge of the tentorium was sectioned to expose the deep venous plexus in a more favorable attitude relative to the underlying tumor. Krause in 1926 reported three patients, each with a different variety of tumor in the pineal or quadrigeminal region, which he approached through the posterior fossa, over the cerebellar hemispheres, and beneath the tentorium. The advantages of this approach are that injury to the deep venous system is avoided and the tumor is approached directly. Stein modified this approach with the introduction of microsurgical techniques and has now used it successfully in 72 cases (Figure 2).

For most tumors in the posterior third ventricular region, the infratentorial supracerebellar approach is appropriate. For a small group of tumors which are eccentric (i.e., to one side), the interhemispheric transcallosal approach has been useful. These tumors can be reached more effectively through one or the other lateral ventricles; generally, the tumor lies more in relation to the pulvinar of the thalamus than to the pineal or posterior third ventricular region. If there is evidence of tumor seeding, conservative management with a shunt and radiotherapy should be considered.

Operative Technique—Posterior Fossa Approach

If the patient has intracranial hypertension and hydrocephalus, the direct approach to the pineal tumor

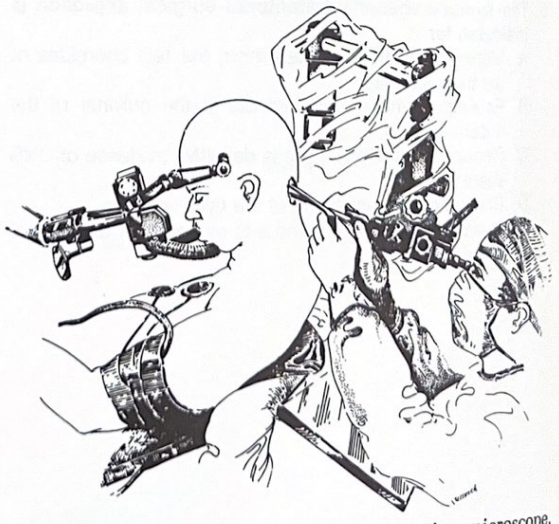

Figure 2. Positioning of the patient, operation microscope, and surgeon for the posterior fossa supracerebellar approach to pineal region tumors.

may be facilitated by a preliminary extracranial shunt in order to gradually reduce ventricular size and compensate for the raised ICP. In most cases, however, this is not necessary and a direct approach to the tumor is performed. Unless the patient is under 2 years of age and suffering from a severe degree of hydrocephalus, the sitting position is preferred. The patient's head must be markedly flexed so that optimal exposure of the tentorial notch can be achieved. For prevention of cervical cord compression secondary to the marked flexion, preoperative testing is performed to determine the safety of this degree of neck flexion. In addition to evaluation by cervical x-rays, evaluation of the patient can be performed by having the patient, while awake, sit for 5 minutes with his neck in forced flexion. The onset of neurological symptomatology such a Lhermitte's sign precludes the use of this position. Positioned on the operating table, the patient is tilted somewhat forward, with marked flexion of the neck, so that the surgeon actually works over the back of the patient in the posterior fossa (Figures 2 and 3). Care is taken to prevent or to recognize an air embolism, with use of a Doppler monitor, CO_2 monitor, central venous catheter, and modest positive-pressure ventilation during the opening or during exposure of large venous sinuses.

