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The Median, Innate Bleb-Pineal Gland Cyst

Anubha Bajaj*

Department of Histopathology, AB Diagnostics, New Delhi, India

***Corresponding author:** Bajaj A, Department of Histopathology, AB Diagnostics, New Delhi, India, Tel: +91 9811693956; E-mail: nac.brain@hotmail.co.uk

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1. Preface

Pineal cyst is a commonly discerned, benign, non-neoplastic, midline, intraventricular, glial cyst emerging from the pineal gland. Although miniature and asymptomatic, pineal cysts may simulate a cystic neoplasm and engender hydrocephalus or depict associated, non-specific clinical symptoms. Neurosurgical intervention is a cogent therapeutic strategy. Additionally, stereotactic aspiration or resection and conservative management may be beneficially employed.

2. Disease Characteristics

Pineal cyst is predominantly situated within the quadrigeminal cistern wherein the anterior cyst wall protrudes into the third ventricle. Majority of pineal cysts are miniature, below <10-millimetre magnitude, asymptomatic and discovered incidentally [1-2].

Pineal cysts are discerned in around 3% of adult population and roughly ~20% of autopsies whereas microscopic lesions are delineated in up to 40% of pineal glands. The cyst may arise in children with short stature or precocious puberty. No age of disease emergence is exempt. A female predominance is observed and the female to male proportion of pineal cysts is around 1.7:1 [3-8].

Pineal cyst is commonly discerned within the third decade or fourth decade. The lesion is infrequent in children below < 10 years or in adults exceeding 70 years. Pineal cyst is usually configured in adolescence wherein cyst enlargement occurs through adulthood followed by cyst regression in elderly individuals.

Of obscure mechanism of initiation, cystic enlargement of pineal gland possibly occurs due to

- enlargement of an embryonic remnant of pineal diverticulum within the pineal gland.
- configuration of a distinct cyst layered by glial epithelium.
- degeneration or necrosis of pineal parenchymal cells.

Mechanisms which ensure emergence of cyst associated symptoms are posited as direct compression of tectum, compression of aqueduct or crowding of quadrigeminal cistern with consequent compression of deep venous system [9-13].

3. Clinical Elucidation

As pineal cyst is associated with clinical symptoms within the third decade or fourth decade, the cyst may emerge prior to clinical representation or symptoms may ensue from enlargement of a pre-existing cyst. Enlarged pineal cysts arising in adults may compress the aqueduct and engender diverse clinical symptoms.

Majority of pineal cysts are asymptomatic and are discerned as an incidental finding upon magnetic resonance imaging (MRI).

Despite an obscure genesis of symptomatic contribution, pineal cyst is commonly associated with obstructive hydrocephalus and Parinaud's syndrome. Besides, symptomatic pineal cyst non representative of hydrocephalus or Parinaud's syndrome may display chronic or paroxysmal headache unlike a typical migraine, intermittent nausea or vomiting, visual disturbances as blurred vision, greying of colours or altered visual perception, transient impaired consciousness, instability of gait or hypersomnolence.

Headache is a common, predominant symptom which manifests as a constant, dull pressure or fullness or may simulate a migraine. Headache is posited to arise due to direct hormonal alterations, hormonal modifications, and redistribution through body fluid content. Imaging may depict an absence of ventriculomegaly.

Visual symptoms associated with non-hydrocephalus pineal cysts emerge as blurred vision, double vision, delayed acquisition or binocular fusion of visual images with altered gaze or 'tired', painful eyes. Prolonged visual stress or frequent switching of gaze may trigger headaches, vertigo, and disorientation. Ophthalmological examination is generally normal and additional abnormalities are absent. Aforesaid dorsal midbrain symptoms possibly ensue due to direct compression by pineal cyst. Vertigo, unsteady gait, nausea, sensory symptoms or. 'neurology- not otherwise specified, 'psychiatric' symptoms as multisensory dysfunction and dissociative disorders may occur as a consequence to chronic headache, sleep disturbance, impaired fulfilment of personal and family, work or diverse social expectations, compounded by lack of efficacious therapy. A combination of 'headache-visual', 'headache-visual-nausea/vomiting', 'headache-visual-vertigo/dizziness- not otherwise specified' symptoms are commonly discerned.

Symptoms may deteriorate in pregnancy, obesity and may worsen in early mornings, a feature described as a 'bad hangover'. However, intracranial pressure is generally not raised in symptomatic, non-hydrocephalic pineal cyst. Besides, symptoms engendered by pineal cyst may resolve or ameliorate following surgery.

