Pineal apoplexy may lead to the development of ill-defined symptoms.\textsuperscript{1} Cases of pineal apoplexy have been reported in the literature over the past 25 years with patients harboring a diverse group of underlying pathological lesions.\textsuperscript{1-9} The premise that pineal apoplexy may more often occur in association with an underlying malignant lesion as opposed to a benign lesion is not supported in the literature.\textsuperscript{1} Hemorrhage within a pineal cyst, leading to an acutely declining neurological state has been reported in patients with and without anticoagulant therapy.\textsuperscript{1,4} Pineal cysts are benign lesions found incidentally in 25-40\% of all autopsies.\textsuperscript{10-12} Pineal cysts have recently gained more attention because of an increased frequency of diagnosis secondary to magnetic resonance imaging. Incidence rates in the...
general population are in the 1.5 to 4.3% range, with a higher rate of 5.8% being demonstrated among young females between 21 and 30 years of age.13-18

The discrepancy between the imaging frequency of these cysts and the number actually ever requiring treatment illustrates their benign nature. It is unusual for these cysts to enlarge to a degree such that they cause major symptoms. When symptoms do occur they are a function of either hydrocephalus secondary to aqueductal compromise or ocular findings secondary to a dorsal midbrain syndrome.

The underlying pathogenesis responsible for the development of these pineal cysts remains controversial. Theories for cyst development have included the persistence of the cavum pineale, invagination of the pineal gland, ependymal invasion of glial lacunae and the coalescence of smaller cysts.10,11,17,19,20

CASE REPORT

A previously healthy, 12-year-old girl presented with a six-day history of headaches, nausea, vomiting and syncopal episodes. Computed tomography performed by her referring hospital revealed a 10 by 14 millimeter hyperdense pineal region mass with associated noncommunicating hydrocephalus (Figure 1). On arrival at our emergency department she was unresponsive secondary to acute hydrocephalus. Her neurological exam dramatically responded after endotracheal intubation and placement of an external ventricular drain. Both serum and cerebrospinal fluid levels of α-fetoprotein and human chorionic-gonadotropin levels were within normal limits. Magnetic resonance imaging (MRI) demonstrated a hemorrhagic lesion within the pineal gland (Figures 2a-b).

Her hydrocephalus was definitively treated with an endoscopic third ventriculostomy. She recovered completely and we elected to follow her clinically and radiographically. Subsequent MRI imaging seven weeks later, demonstrated an increasing lesion within the pineal region with resolution of the blood products. (Figure 3a-3b) The lesion was largely cystic and demonstrated cyst wall enhancement on gadolinium infused magnetic resonance imaging.

Due to the increasing size of the lesion we felt a surgical biopsy, in order to gain a definitive pathological diagnosis, was indicated. As the child was asymptomatic and her hydrocephalus had already been treated with an endoscopic third ventriculostomy, one may have also chosen a more conservative approach of careful observation.

A supracerebellar, infratentorial surgical approach was adopted with

Figure 1: Axial unenhanced CT scan revealing a 10 by 14 mm hyperdensity in the pineal region.

Figure 2a: Sagittal unenhanced T1-weighted magnetic resonance image revealing a 14 by 12 mm lesion with a signal consistent with recent hemorrhage.

Figure 2b: Axial T1-weighted magnetic resonance image revealing a hyperintense lesion in the pineal region.
Figure 3a: Seven weeks after the initial hemorrhage, a sagittal T1-weighted magnetic resonance image, demonstrating a larger cystic lesion extending into the third ventricle with compression of the cerebral aqueduct.

Figure 3b: Seven weeks after the initial hemorrhage, a sagittal gadolinium enhanced T1-weighted magnetic resonance image demonstrating some enhancement of the posterior and superior wall of the cyst.

Figure 4: Histological examination of the specimen revealed three distinct layers. The inner layer was comprised of reactive astrocytes with abundant Rosenthal fibre formation and frequent granular bodies. Also present were occasional hemosiderin containing macrophages at the innermost aspect. The next layer was comprised of compressed normal pineal gland with the outermost layer consisting of a very attenuated fibrous connective tissue investment.

