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Pineal Cyst Apoplexy: A Rare Complication of Common Entity

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Pineal cysts (PCs) are often encountered as incidental findings in intracranial images. The vast majority of cysts are normally asymptomatic and clinically benign. Bleeding into the cysts, which leads to neurological symptoms and signs, is considered to be quite rare. The authors illustrate a newly identified complication of PC in a 56-year-old woman who characterized by headache of sudden onset and vomiting. MRI disclosed a small hemorrhagic PC without narrowing of the cerebral aqueduct. The patient was managed conservatively without any surgical interventions, and she remained symptom-free over a period of 15-year follow-up. The description of this case adds to the limited literature on the series in which nonsurgical treatments had a role in the care for patients with PC complicated by intracystic hemorrhage.

Key Words Apoplexy; Pineal cyst; Pineal gland.

INTRODUCTION

Extensive use of MRI increases the discovery rate of pineal cysts (PCs) in the clinical neurology. In adults, the prevalence of cysts is estimated to be 1.1−4.3% [1]. In terms of therapy, there is no accepted indication for follow up or criteria for intervention. Only a minority of patient with PCs requires specific treatment for their symptoms. A variety of approaches to symptomatic PCs are described in the limited literature [2-4]. Rarely, the patient of PCs was complicated by intracystic bleeding, which manifested with sudden onset of neurological symptoms and signs [5].

This syndrome of apoplexy stands for acute hemorrhage occurring into the pathology in the pineal region, most commonly into a PC. To date, however, only small series or individual case reports of PC apoplexy have been published, and our understanding of the disease is still incomplete [6,7]. For this reason, the proper management of the apoplectic PC remains a controversial issue in the neurological surgery. The author reports a case of small PC which bled out spontaneously, and describes the diagnostic characteristics and treatment options for this rare phenomenon.

CASE REPORT

A 56-year-old woman who suffered from headache and vomiting was admitted as an emergent case. The sudden onset of severe pain in the occipital area started 2 weeks before admission and got worse over 2 days. On arriving, the patient was alert and oriented. She had a history of uncontrolled hypertension, and her presenting blood pressure was 170/90 mm Hg. Intravenous hydralazine to lower blood pressure was administered. When the patient was referred for neurological opinion, she denied photophobia, sensory change or motor weakness. A further evaluation was notable for a normal neurologic examination including cranial nerves and no neck stiffness. No abnormality of ocular motility was observed. The laboratory results for coagulation profiles and tumor markers were within normal limits.

CT showed an 11 mm-sized, noncalcified, and faintly-enhancing mass in the pineal region without ventricular enlargement (Fig. 1). No abnormal finding in the cerebral vasculatures was detected on the reconstructed three-dimensional CT angiography. The pineal lesion appeared at MR images as ovoid, thin-walled, and well-defined cyst. The cyst was hypointense on T1-weighted and hyperintense on T2-weighted sequence. There was a blood-fluid interface within the cyst on

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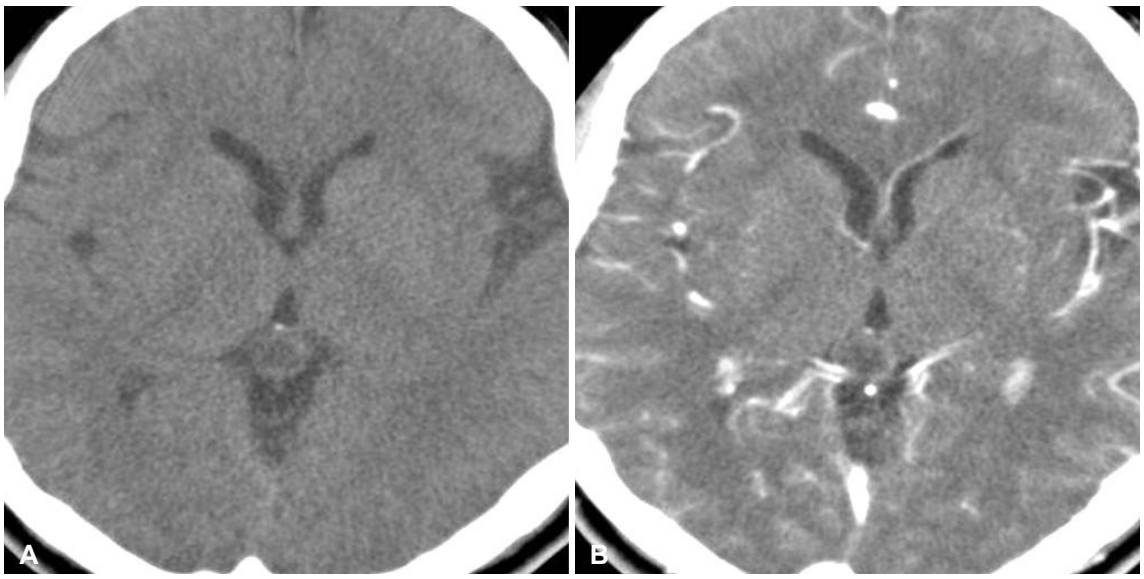