4. Histological Elucidation

Upon macroscopic examination, the cyst exhibits a smooth extraneous surface and an intrinsic layer. Upon cytological assessment, the cyst is layered with miniature, uniform polygonal cells.

Upon microscopy, the pineal cyst manifests as a three-layered cyst wherein the intrinsic, hypo-cellular coating is composed of extensively fibrillary, glial tissue with innumerable Rosenthal fibres. Foci of hemosiderin pigment deposition are commonly discerned. The glial layer is circumscribed by compressed parenchyma of pineal gland with superimposed focal calcification.

The extraneous layer is comprised of leptomeningeal connective tissue. Characteristically, pineal cyst is composed of an intrinsic layer of glial tissue with a circumscribing perimeter of pineal gland tissue.

Pineal cyst may manifest as simple, unilocular cyst or complex, multilocular cyst traversed with septa and an absence of an epithelial layer.

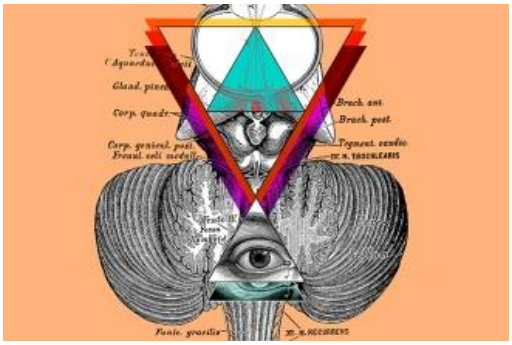

FIG. 1. **Pineal cyst depicting a cystic cavity within the midline pineal gland [14].**

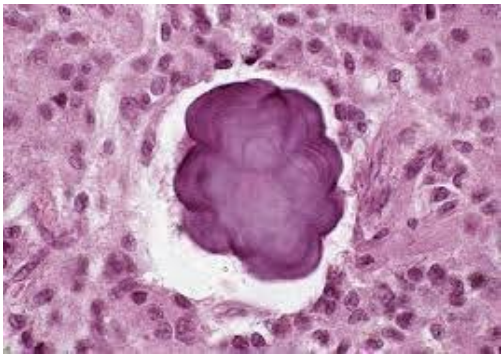

FIG. 2. **Pineal cyst exhibiting a cyst within pineal gland region with circumscribing fibro-connective tissue [15].**

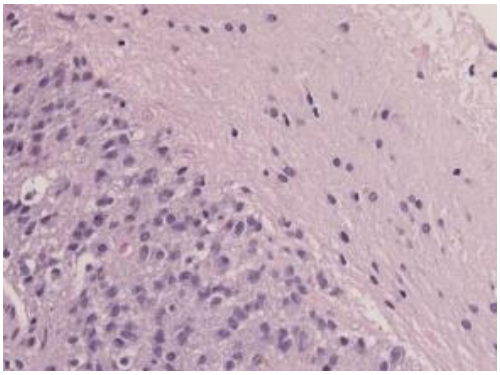

FIG. 3. **Pineal cyst delineating a cystic cavity lined by hypo-cellular, fibrillary glial tissue with Rosenthal fibres [16].**

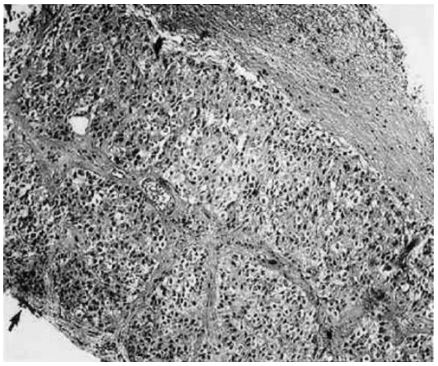

FIG. 4. **Pineal cyst displaying a cystic cavity layered with fibrillary glia and admixed Rosenthal fibres [17].**

