Lesions of the Pineal Region: A Practical Approach

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Learning objectives

A wide variety of lesions involve the pineal region, including neoplastic and non neoplastic processes. Tumors of the pineal region can be classified into those arising from the pineal parenchyma, germ cell neoplasms and lesions arising from adjacent structures. Overall they make up less than 1% of intracranial tumors in adults, however pineal region neoplasms account for 3-8% of intracranial neoplasms in the pediatric population.

We propose to attend these objectives:

Provide a practical differential diagnosis of lesions of the pineal region, with examples of germ cell tumors, neoplasms arising from the pineal parenchyma and other pineal region masses.

Discuss the imaging characteristics of pineal region lesions.

Describe the clinical features of pineal lesions.

Background

Lesions of the pineal region include a diverse group of entities. Although they do not have pathognomonic imaging findings, imaging in combination with laboratory evaluations sometimes helps narrow the differential diagnosis.

In order to interpret and understand the pineal lesions, it is important to highlight the normal anatomy of the pineal region. Then we will focus on signs and symptoms of pineal region masses and finally we will list the imaging features of lesions of the pineal region, including tumors of pineal parenchymal origin, germ cell neoplasms, pineal cyst, and other entities.

1. NORMAL PINEAL REGION ANATOMY

The pineal gland is usually approximately 8 mm long and 4 mm wide; it is located in the midline, above the tentorium and superior colliculi and below the splenium of the corpus callosum and the vein of Galen, and is attached to the superior aspect of the posterior border of the third ventricle (Fig 1).
Internal cerebral veins joins with the basal veins to form the great vein of Galen. Pineal and parapineal masses elevate these veins, whereas callosal masses depress them.

The pineal gland secretes melatonin which is involved in diurnal rhythms and does not have a blood-brain barrier and therefore enhances on contrast material-enhanced images.

2. SIGNS AND SYMPTOMS RELATED TO PINEAL MASSES

Signs and symptoms of pineal region masses are most often related to mass effect on the adjacent structures, but higher-grade structures, such as a pineoblastoma, may also invade the surrounding tissue. These signs and symptoms include Parinaud syndrome, hydrocephalus and precocious puberty.

Parinaud syndrome consists of a failure of conjugate vertical eye movement, mydriasis, failed ocular convergence, and blepharospasm due to compression or invasion of the tectal plate. When a patient presents with a paralysis of upward gaze, the possibility of a pineal mass should be considered.

Hydrocephalus results from obstruction of the aqueduct of Sylvius; patients may also develop headache, nausea, and vomiting as a result of increased intracranial pressure.

Precocious puberty is more commonly associated with germ cell tumors (GCTs) and may be related to increased human chorionic gonadotropin (hCG) secreted by the tumor.

Hemorrhage into a pineal tumor or cyst (rarely occurs) is referred to as pineal apoplexy; the most common presenting symptom is a sudden decrease in consciousness associated with headache.

3. TUMORS OF PINEAL PARENCHYMAL ORIGIN

Pineal parenchymal tumors are rare lesions, accounting for less than 0.2% of intracranial neoplasms. These group of lesions include the low-grade pineocytoma, the intermediate-grade pineal parenchymal tumor of intermediate differentiation (PPTID), and the highly malignant pineoblastoma.
3.1 Pineocytoma

Pineocytoma is a slow-growing grade I lesion, World Health Organization (WHO). Accounts for 14%-60% of pineal parenchymal neoplasms. Predominantly manifest in adults (mean age, 38 years). No gender predilection. The 5-year survival is 86%-100%. Cerebrospinal fluid (CSF) dissemination rarely occurs.

3.2 Pineal Parenchymal Tumor of Intermediate Differentiation

PPTID is classified as a WHO grade II or III neoplasm. They make up at least 20% of all pineal parenchymal tumors and affect patients of any age, but the peak prevalence is in early adulthood; a slight female preponderance is reported. The 5-year survival is 39%-74%. Cerebrospinal fluid (CSF) dissemination or other metastases rarely have been reported.

