

Atypical Presentation of an Apoplexy in a Pineal Cyst

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Abstract

Objectives: Apoplexy in a pineal cyst is a rare condition, with always headache or signs of increased intracranial pressure due to secondary hydrocephalus.

Methods: We describe a patient with recent intra-cystal haemorrhage with vertigo, but without any headache symptoms. We also conducted a literature review.

Results: This study focusses on a case report of a patient in whom apoplexy in the pineal cyst was detected as an incidental finding. Magnetic Resonance Imaging showed a recent haemorrhage within a 15 × 10 × 8 mm cyst of the pineal gland, without changes during follow-up. Clinically, however, the patient did not have any typical symptoms of headache or deterioration in the level of consciousness.

Conclusion: Pineal apoplexy can also present as an asymptomatic incidental finding, or as the cause of transient vertigo and eye-movement disorder.

Keywords: Pineal cyst; Pineal gland; Pineal apoplexy; Haemorrhage; Headache

Introduction

Pineal cysts are often found incidentally on cranial magnetic resonance (MR) examination. The majority are asymptomatic, but neurological symptoms can occur in large cysts (>5 mm), [1,2] and after intra-cystic haemorrhage (so-called pineal apoplexy) which has only been described a few times in the literature, almost always presenting with severe headache [2]. Here we describe a pineal apoplexy in a patient who presented with vertigo and transient eye movement abnormalities, but without headache. Magnetic resonance imaging was performed at presentation and during follow-up. We also conducted a literature review.

Methods

We describe a patient with recent intra-cystal haemorrhage with vertigo, but without any headache symptoms. Magnetic resonance imaging was performed at presentation and during follow-up. We also conducted a literature review.

Case History

Case presentation

A 47 year old male patient, with no previous medical history presented with acute vertigo, nausea and vomiting, aggravating when turning around, sitting or standing up. There was no accompanying headache and he also had not suffered from any headaches in the past years. Upon examination, blood pressure was 170/112 mmHg (which normalized spontaneously to 140/82 mmHg within one day) and he had a heart rate of 72/min. The EKG was normal. Neurological examination revealed a first degree horizontal nystagmus and a limited upward gaze. The vestibulo-ocular reflex was difficult to determine, as the patient felt very sick during the examination and had to vomit on all head-movements. Cerebral CT and MR scans showed no parenchymal lesions, but the MR scan did show a 15 × 10 × 8 mm cyst of the pineal gland with a fluid-fluid interface, suggestive of a recent haemorrhage. There was no mass effect on the surrounding tissue (Figure 1).

Case follow-up

Follow-up MR scans three days and two months later did not

show new lesions or any changes in cyst size or fluid-fluid interface. The pineal abnormality was interpreted as a co-incidental finding, based on the physical findings (first degree nystagmus), the fact that the patient quickly became symptom free including normalization of

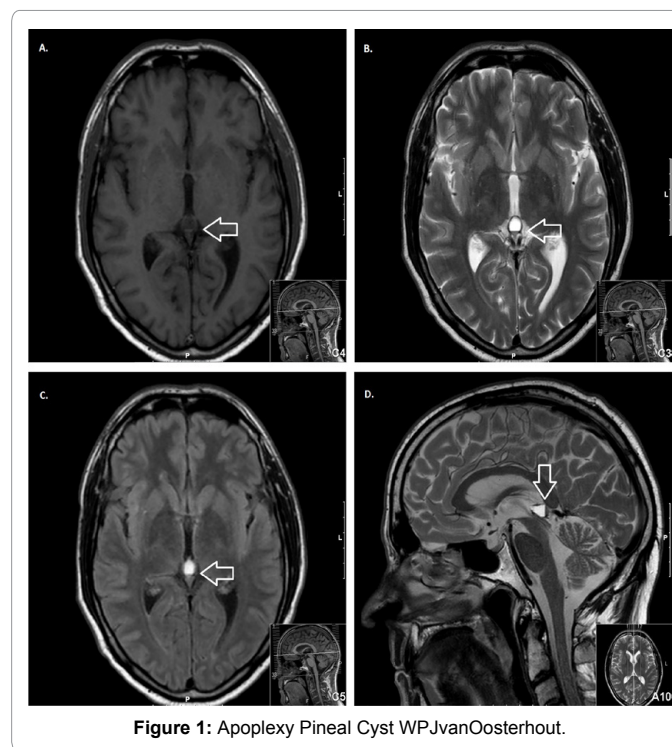


Figure 1: Apoplexy Pineal Cyst WPJvanOosterhout.