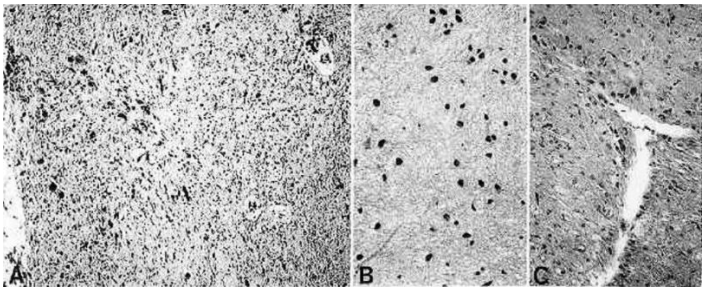

FIG. 5**. Pineal cyst exemplifying a cystic cavity with hypo-cellular coating, Rosenthal fibres and compressed pineal gland tissue [17].**

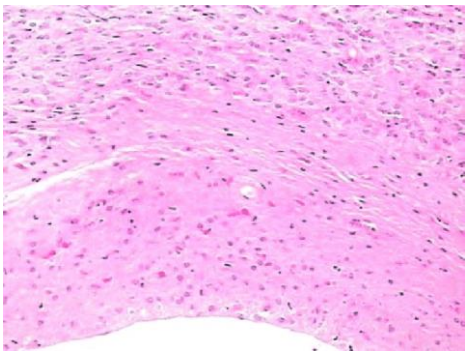

FIG. 6. **Pineal cyst enunciating a cavity with hypo-cellular coating, fibrillary glia and compressed, surrounding pineal gland [18].**

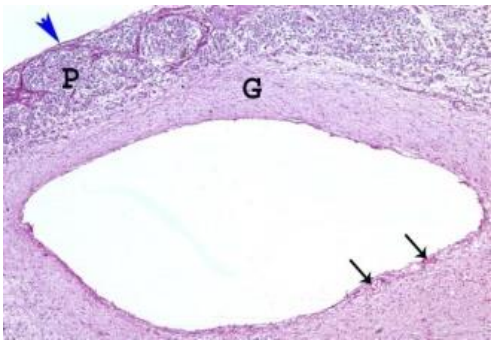

FIG. 7. **Pineal cyst exhibiting a cavity layered with hypo-cellular, fibrillary glia and circumscribing compressed pineal gland tissue [18].**

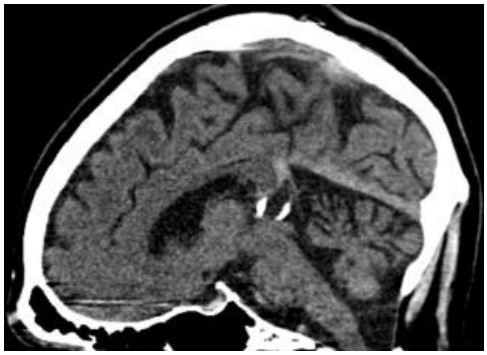

FIG. 8. **Pineal cyst demonstrating a midline, well circumscribed, enhancing lesion with an attenuated perimeter [19].**

5. Differential Diagnosis

Pineal cyst requires a segregation from lesions such as

- pilocytic astrocytoma which is a well circumscribed neoplasm depicting a predominantly solid pattern with limited tumour cell infiltration upon the periphery. Nevertheless, tumefaction may extend into the subarachnoid space. The neoplasm exhibits a biphasic appearance comprised of compact segments of fibrillary cells with elongated nuclei, bipolar piloid processes and Rosenthal fibres. Alternating loose, microcystic, loosely configured cellular zones exhibit spherical to elliptical nuceli, cobweb-like cytoplasmic processes and granular, eosinophilic bodies. Occasional multinucleated cells and glomeruloid vascular articulations are observed. Focal degenerative alterations are comprised of degenerative atypia, vascular hyalinization, infarct-like necrosis, focal calcification and perivascular accumulation of lymphocytes. Few instances depict anaplasia with enhanced mitotic activity exceeding >4 mitoses per 10 high power fields. Focal necrosis may or may not be discerned.
- pineocytoma is a well differentiated, non-infiltrative, hyper-cellular neoplasm which simulates normal pineal gland. Fibro-vascular stroma circumscribes expansive lobules of tumour cells with pinocytomatous rosettes demonstrating enlarged, loose, Homer Wright- like rosettes with centric fibrillary zones encompassed by neoplastic cells imbued with uniform, spherical nuclei. Focal neuronal differentiation with ganglion cells can be discerned. Mitotic figures and focal atypia are exceptional. Tumour necrosis is usually absent.

6. Investigative Assay

Upon imaging, a midline, well circumscribed cyst of up to 3-centimetre magnitude is observed. Upon magnetic resonance imaging (MRI) pineal cyst is categorised as a simple cyst or atypical cyst. Simple pineal cyst is unilocular with a smooth, attenuated cyst wall which may lack enhancement upon imaging. Cyst magnitude varies from below <5-millimetres or may exceed >25-millimetres.

