

Pineal Cyst Associated with Apoplexy and Hydrocephalus: A Case Report

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ABSTRACT: Pineal cysts (PCs) are common findings on (Magnetic resonance Imaging) MRI, often incidental in females and asymptomatic throughout life. Rare complications, like pineal apoplexy with acute hydrocephalus, require differential diagnosis and urgent intervention. We report a 19-year-old male with a progressive headache and visual decline. MRI showed a 2.3 cm pineal cyst with hemorrhagic features (apoplexy) causing hydrocephalus. He underwent endoscopic third ventriculostomy and microsurgical resection, both successful. Postoperatively, symptoms resolved completely. Though often benign, complicated PCs can be life-threatening. This case highlights their management, aiding understanding of etiologies, differential diagnoses, and treatments, enhancing medical knowledge

KEYWORDS: Pineal cyst, Apoplexy, Hydrocephalus, Endoscopic treatment, Pineal Gland.

Background

Pineal gland cysts are generally incidental findings, typically not associated with neurological symptoms.

When neurological symptoms are present, they are usually due to the cyst's growth, leading to mass effect, or following intracystic hemorrhage, termed pineal apoplexy, a rare and uncommon complication with few cases reported.

In patients exhibiting symptoms characteristic of pituitary growth and mass effect, cysts were present in 1.5% to 4.3% of MRI scans.

However, even in asymptomatic patients across all age groups, their incidence ranges from 1.2% to 10.8%, representing incidental findings [1].

The treatment varies depending on the patient, with options ranging from monitoring to surgical intervention.

We present a case involving a young male patient with a bleeding pineal cyst accompanied by hydrocephalus, along with a review of the existing literature on the topic.

Case Report

A 19-year-old male patient with a prior diagnosis of attention deficit hyperactivity disorder (ADHD), without other comorbidities, presented with a subacute onset of intense, progressive holocranial headache, which subsequently led to a decline in visual acuity.

Neurological examination did not reveal focal deficits; however, the patient exhibited left superior homonymous quadrantanopia and bilateral papilledema.

MRI revealed an expansive lesion in the pineal gland region, with a cystic appearance, measuring 2.3cm at its largest axis.

The imaging also showed a markedly hypointense focus on susceptibility-weighted sequences, indicating a fluid-fluid level suggestive of previous hemorrhage, likely related to pineal apoplexy.

This lesion was compressing the superior portion of the quadrigeminal plate and causing dilation of the supratentorial ventricular system, consistent with hydrocephalus.

It was also associated with hyperintensity on T2/FLAIR sequences in the white matter surrounding the lateral ventricles, which may be related to cerebrospinal fluid (CSF) transudation.

Additional laboratory tests were performed, along with an electrocardiogram and chest imaging, all of which showed no abnormalities. (Figure 1).

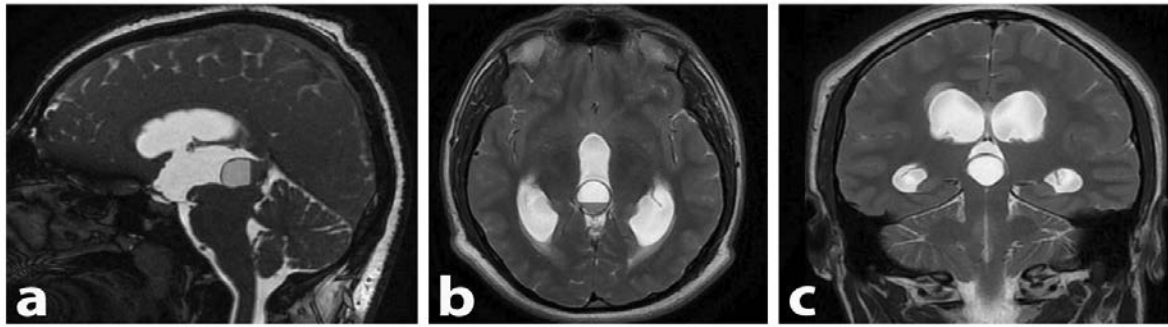


Figure 1. Pre-operative MR imaging. a) Sagittal T2-weighted MRI, evidencing a mass in the pineal region, with both solid and liquid components, obstructing the aqueduct of Sylvius and causing enlargement of the lateral and third ventricles. b) Axial T2-weighted MRI, confirming the clear boundary between the solid and liquid components inside the pineal mass, and important enlargement of the lateral and third ventricles. c) Coronal T2-weighted MRI, highlighting the enlargement of the lateral and third ventricles.

A two-stage surgical treatment was indicated, beginning with an endoscopic third ventriculostomy, followed two days later by microsurgical resection of the lesion through a supracerebellar infratentorial approach.

