Pineal Region Mass

Matthew Minor, M.D., 1 William T. O’Brien, Sr., D.O. 2,3

1Department of Radiology, Wilford Hall Ambulatory Surgical Center, Joint Base San Antonio-Lackland, TX
2Department of Radiology, David Grant USAF Medical Center, Travis Air Force base, CA
3Department of Radiology, Uniformed Services University of the Health Sciences, Bethesda, MD

Case Presentation

A 15-year-old boy presented with acute exacerbation of chronic headache, as well as new onset nausea, vomiting, and visual changes. Past medical history and review of systems were noncontributory. Physical examination revealed mild papilledema and paralysis of upward gaze. The patient was subsequently referred for an emergent head CT, which was initially performed without contrast but then with contrast based upon the preliminary findings, followed by an MRI of the brain with and without contrast (Fig.).

Figure. Axial contrast-enhanced CT image (A) reveals an enhancing pineal mass with central regions of calcification. There is dilatation of the visualized portions of the third and lateral ventricles, as well as mild transepicondymal flow of CSF along the margins of the lateral ventricles. Sagittal T1 weighted MR image (B) with contrast demonstrates a lobulated, enhancing pineal region mass with compression of the underlying tectal plate and cerebral aqueduct. There is dilatation of the third ventricle and a normal-sized fourth ventricle.
Key Imaging Finding

Pineal region mass

Differential Diagnoses

Pineal cyst
Pineal germ cell tumor
Pineal cell tumor
Tectal plate glioma
Meningioma

Discussion

Pineal region masses include those that originate from the pineal gland, as well as those that arise from adjacent structures. Masses of the pineal region range from simple, benign cysts to high-grade neoplasms. Imaging plays an important role in establishing the appropriate diagnosis or a reasonable list of differential considerations, as well as for identifying underlying complications.

Tumors originating from the pineal gland represent 3-8% of all pediatric intracranial neoplasms and 0.4-1.0% of all adult intracranial neoplasms. Germ cell tumors are most common, particularly germinoma, followed by pineal cell tumors, to include pineocytomas and pineoblastomas.

The pineal gland ranges in size from 10-14 mm and is located within the midline above the tentorium and superior colliculi and below the splenium of the corpus callosum and vein of Galen. It develops as a diverticulum in the diencephalon roof of the 3rd ventricle during the second month of gestation. The gland itself is attached to the posterior aspect of the 3rd ventricle by the pineal stalk. The mature gland secretes melatonin, an endocrine hormone involved in multiple pathways, but most commonly known for its association with circadian rhythms. The pineal gland is composed of 95% pineocytes (specialized neurons related to retinal rods and cones) and 5% astrocytes. Unlike most intracranial structures, the gland is outside of the blood-brain barrier.

Small pineal masses may be asymptomatic, but as lesions increase in size, they compress adjacent structures and may become symptomatic. Poppen and Marino initially suggested 3 clinical phases to pineal region masses: 1) headaches with nausea and vomiting; 2) blurred vision, diplopia, changes in mental states, drowsiness, papillary changes, ataxia or dizziness, and paralysis of the extra-ocular muscles; 3) papilledema, weakness, and spasticity. Two common syndromes associated with pineal region masses include the Sylvian aqueduct syndrome and Parinaud syndrome. These syndromes are similar and result from compression of the mesencephalon. Typical clinical findings include paralysis of upward gaze, abnormalities of the pupil, and nystagmus retractorius. One dreaded complication of a pineal region mass is pineal apoplexy, which is a sequela of hemorrhage into a pineal cyst or tumor with sudden decrease in consciousness and headache.

Pineal cyst

Pineal cysts are very common and have an outer wall composed of 3 layers: inner gliotic tissue, middle pineal parenchymal tissue, and outer connective tissue. Pineal gland cysts are usually asymptomatic and range in size from 2-15 mm. If the cyst reaches 15 mm or larger, however, it may become symptomatic due to compression of the tectum. The vast majority of cysts are simple, unilocular, and follow CSF density (CT) and signal intensity (MRI), although the cyst may not completely suppress on FLAIR sequences secondary to proteinaceous content. Peripheral enhancement may be seen but is usually incomplete. Complex cysts may be multilocular with variable signal intensity but do not have solid enhancing components. Atypical cysts are followed on imaging and clinically to ensure stability and benignity.