A long midline incision extending from C2 high up into the occipital region is used so that the pericranium and muscle attachments can be elevated to either side without disrupting their continuity. This greatly facilitates closure, since all muscle attachment must be elevated. Generally, a wide craniectomy which extends to the lateral sinuses and torcular is performed. In the initial series, the craniectomy was carried to the foramen magnum, and the posterior arch of C1 was removed. In the recent series, we have performed a more limited craniectomy which involves the superior region of the posterior fossa but does not extend to the foramen magnum (Figure 4). Spinal or ventricular drainage may be used to reduce volume and to facilitate retraction of the cerebellar hemispheres. The opening of the dura is important, for it must be done bilaterally toward the lateral sinus. The dural incision(s) should not be carried too far laterally, however, since in doing so one is unable to retract the flap of dura adequately in the region of the torcular (Figure 5). Once this is accomplished, the tentorium can be elevated with a self-retaining retractor. The weight of cottonoids or a retractor is sufficient to depress the cerebellum. Also, gravity helps with cerebellar retraction. The operating microscope with either a 250- or 275-mm objective lens is used. If a greater focal distance is used, the operator is too far removed from the incisural region and, even with an armrest, finds the position extremely fatiguing. Therefore, the microscope and operator must be relatively close to the field. Some type of armrest is recommended. In addition, relatively long microsurgical instruments are required because of the distance to the pineal region and beyond, as many of these tumors extend well into the third ventricle and some extend into the region of the foramen of Monro (Figure 6). All bridging veins over the dorsal surface of the cerebellum, including the hemispheres of the vermis, may be sacrificed in order to open the quadrigeminal region and the incisura. The arachnoid in this region is almost always thickened and opaque in the presence of tumors and must be opened by microdissection techniques to expose the surface of the tumor. The great vein of Galen and the internal cerebral veins generally are well above the tumor and are not encountered in these initial maneuvers. Laterally, the medial aspect of the temporal lobe is visualized, and the veins of Rosenthal can be seen as they course upward toward the confluence of veins in this region. The tumor capsule is cauterized and opened by sharp dissection. Depending on its consis-

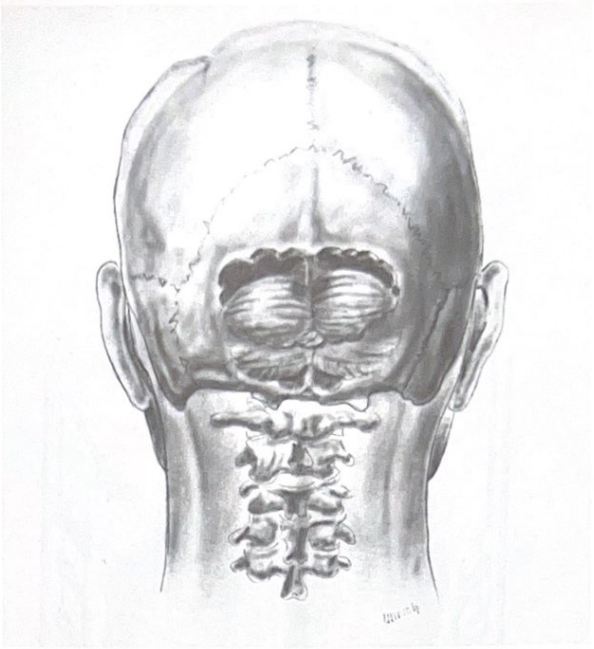

Figure 4. Area and size of the craniectomy.

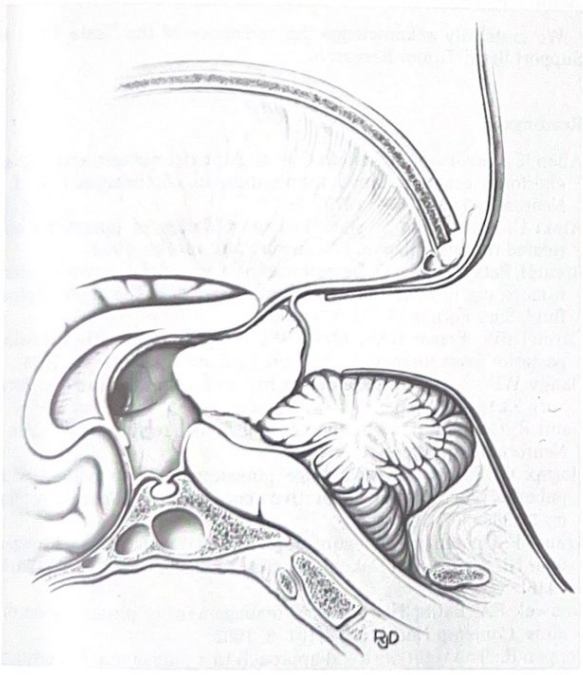

Figure 3. Sagittal brain section showing the posterior fossa supracerebellar approach to pineal region tumors.