Fig. 1. CT scans of pineal cyst apoplexy. The ventricle width is normal (A) and the cyst is mildly enhanced at the periphery (B).

gradient echo sequence (Fig. 2A-E). No tumor or vascular malformation in the pineal region was visualized on enhanced MR studies. An imaging diagnosis of intracystic hemorrhage of PC was made by staff neuroradiologists without the need for clinical information about patient's history and findings.

Since it was unlikely that the cyst would compress the cerebral aqueduct, the patient was scheduled for managing the PC apoplexy conservatively. Her symptoms had completely resolved with use of pain medication and antiemetic drugs. The patient was discharged home at the ninth day of hospitalization with normalized blood pressure. The close observation and regular follow-up with MRI were conducted. At the 6-month follow-up, MR scans revealed the stabilized cyst with resolution of blood products. No recurrent mass or other complication was detected on MR series taken 15 years later after the apoplexy (Fig. 2F). The patient was still clinically asymptomatic at the last outpatient visit.

The IRB exempted informed consent due to its retrospective nature and minimal risk for harm to the patient, and this report was conducted according to the guidelines of the Declaration of Helsinki for biomedical research.

DISCUSSION

The PC is a frequently identified cyst in the pineal body. Histologically, the cysts consist of an inner glial layer, a middle layer of pineal tissue, and an outer fibrous capsule. Its incidence is higher in females and adults, and the size is not related to the patient's age or gender [8]. A follow-up MRI study observed that PCs usually remain stable without significant dimensional and natural changes [9]. On these grounds, only a few PCs might enlarge in time, and then make them of neurological significance. Large cysts typically exert the mass effect on the cerebral aqueduct, surrounding venous structures, and the dorsal midbrain. Symptomatic PCs were classified into three distinct syndromes: 1) paroxysmal headache and gaze palsy; 2) chronic headache, papilledema, gaze paresis, hydrocephalus; and 3) pineal apoplexy with acute hydrocephalus [10]. Of these neurological features, apoplexy is the rarest but most dangerous form.

The most common symptoms of pineal apoplexy were headaches, followed by gaze paresis and visual deficits in the analysis of the affected [11]. The bleeding into PCs brought on or worsened headaches in almost all cases reported at the time of their presentations [12]. Since the current patient had no evidence of hydrocephalus in imaging tests, the author considered the headache to be the result of the apoplectic event. Primary headaches were also excluded following a thorough history and focused neurological examination. In addition, infectious and vascular diseases that cause severe headache were not evident in thin-sliced CT and MR images for this case. Some degree of abnormalities on ocular examination in these patients could be either from direct compression on the midbrain or secondary to obstructive hydrocephalus. Additional neurological manifestations included nausea, vomiting, ataxia, dizziness, syncope, hemiparesis, impotence, depression, insomnia, deafness, and seizure [13-15]. The duration of symptoms and signs before hospital admission ranged from a few days to months or even years [16]. In rare instances, the clinical pattern was of sudden collapse, leading to death and diagnosis of apoplexy being confirmed at autopsy [17].

The precipitating or concomitant factors for intracystic pineal hemorrhage have been proposed in the previous studies.