Both procedures were performed without complications and, in the postoperative period,

the patient reported no new complaints and experienced a complete remission of symptoms.

Also, the patient gave permission for the publication of this case, and a consent form was signed by hand by him (Figures 2 and 3).

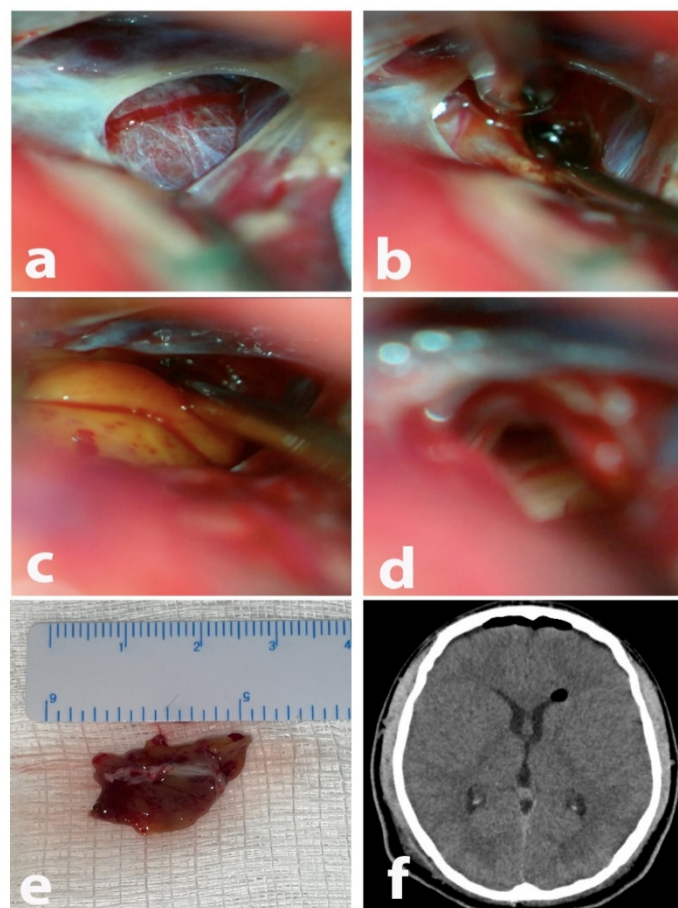


Figure 2. Intraoperative MR imaging. a) Initial exposure, evidencing the pineal mass after careful dissection of the quadrigeminal cistern arachnoid. b) After opening of the capsule of the lesion, the hematic component (previous bleeding inside the cist) drained spontaneously. c) Dissection of the cist capsule. d) Exposure of the third ventricle after total resection of the lesion. e) Surgical specimen. f) Post operative CT scan, evidencing resolution of the hydrocephalus and complete resection of the cist.

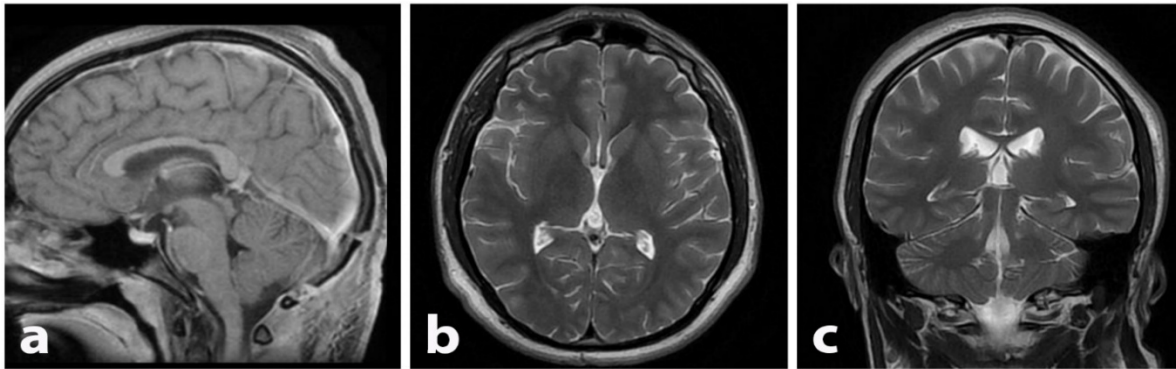


Figure 3. Post-operative MR imaging. a) Sagittal T1-weighted MRI, with gadolinium enhancement, evidencing a complete resection of the cyst in the pineal region. b) Axial T2-weighted MRI, confirming the resection and resolution of the enlargement of the ventricles. c) Coronal T2-weighted MRI, highlighting the total resection of the cyst and resolution of the hydrocephalus.

Discussion

The exact cause of PCs is still not clearly understood.