Germ cell tumor

Germinomas are the most common intracranial germ cell tumor (GCT) and represent 1-2% of all intracranial neoplasms. They represent the most common pineal malignancy. On CT, germinomas are typically hyperdense due to high cellularity and tend to engulf pineal calcifications centrally within the mass. On MRI, germinomas are typically iso- to hyperintense to gray matter on T1 and T2 sequences and demonstrate avid, homogenous enhancement. They may also have cystic components. Germinomas are
prone to dissemination throughout the cerebrospinal fluid (CSF); therefore, imaging of the remainder of the neuroaxis should be performed. Pathologically, germinomas are very cellular tumors and highly responsive to radiation therapy. In the presence of a germinoma, hCG will often be elevated within the CSF.\textsuperscript{3} Teratomas are less common GCTs composed of elements from all three germ layers and typically have cystic and solid components with macroscopic fat.

**Pineal cell tumor**

Pineal parenchymal tumors range from low-grade pineocytomas (WHO grade I) to high-grade pineoblastomas (WHO grade IV). Pineocytomas are typically well demarcated, hypo- to isointense on T1, and hyperintense on T2 with avid enhancement. The lower grade masses may have cystic change with solid nodular components. More aggressive lesion are often large, lobulated, and highly cellular. Regions of increased cellularity are hyperdense on CT and may demonstrate restricted diffusion. Pineal calcifications may be seen along the periphery in an “exploded” pattern. Pineoblastomas have heterogeneous MR signal intensity and avid but more heterogeneous enhancement. Obstructive hydrocephalus due to compression of the cerebral aqueduct is common. Aggressive pineal cell tumors are prone to CSF dissemination; therefore, imaging of the entire neuroaxis should be performed.\textsuperscript{4}

**Tectal plate glioma**

A tectal plate glioma is a subset of midbrain gliomas that are typically indolent in nature and low-grade. The most common associated finding will be hydrocephalus as a sequelae of tectal plate enlargement and compression of the Sylvian aqueduct. On MRI, the tumor is most commonly well-circumscribed, iso- to hypointense on T1, and hyperintense on T2 weighted sequences. The glioma will demonstrate variable enhancement. Given that progression of tectal plate gliomas is rare, the overall prognosis is favorable with symptomatic treatment for hydrocephalus.\textsuperscript{5}

**Meningioma**

Meningiomas are benign extra-axial tumors (WHO grade I) of the meninges which result from over-proliferation of arachnoidal cap cells. They are the most common extra-axial tumors and comprise 13-20% of intracranial tumors in adults.\textsuperscript{4} Most lesions are asymptomatic unless there is compression of adjacent structures. Meningiomas are typically iso- to hypointense to gray matter on T1 and variable in signal intensity on T2 weighted sequences, depending upon their cellularity and presence of calcifications. A CSF cleft and broad dural base confirms an extra-axial location. Lesions enhance homogeneously and typically have a dural tail, which is nonspecific, but highly suggestive of a meningioma. On CT, meningiomas present as well-circumscribed dural-based masses which are iso- to hyperdense compared to brain parenchyma with intense, homogenous enhancement. Calcifications and adjacent bony hyperostosis are characteristic.\textsuperscript{4}
Diagnosis

Germ cell tumor (germinoma)

Summary

A variety of pathological entities may involve the pineal region, ranging from benign cysts to malignant neoplasms. Patients may be asymptomatic or present with symptoms most often related to compression of adjacent structures. Cross-sectional imaging, especially MRI, is essential in the work-up and management of pineal region masses. Simple, uncomplicated pineal cysts are benign and do not need further imaging. When presented with an enhancing mass originating from the pineal gland, on the other hand, it is important to image the entire neuroaxis, as aggressive lesions in this region are prone to CSF dissemination.

References


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