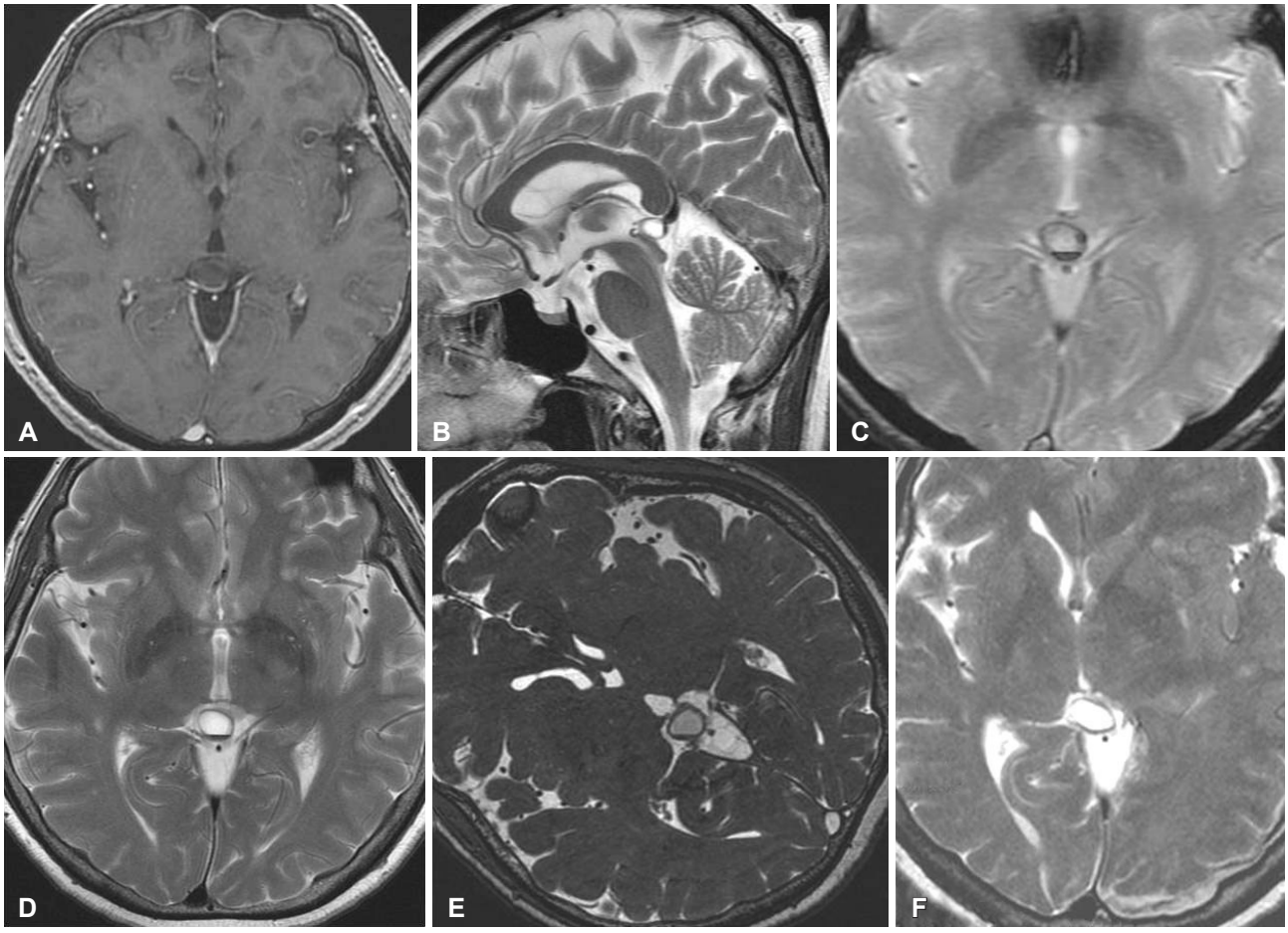

Fig. 2. MRI for pineal cyst (PC) apoplexy. A: Initial examinations display the PC measuring 11×9×7 mm with incomplete rim enhancement. B: The dorsal midbrain remains intact, and the cerebral aqueduct is patent. C: The sign of hemorrhage is visible on gradient echo sequence image. D, E: MR series taken from lying supine and inclined at 45 degrees depict fluid-blood level within the cyst suggesting intracystic hemorrhage. F: At the 15-year follow-up post-apoplexy, the scan reveals the stable cyst with hemosiderin stains in the pineal body.

Firstly, a few events of apoplexy have occurred in patients who receiving antiplatelet or anticoagulant therapy [18,19]. Secondly, the studies reported the interesting cases in which an anatomical cause for apoplexy was vascular malformation in the wall of the cyst [20]. Thirdly, based on this case, and backed by the literature, the author cannot ignore the effect of hypertension as a medically correctable etiology for acute hemorrhage in the underlying PCs [21]. Fourthly, the pathological study demonstrated that one third of patients with PCs had foci of bleeding in the form of hemosiderin pigments within the glial and pineal layers in surgical specimens [22]. A similar number of cases showed hyalinization of their fine blood vessels [23]. Therefore, these degenerative changes in the structure could be a factor favoring the apoplectic hemorrhage within the long-standing PCs.