3.3 Pineoblastoma

Pineoblastomas are highly malignant WHO grade IV lesions that represent the most primitive form of pineal parenchymal tumors and account for 40% of pineal parenchymal tumors. They most commonly occur in the first 2 decades but can occur at any age, and there is no gender predilection. CSF dissemination and infiltration into adjacent structures commonly occurs and is the most common cause of death. The 5-year survival is 58%.

4. GERM CELL TUMORS

Several theories exist regarding the origin of intracranial GCTs. All of them suggest the aberrant migration of primordial germ cells. These lesions account for greater than half of the pineal region neoplasms. The WHO classifies them into germinomas and nongerminomatous GCTs. The nongerminomatous GCTs include teratomas, embryonal carcinoma, yolk sac tumor, choriocarcinoma, and the mixed GCTs. Germinomas represent the majority of these neoplasms, and teratomas are the second most common.

Central nervous system GCTs are most commonly located in the pineal and suprasellar regions. These lesions result in increased serum and CSF levels of tumor-produced oncoproteins (#-fetoprotein, #-hCG, placental alkaline phosphatase).

4.1 Germinoma
Germinomas account for 1%-2% of all cranial neoplasms, and 90% of patients are less than 20 years old. Central nervous system germinomas are similar histologically and genetically to dysgerminoma in the ovary and seminoma in the testis. Fifty percent to 65% of intracranial germinomas occur in the pineal region, and 25%-35% are located in the suprasellar region. Those that occur in the pineal region are 10 times more common in males. Dissemination by CSF and invasion of the adjacent structures commonly occur, but the prognosis is good (5-year survival at least 90%) and the lesions are highly responsive to radiation therapy.

4.2 Teratoma

Teratomas differentiate along ectodermal, endodermal, and mesodermal lines. There are three types of teratoma: mature teratoma (fully differentiated tissue), immature teratoma, and teratoma with malignant transformation. The last one is the least common form and demonstrates malignant degeneration of the mature tissues.

5. PINEAL CYST

Pineal cysts are reported in 25%-40% of cases in autopsy series. Pineal cysts occur in all age ranges but are most predominant in adults 40-49 years of age. These lesions are typically asymptomatic and are usually 2-15 mm in size. Follow-up studies have indicated that these lesions remain stable in size over time. When they exceed 15 mm, patients may become symptomatic, typically with headache or visual changes.

6. OTHER PINEAL REGION MASSES

Many other lesions occur in the pineal region but are less common and derive from the cell types that reside in the adjacent structures. These include meningioma, ependymoma, choroid plexus tumors, astrocytoma, ganglioglioma, epidermoid and dermoid cysts, and lipomas. Although rare, metastases also occur in the pineal region.

6.1 Lipoma

Lipomas arise from abnormal differentiation of the meninx primitiva. Lipomas represent malformations and not neoplasms. Blood vessels and nerves course through them, making resection difficult if required.
Fig. 1: Normal pineal anatomy. Sagittal T1-weighted magnetic resonance (MR) image shows the normal anatomy of the pineal region. The pineal gland (arrow) lies below the splenium of the corpus callosum. The vein of Galen crosses just above the pineal gland. The tectal plate is located immediately inferior to the gland.
1. TUMORS OF PINEAL PARENCHYMAL ORIGIN

Pineal parenchymal tumors expand and obliterate pineal architecture, "exploding" the normal pineal calcification toward the periphery.

1.1 Pineocytoma

At computed tomography (CT), pineocytomas are well demarcated, usually less than 3 cm, and iso-to hyperattenuating (Fig 2).

At MR imaging, pineocytomas are also well-circumscribed lesions that are hypo-to isointense on T1-weighted images and hyperintense on T2-weighted images. On post-contrast images, they typically demonstrate avid, homogeneous enhancement.