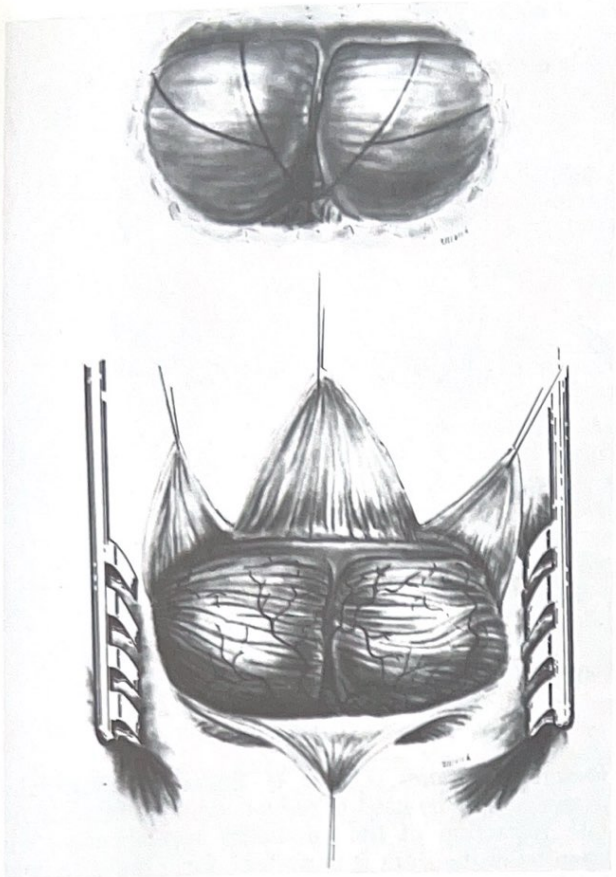

Figure 5. Configuration of the dural opening and the actual opening of the dura, exposing the posterior and superior surfaces of the cerebellum.

tency, the tumor is removed with tumor forceps, small curettes, suction, or cautery. The recent introduction of the ultrasonic aspirator (Cavitron) has added greatly to the safety and speed of removal of these tumors. With the extra-long Cavitron handle and a 275-mm microscope objective lens, this instrument rapidly removes tumor, even deep within the third ventricle. In our experience, the laser (through the microscope) has not added to facilitating tumor removal. In general, the trajectory of the operation is toward the velum interpositum. This must be considered when attempts are made to remove portions of the tumor in the inferior portion of the third ventricle or directly over the quadrigeminal plate. Operation in this area is most difficult, and small dental mirrors and angled instruments may be required to accomplish removal of tumor in this region. The lateral extent of the tumor can be pursued through this particular exposure.

Even with nonresectable tumors, benefit may be accrued through internal decompression of or an opening through the tumor adequate to expose the posterior third ventricle. This will allow placement of an internal shunt tube from directly within the third ventricle, through the interior of the tumor, and over the cerebellar hemisphere to the region of the cisterna magna. If the tumor cannot be removed completely or an internal shunt placed, ventricular drainage with subsequent conversion to a shunt may be necessary in the postoperative period.

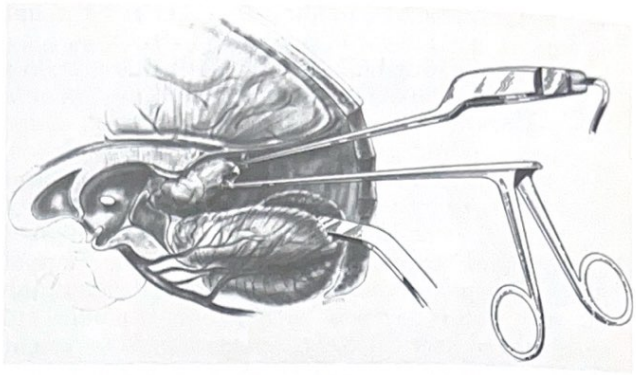

Figure 6. Sagittal plane of the brain, showing the location of the tumor and the extra-long instruments used in tumor removal.