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eye movements and the lack of vascular risk factors. The diagnosis was an acute peripheral vestibular syndrome, most probably a vestibular neuritis. Neurosurgical intervention on the pineal cyst was not necessary in this case.

Discussion

Epidemiology

Pineal cysts are benign intracranial structures which can be observed at all ages but which are mostly found in women between the ages of 20 and 30 years [3,4]. In MR imaging studies, the prevalence of cysts larger than 5 mm is estimated to be approximately 4% [4]. Data from autopsy series have suggested prevalence rates of 21-41% for pineal cysts of all sizes (also including <5 mm) [5]. Most cysts remain stable in size over time, a minority regress, and seldom cyst size increases, mainly in children [6]. Due to the widespread use of MR, the prevalence of these incidentalomas is expected to increase, although cysts <5 mm in diameter are more difficult to detect on conventional 3T MR used in clinical practice [5,7].

Pathogenesis

How pineal cyst develop is unknown, and several theories have been proposed: 1) congenital dysembryogenetic origin due to lack of obliteration of the cavum pineale; 2) congenital remnant of the embryological diverticulum that develops as an outgrowth from the floor of the third ventricle and form the pineal gland; 3) degeneration of pineal cells; and 4) ischemic necrosis with cyst formation. Finally, pineal cysts could be seen as a normal variant since their prevalence is high [2].

Imaging of pineal cysts

A consistent number of pineal cysts is being diagnosed using a CT scan, as this usually is the first imaging modality of choice in patients with a head injury. Cysts are hypodense, round-shaped lesions in the pineal region, which in 30% have slight hyperdensities within the cyst or on its wall, corresponding to haemorrhage or calcifications [8,9]. MR is the preferable standard for both cyst assessment and follow-up, and usually show well-circumscribed, round-shaped lesions. The wall is thin and usually shows heterogeneous enhancement after gadolinium. Uniform enhancement during delayed gadolinium imaging can mimic a solid neoplasm. Characteristically, the intra-cystic content is iso-intense to CSF on both T1 and T2 sequences. Atypical pineal cysts, with aberrant enhancement or intra-cystic septations give rise to a differential diagnosis with cystic gliomas and pineal parenchymal tumours. These intra-cystic septations, however, sometimes are seen in pineal cysts when high-resolution MR is used [1].

Cyst size and clinical symptoms

Cyst with diameters of over 10-15 mm have been reported more likely to be linked to neurologic signs and symptoms, compared to smaller cysts [7,10,11]. However, correlation between cyst size and neurological symptoms is not considered to be rule. In 20-50% of asymptomatic individuals with a pineal cyst, diameters were found larger than 10 mm [1,12,13].

Rarely, pineal cysts increase in size and cause neurological symptoms, consisting of paroxysmal headache and vertical gaze palsy, also known as Parinaud syndrome; chronic headache, gaze paresis, papilledema and occlusive hydrocephalus, due to blockage of the aqueduct, obstruction of the vein of Galen, or compression of the collicular plate [1]. The increase in size could be due to coalescence

of small colloid cysts into one larger cyst or due to hormonal effects through pregnancy or the ovulatory cycle [2].

Pineal apoplexy is another – very rare – cause for symptoms ranging from mild to sudden death. Unfortunately, no data on the risk of intra-cystic haemorrhage are available [14]. It can present with acute or chronic headache, nausea, vomiting, syncope, ataxia, visual field defects, (vertical) gaze paresis and even acute death. In a recent case-series, 28/31 (90%) cases presented with headache, making this the most commonly reported symptom in pineal apoplexy [2]. In exceptional cases, a fluid-fluid level can be observed in asymptomatic pineal cysts secondary to a clinically silent apoplexy [1].

Treatment

Consensus is that asymptomatic cysts (incidentalomas) do not require any surgical treatment or routine follow-up in adults. In children, especially during puberty, some recommend to perform clinical and imaging follow-up [1,15,16]. Since symptoms of symptomatic pineal cysts are caused by secondary hydrocephalus or venous intracranial hypertension, surgical treatment is recommended. Apoplexy in a pineal cysts is almost always symptomatic and due to the expanding pineal mass. Neurosurgical interventions are aimed at relieving direct pressure on the tectal plate and secondary hydrocephalus, usually with a combination of a trans-ventricular marsupialization technique and third ventriculostomy [1].

The patient described here did not suffer from headaches at all. Thus, the pineal apoplexy could either be regarded as an asymptomatic incidental finding (which has not been described in literature so far), or as the cause of transient vertigo and eye-movement disorder, which has also never been described previously.

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