The three arrowheads at the inner aspect point to hemosiderin macrophages. The arrows within the wall point to representative Rosenthal fibres in the astroglial portion of the cyst wall. The box encloses a portion of the compressed normal pineal gland. The large arrow points to a blood vessel in the outer fibrous connective tissue layer. (H&E stained section x 200)

Figure 5: A sagittal gadolinium enhanced magnetic resonance image, three months postoperatively demonstrating no residual cyst.
the patient received gadolinium.17

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proton-density imaging these cysts were often much more

cerebrospinal fluid on T1-weighted and T2-weighted images. On

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DISCUSSION

Symptomatic pineal apoplexy is a rare occurrence, first being
described within the neurosurgical literature by Apuzzo et al1 in
1976. This case was of a 56-year-old man on anticoagulants who
presented with headache, lethargy, and a gaze palsy due to the
hemorrhage into a pineal cyst.

The first craniotomy for a pineal cyst was performed in 1914, and
since that time less than one hundred cases have been
reported.1,3-6,13-17,21-35 The majority of these cases represent
cysts that presented with mass effect on the dorsal midbrain
leading to visual complaints or symptoms secondary to
obstruction of cerebrospinal fluid pathways.

Symptomatic pineal cysts can occur in all age groups and may
be more common in females.6,9,22 Pineal cysts are relatively
common and have been found with increasing frequency in vivo
with the advent of modern imaging techniques.14,16 In spite of the
common occurrence of pineal cysts, their underlying
pathogenesis is not completely understood. Some authors have
suggested that these lesions result from minor imperfections in
the gland’s normal embryogenesis.5 Other theories concerning
their formation include normal involution of the pineal gland,
ischemic glial degeneration with secondary cyst formation, and
ependymal invasion of glial lacunae.12,21

It has also been postulated that larger pineal cysts may
actually represent a coalescence of smaller pineal cysts.36 This
coalescence explanation seems inadequate based on the
discrepancy between the rarity of large pineal cysts and the
significant number of smaller cysts found in autopsy series.12
This theory is further flawed by the predominance of young
patients within the symptomatic pineal cyst population.21 Various
reports have also demonstrated women first becoming
symptomatic with their pineal cysts during pregnancy.21,35 This
information, coupled with the higher rates of cyst formation in
younger females, has led many to believe that hormonal
influences associated with pregnancy or the menstrual cycle may
play a role in the pathogenesis of these cysts. Other theories for
the growth of large pineal cysts have included cerebrospinal fluid
that flows through a direct connection with the third ventricle.35

With the advent of magnetic resonance imaging, these lesions
are now often identified incidentally and neurosurgical opinion is
obtained. Pineal cysts greater than 5 millimeters in size have
been identified radiographically in 1-4% of the general
population with rates being slightly higher in the young female
population.13,18 Magnetic resonance imaging reveal these pineal
cysts to be slightly higher intensity when compared to the
cerebrospinal fluid on T1-weighted and T2-weighted images. On
proton-density imaging these cysts were often much more
intense than the cerebrospinal fluid. A thin wall can often be
appreciated on T1-weighted images with rim enhancement when
the patient receives gadolinium.17

Our report describes a case of pineal apoplexy resulting in an
expanding pineal cyst. It is unclear in our case, if the initial
hemorrhage took place within a small pineal cyst that
subsequently enlarged or within the stroma of the pineal gland
itself. Evidence of previous hemorrhages within pineal cysts is
supported by xanthochromia and hemosiderin laden
macrophages within specimens from symptomatic patients.5 It is
possible that microhemorrhages are responsible for the
expansion of pineal cysts. In a series by Fain et al22 histological
evidence of prior hemorrhage in the form of hemosiderin
deposits within the glial layer in the predominantly perivascular
distribution was found in eight of 24 cases. We may also
speculate, that in our case, simple osmotic forces may have been
responsible for an increase in the lesion size.

Our case demonstrates how a pineal hemorrhage may be an
instigator for the development of a pineal cyst within a short
period. Knowledge of this mechanism of cyst growth is helpful
to surgeons when approaching patients with lesions in the pineal
region. Surgery done only for the sake of defining the
pathological nature of this type of lesion should be avoided. The
theory of pineal hemorrhage leading to the development of an
expanding pineal cyst is supported by this case, although other
models of pathogenesis may still exist.

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