Dural closure is facilitated by a significant decompression or removal of the tumor. Dural closure makes the postoperative course smoother and may limit the degree of aseptic meningitis.

Conclusion

Advancement in microsurgical technique as well as the knowledge that up to 25% of tumors in the posterior third ventricular region may be resectable has led to a more aggressive surgical approach to these tumors. Even with the malignant tumors there exists the potential for evacuation of a cyst and removal of some portion of the lesion for decompression. The identification of the exact histological nature of these tumors permits a more rational use of contemporary therapeutic techniques such as focal or neuroaxic radiation and/or chemotherapy.

Acknowledgment

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Answer questions on response card for volume 007, lesson 04

IMPORTANT: MARK ONLY ONE OVAL AND USE NUMBER 2 PENCIL ONLY

Response card must be mailed on or before May 9, 1985

1. **The arterial supply to pineal region tumors generally does not include**
   A. Posterior choroidal arteries
   B. Quadrigeminal artery branches
   C. Posterior cerebral artery branches
   D. Posterior inferior cerebellar artery branches
   E. Superior cerebellar artery branches

2. **The incidence of pineal region tumors is about**
   A. 1% in all reported series of gliomas
   B. 10% in Japanese series of gliomas
   C. 25% in Japanese series of gliomas
   D. 10% in American series of gliomas
   E. 25% in American series of gliomas

3. **Which of the following vessels cannot be sacrificed safely during pineal region tumor dissection?**
   A. Pontomesencephalic veins
   B. Precentral cerebellar vein
   C. Quadrigeminal artery branches
   D. Posterior medial choroidal arteries
   E. Cerebellar bridging veins

4. **Parinaud's syndrome consists of**
   A. Failure of upward gaze
   B. Lack of convergence
   C. Failure of the near reflex
   D. Irregular and unequal pupils
   E. All of the above

5. **The supracerebellar infratentorial surgical approach is indicated for**
   A. Meningiomas that originate from the tela choroidea of the third ventricle
   B. Eccentric tumors that originate in the pulvinar of the thalamus
   C. Pinealomas in which there is definitive evidence of CNS seeding
   D. Choroid plexus papilloma of the third ventricle
   E. Pineal tumors in which there is evidence of a chiasmal lesion

6. **Decompression of the hydrocephalus associated with a posterior third ventricle tumor can be accomplished by**
   A. Ventriculoperitoneal shunt
   B. External drainage of lateral ventricles perioperatively
   C. Spinal drainage intraoperatively
   D. An internal shunt from the third ventricle to the cisterna magna
   E. All of the above

7. **Precocious puberty and diabetes insipidus**
   A. Occur in approximately 10% of cases of pineal region tumors
   B. Are invariably found in patients with pineal region tumors
   C. Preclude the diagnosis of a pineal region tumor
   D. Occur in approximately 50% of cases of pineal region tumors
   E. Always present together

8. **Pineocytomas**
   A. Are malignant neoplasms
   B. Are uniformly fatal
   C. Should always be radiated
   D. Can be subdivided into a group that can be shown to be more differentiated and slower growing
   E. Are of a cell type that is suggestive of a lymphocytic infiltrate

9. **Calcified pineal on a plain x-ray is abnormal if**
   A. It is present in a 1-year-old child
   B. It measures more than 2 mm
   C. The habenular region is also calcified
   D. It is curvilinear
   E. All of the above

10. **When the supracerebellar infratentorial approach to the pineal region is utilized,**
    A. The craniectomy must extend above the sinus and torcular
    B. The ventricular system must be decompressed
    C. The bridging veins over the superior aspect of the cerebellum may be sacrificed
    D. Dural closure is useful in preventing postoperative aseptic meningitis
    E. All of the above