Atypical pineal cyst is multilocular with multiple septa traversing the cyst and demonstrates a solid, enhancing component within the posterior segment abutting internal cerebral veins.

Majority of pineal cysts appear isointense with constituent cerebrospinal fluid (CSF) upon T1 weighted imaging and T2 weighted imaging although remain hyper-intense upon Fluid Attenuated Inversion Recovery (FLAIR) due to impacted

proteinaceous contents. Occasionally, focal calcification, preceding foci of haemorrhage and hemosiderin pigment can be incorporated within the cyst. Although debatable, imaging with gadolinium contrast at 12-month interval can be adopted for pineal cysts exceeding >10-millimetre magnitude.

Fine needle aspiration cytology can be employed to obtain a rapid determination of lesion. Tissue sampling of the cyst wall can be obtained during cogent surgical procedures for pertinent diagnosis. Serum and cerebrospinal fluid (CSF) germ cell tumour markers aid in segregating diverse pineal neoplasms.

7. Therapeutic Options

Appropriate management of pineal cyst is contingent to age of incriminated individual and associated comorbidities. Generally, management of incidental pineal cysts or cysts engendering hydrocephalus or symptomatic pineal cysts devoid of accompanying hydrocephalus is variable.

Pineal cysts engendering hydrocephalus may manifest with elevated intracranial pressure. Intra-cystic haemorrhage may precipitate hydrocephalus, a feature designated as “pineal apoplexy”.

An endoscopic, third ventriculostomy can be employed to relieve the hydrocephalus and achieve endoscopic fenestration of pineal cyst wall. Monitoring with magnetic resonance imaging (MRI) imaging and CSF flow studies is recommended at 3 months, one year and three years to ensure functioning and patency of endoscopic third ventriculostomy is beneficial. Although not recommended, alternative diversion procedure of cerebrospinal fluid as insertion of a ventriculoperitoneal shunt, craniotomy and open resection of pineal cyst may be undertaken. Incidental pineal cyst arising in children necessitate monitoring with an interval MRI at one year or two years as the cysts may enlarge or depict cogent clinical symptoms. Incidental pineal cysts discovered upon imaging for head injury, tinnitus, neurological symptoms as new-onset seizures or cerebrovascular disease demonstrates altered cyst magnitude upon serial magnetic resonance imaging. However, majority of incidental pineal cysts occurring in adults may be managed conservatively with reassurance. Singular or contrast-enhanced MRI is usually not required.

Simple pineal cyst below ≤ 10 mm magnitude does not require monitoring and may be contemplated as a normal variation of pineal gland.

Atypical pineal cyst emerges as a multiloculated cyst with a posterior, enhancing, solid component and can be monitored with an annual, elective MRI for five years. However, atypical pineal cysts simulating a solid tumour are stable and extensive monitoring is unnecessary. Enlargement of incidental pineal cyst with occurrence of symptomatic hydrocephalus is exceptional. Incidental, asymptomatic pineal cysts exceeding ≥ 15-millimetre magnitude may display hydrocephalus or visual impairment. Thus, clinical monitoring and magnetic resonance imaging is beneficial and recommended.

Although debatable or associated with complications, surgical eradication of the exceptional, symptomatic pineal cyst generating non-hydrocephalic, nonspecific clinical symptoms as headache, altered sleeping pattern or gait, visual and sensory disturbances may be warranted for amelioration of symptoms.

Besides, symptoms such as headache, episodic loss of consciousness, seizures and psychiatric symptoms are ameliorated in a majority of instances. Typically, ‘non-migraine headaches’ resolve following surgery whereas ‘migraine headaches’ may resolve or the frequency and duration may decimate.

However, symptoms such as cognitive deficit, fatigue, sleep disturbances are minimally relieved. Following surgery, pineal cyst of up to 28-millimetre magnitude may display an amelioration or comprehensive resolution of preoperative symptoms.

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14. Image 1 Courtesy: MUSC Health.com
15. Image 2 Courtesy: King George Medical University
16. Image 3 Courtesy: Libre Pathology
17. Image 4 and 5 Courtesy: Journal of Neurosurgery
18. Image 6 and 7 Courtesy: Research gate
19. Image 8 Courtesy: Wikipedia.