1.2 Pineal Parenchymal Tumor of Intermediate Differentiation

No specific imaging findings separate PPTID from pineoblastoma or pineocytoma. PPTIDs demonstrate high signal intensity on T2-weighted images and enhance on postcontrast images. Cystic areas may also be seen.

1.3 Pineoblastoma

CT reveals a large (typically #3 cm), lobulated, hyperattenuating mass. Nearly 100% of patients have obstructive hydrocephalus.

At MR imaging, pineoblastomas are heterogeneous in appearance, with the solid portion hypo-to isointense on T1-weighted images and iso-to mildly hyperintense to the cortex on T2-weighted images. Pineoblastomas demonstrate heterogeneous enhancement on postcontrast images.

Necrotic regions and hemorrhage may be present (Fig 3,4). Extensive cystic change rarely occurs in pineoblastomas. Cerebrospinal fluid dissemination is a common finding so examination of the entire craniospinal axis is mandatory.
2. GERM CELL TUMORS

These lesions may result in increased serum and Cerebrospinal fluid levels of tumor-produced oncoproteins (#-fetoprotein, #-hCG, placental alkaline phosphatase) (Table 1).

2.1 Germinoma

CT demonstrates a sharply circumscribed, hyperattenuating mass that engulfs the pineal calcifications. Hydrocephalus may be present

(Fig 5).

MR imaging typically reveals a solid mass that may have cystic components. Germinomas are iso-to hyperintense to gray matter on T1- and T2-weighted images and demonstrate avid, homogeneous enhancement on postcontrast images (Fig 6).

The possibility of CSF seeding necessitates imaging evaluation of the entire neuroaxis.

The differential diagnosis for these lesions includes the primary pineal neoplasms. However, if oncoproteins are present or engulfment of the pineal calcifications is noted at CT, these findings help narrow the differential diagnosis.

2.2 Teratoma

CT usually reveals a multiloculated, lobulated lesion with foci of fat attenuation, calcification, and cystic regions.

T1-weighted MR images may show foci of T1 shortening due to fat and variable signal intensity related to calcification (Fig 7). On T2-weighted images, the soft-tissue component is iso-to hypointense. The soft-tissue component demonstrates enhancement on postcontrast images.

The malignant form may have a more homogeneous imaging appearance (fewer cysts and calcifications), thus making it difficult to distinguish from other neoplasms.

2.3 Other germ cell tumors
Choriocarcinoma, yolk sac tumors, and embryonal carcinoma are rare neoplasms. These neoplasms may have imaging findings similar to those of other germ cell neoplasms or primary pineal neoplasms. Evaluation of serum oncoproteins assists in making the appropriate diagnosis. These lesions may also hemorrhage, resulting in T1 shortening (Fig 8).

3. PINEAL CYST

At MR imaging, pineal cysts are round or oval, thin-walled, and well-circumscribed. They typically demonstrate signal intensity similar to that of Cerobrospinal fluid on T1- and T2-weighted images. On fluid-attenuated inversion-recovery (FLAIR) images, the signal may not be completely suppressed due to the proteinaceous contents. On gadolinium-enhanced images, enhancement of the cyst wall occurs in most pineal cysts but is typically incomplete; this finding has been attributed to fragmentation of the pineal parenchyma as the cyst enlarges (Fig 9).

4. LIPOMA

At CT, lipomas have low attenuation, consistent with fat.

At MR imaging, they have the same signal characteristics as fat (hyperintense on T1-weighted images with saturation on fat-saturated images). No enhancement is seen on postcontrast images (Fig 10).