One prevailing theory links their development to abnormalities during embryogenesis.

In early brain development, the pineal gland emerges from the proliferation of tissue in the third ventricle's diverticulum.

Occasionally, remnants of this diverticulum may persist, forming a fluid-filled space lined by cells capable of becoming ependymal in nature.

Such PCs, characterized by ependymal lining, are believed to be the result of an expansion of this embryonic cavity.

Conversely, some cysts lack this ependymal layer and are instead enclosed by a glial scar.

This finding has led some scientists to propose that these types of PCs may originate from ischemic damage to glial tissue.

Additional hypotheses suggest that they could form through the necrosis of pineal gland tissue, the breakdown of pineal cells, or ischemic injury affecting the entire gland. [2].

PCs are more frequently observed in females aged 30 to 40, which has led to the hypothesis that hormonal factors, such as those related to ovulation and pregnancy, might contribute to their development or enlargement. [1].

However, the reported case contrasts with this statistic, as the patient is a young male, which adds to the rarity of the described case.

Most PCs are small, with 80% measuring under 10mm and often asymptomatic.

Larger cysts (>15mm) can cause neurological symptoms due to mass effect or hydrocephalus from aqueduct compression [3].

Wisoff and Epstein described three distinct clinical patterns: 1) sudden-onset headaches accompanied by gaze palsy, 2) persistent

headaches associated with motor weakness, optic disc swelling, and buildup of CSF (hydrocephalus), and 3) hemorrhage within the pineal region (pineal apoplexy) presenting with acute hydrocephalus [4].

Symptoms mainly result from compression of nearby structures, such as the quadrigeminal plate, leading to Parinaud's syndrome with upward gaze paralysis, light-near dissociation, and convergence-retraction nystagmus [5].

Other compressed structures may cause hydrocephalus, intracranial pressure, gait issues, and endocrine dysfunctions.

Pinealocytes, essential for melatonin regulation, may be disrupted, leading to hormonal imbalances like precocious puberty, hypogonadism, or diabetes insipidus [1].

Rare complications include hemorrhage, parkinsonian symptoms, papilloma, or aseptic meningitis from cyst rupture [6].

Although computed tomography (CT) is frequently the first imaging technique used in neurological assessments, MRI is still considered the most reliable method for accurately identifying and distinguishing various lesions.

MRI may indicate pineal cystic lesions when it reveals an oval-shaped signal anomaly in the pineal recess, with uniform appearance on T2-weighted images, signal intensity similar to CSF, absence of irregular borders or nodular enhancement after contrast administration, and a cyst wall measuring less than 2mm in thickness.

These features distinguish PCs from other masses. MRI is also preferred for follow-up [7].

These lesions typically show a uniform signal on T1-weighted images, appearing slightly brighter than CSF fluid, which is attributed to their elevated protein concentration. T2-weighted images show uniform hyperintensity, and

contrast administration enhances the thin cyst wall uniformly [7].

PCs can be distinguished from other abnormalities in the pineal area, such as tumors, using both tissue analysis and imaging studies.

On MRI, PCs usually present as oval or circular structures with well-defined edges.

A cyst wall measuring no more than 2mm in thickness is a typical feature of this lesion.

On T1-weighted scans, PCs display a consistent signal that is marginally more intense than that of CSF.

This minor increase in intensity is thought to be due to elevated protein levels within the cyst contents.

In contrast, T2-weighted imaging shows a consistently bright signal.

After contrast is given, there is even enhancement of the thin wall of the cyst.

From a histological perspective, PCs are made up of three separate layers.

The innermost portion is formed by fibrillary glial tissue, which may or may not contain hemosiderin.

This is encased by a middle layer composed of pineal gland parenchyma, sometimes featuring calcifications.

The outermost component is a delicate layer of fibrous tissue originating from the leptomeninges [8].

Intraoperatively, PCs generally present as smooth, single-chambered structures with an external layer exhibiting a brownish hue.

The material inside the cyst can differ, ranging from clear fluid to blood-tinged or clotted substances [4].

Pineal tumors, the main differential for PCs, account for up to 1% of brain neoplasms [9].

These include germ cell tumors (GCTs), such as germinomas and teratomas, and parenchymal tumors, including pineocytomas and pineoblastomas.

GCTs are the most common pineal tumors, comprising 0.5-4% of adult and 3-15% of pediatric brain tumors.

They often invade CSF spaces, requiring spinal MRI with contrast for diagnosis and follow-up [11].

Germinomas are frequent central nervous system (CNS) GCTs and pineal masses. CT shows well-circumscribed hyperattenuated masses, and MRI reveals solid lesions, sometimes cystic, with hyperintensity on T1/T2, homogeneous post-contrast enhancement, reduced diffusion, and possible hydrocephalus [12].