Images for this section:
**Fig. 2:** Pineocitoma in a young male discovered incidentally. Axial nonenhanced CT image shows a well defined pineal region mass iso-to hyperattenuating. The pineal calcification is exploded toward the periphery (arrow)
**Fig. 3:** Sagittal T1-weighted MR image shows a large mass in the pineal region with resultant hydrocephalus. The mass is isointense relative to gray matter with a central area of necrosis.
Fig. 4: Same patient as in Fig 3. a) Axial FLAIR-weighted MR image shows a large mass in the pineal region hyperintense relative to gray matter. Flow voids within the mass, hydrocephalus and transependymal edema are also present. b) Postcontrast T1-weighted MR image shows homogeneous enhancement within the mass and a central area of necrosis. Both thalami and tectal plate are affected.
Germ cell Tumors and associated oncoproteins

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<tr>
<th>Type of GCT</th>
<th>Oncoprotein*</th>
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<tr>
<td></td>
<td>AFP</td>
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<tr>
<td>Pure germinoma</td>
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<td>Syncytiotrophoblastic germinoma</td>
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<td>Choriocarcinoma</td>
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<td>Embryonal carcinoma</td>
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<td>Mixed GCT</td>
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Source.—Reference 1.

*AFP = α-fetoprotein, PLAP = placental alkaline phosphatase, + = present, – = absent.

Table 1
**Fig. 5:** Axial nonenhanced CT image shows a hyperattenuating lesion in the pineal region that has engulfed the pineal calcification(arrow).
**Fig. 6:** Same patient as in Fig 5. a) Axial FLAIR weighted MR image shows that the mass is hyperintense relative to gray matter. b) Axial postcontrast T1 weighted image demonstrates avid, homogeneous enhancement of the mass. Note the associated mild hydrocephalus. On sagittal T1-weighted MR image the solid mass is isointense relative to gray matter (not shown).
**Fig. 7:** Sagittal T1-weighted MR image shows a lobulated mass in the pineal region with foci of T1 shortening due to fat and variable signal intensity related to calcification.
Fig. 8: a) Sagittal T1 weighted MR image shows an heterogeneous pineal region mass with foci of T1 shortening due to hemorrhage. Note the associated hydrocephalus. b) Axial postcontrast T1-weighted MR image shows that the pineal region mass also has an heterogeneous enhancement with foci of necrosis/cyst. Involvement of the tectal plate and both thalami are also present (not shown). Evaluation of serum oncoproteins demonstrated high level of b-hCG. Biopsy of the lesion revealed that it corresponds to Choriocarcinoma.
Fig. 9: Pineal cyst (incidentally found) in a 22-year old woman. 
a) Sagittal postcontrast T1-weighted MR image shows an oval, low-signal-intensity, 20mm lesion in the pineal region, a finding consistent with a cyst. The lesion has a thin incomplete enhancing rim (arrow). No nodularity of the wall and no associated hydrocephalus are seen. 
b) Axial T2-weighted MR image shows an oval hyperintense lesion similar to cerebrospinal fluid (arrow), in the pineal region. 
c) Axial FLAIR image shows that the signal of the lesion (arrow) is not completely suppressed, due to the proteinaceous contents.
Fig. 10: Lipoma in a 44-year-old man (found incidentally). a) Sagittal nonenhanced T1-weighted MR image shows a well-circumscribed, hyperintense lesion in the pineal region. b) Saturation of the lesion on sagittal fat-saturated image demonstrate that the entire lesion is consistent with fat.
Conclusion

The differential diagnosis for pineal region lesions includes basically germ cell neoplasms and pineal cell neoplasms. Germinoma and pineoblastoma, have high attenuation at CT due to their high cellularity, but if pineal calcification is seen the germinoma will tend to engulf it, whereas in pineoblastomas it will be exploded to the periphery.

The presence of lipid or fat attenuation at CT leads to the differential diagnosis of a teratoma, or lipoma. Intrinsic increased T1 signal intensity may be seen in pineal parenchymal neoplasms or GCTs with hemorrhage, and in lipomas or teratomas. Use of fat saturation and gradient-echo sequences can help differentiate these lesions.

None of these tumors have a truly pathognomonic imaging pattern. Therefore histologic verification is necessary for almost every pineal region mass that appears to be neoplastic. Knowledge of the various imaging findings and use of CSF and serum laboratory studies will help narrow the differential diagnosis for pineal region neoplasms.

References