Histology shows large cells with clear cytoplasm, round nuclei, and lymphocytic infiltration.

Granulomatous inflammation may obscure tumor parenchyma; PAS and placental alkaline phosphatase stains assist diagnosis.

Syncytiotrophoblastic giant cells are sometimes present [13].

Teratomas, the second most common pineal tumors (15%), mainly affect boys under 10 [14].

CT and MRI show multicystic masses with calcifications, fat, and strong heterogeneous enhancement [15].

Teratomas contain ectodermal, mesodermal, and endodermal tissues, classified as mature or immature.

Mature types have differentiated tissues like skin, CNS, cartilage, and gastrointestinal elements.

Immature types include embryonal tissues such as primitive mesenchyme, neuroectoderm, and fetal glands positive for α -fetoprotein [13].

Pineocytomas are rare, slow-growing WHO Grade I tumors in adults over 40 [10].

They are well-circumscribed, under 3 cm, and compress nearby structures with rare CSF dissemination.

CT shows iso- to hypodense masses, while MRI reveals iso-signal on T1, hyperintense signals on T2-weighted images, and pronounced enhancement after contrast administration [12].

Histology shows moderate cellularity, pseudorosettes with fibrillar/granular centers, and occasional ganglion cells [13].

Pineoblastomas are rare, highly malignant tumors affecting children.

They often exceed 3 cm, infiltrate surrounding structures, and disseminate via CSF, causing hydrocephalus.

CT shows large, lobulated, or poorly defined hyperdense masses with intense post-contrast enhancement.

MRI shows hypo- to isointense T1 and heterogeneous enhancement [12].

Histology reveals small hyperchromatic cells, neuroblastic rosettes, photoreceptor differentiation, and necrosis.

Silver staining highlights fine processes, and mesenchymal components are rare [13].

Pineal apoplexy, indicated by the MRI findings in this case and linked to obstructive hydrocephalus, is regarded as the rarest and most severe manifestation of PCs [16].

The factors causing pineal apoplexy are diverse, with the most frequent being bleeding

within a pineal cyst or tumor, or hemorrhage originating from a nearby vascular anomaly [17].

No cases of bleeding in a healthy pineal gland have been documented, although such an event appears possible, especially in patients undergoing anticoagulant treatment [18].

Treatment depends on the symptoms presented by the patient.

Most authors agree that asymptomatic PCs do not require specific surgical intervention, and others find no benefit in performing routine imaging for asymptomatic adult patients [19].

Surgical management is reserved for symptomatic patients, except those with chronic headache without associated hydrocephalus.

The procedure can be performed through conventional craniotomy, typically using a supracerebellar infratentorial or occipital transtentorial approach, or through minimally invasive techniques such as stereotactic aspiration or endoscopic surgery.

Intracranial endoscopic techniques offer the advantage of addressing both the lesion and associated hydrocephalus via a third ventriculostomy [20].

Among these options, there have been reports of spontaneous regression of the cyst following the placement of a ventriculoperitoneal shunt or endoscopic third ventriculostomy.

A detailed explanation of this phenomenon is described as a change in the pressure gradient between the cyst and the ventricular cavity, resulting in fluid displacement from the cyst to the third ventricular space as a consequence of treatment aimed at normalizing ventricular pressure.

However, in hemorrhagic cases, it is important to differentiate from neoplasms such as glioma, pineocytoma, pineoblastoma, and GCTs.

Therefore, histological diagnosis is crucial, and micro-surgical or endoscopic resection is a viable approach for hemorrhagic PCs [21].

Conclusion

The origin of PCs remains uncertain; they are more frequently observed in women and are mostly asymptomatic.

Symptoms may be present in some cases due to mass effect and subsequent compression of adjacent structures.

Treatment options may be either conservative or surgical, involving resection of the lesion.

It is clear that our knowledge of this condition remains limited, highlighting the necessity for additional studies to clarify its causes and confirm

hypotheses about how it develops, ultimately aiming to enhance current therapeutic options

Abbreviations

MRI: Magnetic resonance Imaging; ADHD: attention deficit hyperactivity disorder; PCs: pineal cysts; CT: Computed tomography; CSF: cerebrospinal fluid; GCTs: Germ cell tumors; CNS: central nervous system.

Consent for publication

The patient gave approval for publication of this case.

A consent form has been signed by the patient.

The original of the signed form is held by the institution and can be made available to the editors upon request.

The manuscript was exempted from approval by the ethics committee.

The article was delivered to and read by the patient prior to its submission and publication.

The study was conducted in accordance with the declaration of Helsinki.

Competing interests

The authors declare that they have no competing interests

Conflict of interest

None to declare.

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