On review of the pertinent literature, the maximal diameter of the hemorrhagic PCs was measured from 15 mm to 36 mm [5,7,11]. The cyst of this patient is of the smallest size when compared to the previous reported cases [3,4]. Extent and severity of apoplexy can vary from minimal xanthochromia to massive subarachnoid or intraventricular hemorrhage [16,18]. MRI feature of PCs might be variable, ranging from that of a simple cyst to a complex mass associated with bleeding, calcification, enhancement, or hydrocephalus [24,25]. A hallmark on MR scan of PC apoplexy is thin-walled, evenly enhanced cyst having the hemorrhage products, without accompanying soft-tissue mass as it was in our case [26,27]. In head CT, the dependent fluid level may not be detected in hemorrhagic PCs like this illustration. For these cases, performing MR scans with head tilted is useful to confirm the existence of small amount of blood inside the cysts. In addition, CT and MR scans with angiography can assist the clinician to rule out the underlying vascular processes in the patient presenting with pineal hemorrhage [28].

On routine MR images for this location, a cystic configuration is commonly related with non-neoplastic lesions rather than with a tumor, but studies do not allow cystic tumors to be differentiated from PCs with clarity. Arachnoid cysts and PCs

have MR signal intensities similar to cerebrospinal fluid. But, the findings of non-calcification, no contrast enhancement, and signal suppression on fluid-attenuated inversion-recovery image are specific to arachnoid cysts [13,29]. Additional differential diagnostics for cystic masses include teratomas, epidermoid and dermoid tumors, and cysticercosis. Apoplectic hemorrhage can happen very rarely in these uncommon circumstances. Besides, the radiographic appearances of these benign cysts are sufficiently unique so that they can be discriminated from other more common pineal neoplasms [22,30]. A heterogeneous group with pathologies, such as pineoblastomas, astrocytomas, meningiomas, and pineocytomas, may also feature a cystic nature on MRI, but they do not typically show hemorrhage within the masses [7,24,25,27]. Characteristically, pineal meningiomas are dural-based lesions which involve the velum interpositum, tela choroidae or falcotentorial junction. The sign of dural tail is a reliable criterion for the differential diagnosis of cystic meningiomas from PCs [26]. Pineocytomas may mimic PCs, however, these are described as predominantly solid, uniformly enhancing masses [31]. In general, it is not always possible to distinguish a non-neoplastic cyst from pineal gliomas with cysts only by radiographic findings [32,33]. Finally, germ cell tumors can be also the differential diagnosis of a hemorrhagic pineal lesion. An imaging analysis compared with blood and cerebrospinal fluid level of tumor markers can narrow the spectrum of diagnosis for hemorrhagic pineal pathologies.

Surgical intervention is primarily required for patients with hemorrhagic PCs, in particular for the subset with symptoms of hydrocephalus and brainstem compression. In patients with neurologically significant symptoms, aspiration, shunting, or resection of the cyst has been introduced as a successful procedure [11,18,27]. Excellent outcomes were obtained even in apoplectic patients presenting altered mentality, if the prompt management was taken. However, it is difficult to determine the best treatment modality for an individual with apoplexy, as published data only reported shorter follow-up results [13,19,34]. Microscopic or endoscopic cystectomy relieved the hydrocephalus by opening the aqueduct, thus avoiding the shortcomings of shunting and aspiration for the PCs, and eliminating the risk of recurrence. On gross examination, hemorrhagic PCs were smooth surfaced, soft, and opaque to yellow or chocolate-brown. The cyst contents have been reported to be clear yellow, cloudy milky, or coffee to amber in the case of older hemorrhages [10].

Owing to the rarity of cases with PC apoplexy, very little is known about the conservative treatment for them. To the best of our knowledge, this patient with apoplexy is the sixth case that was managed nonsurgically in the English literature [12,35]. Given the lack of hydrocephalus, clinical status of the patient, and MR appearance of the cyst, we decided to observe the patient closely instead of doing the surgical interventions. In this patient, a favorable course with conservative management lasted for 15 years in both clinically and radiographically. However, unfortunately, this kind of management for hemorrhagic PCs still lacks sufficient evidence. Besides, the disadvantage of the treatment is that there is risk for repeated bleeding or developing an expanding cyst [8,36]. In contrast, several investigators mentioned that spontaneous involution of the cysts was attributed to pineal apoplexy [16,37]. Nonetheless, the clinician should be stay alert in serial MR scanning even after the intracystic hemorrhage has resolved. In practice in the neurosurgery, the curative pineal surgery should be reserved for the cases showing definitive abnormalities on the clinical and imaging examination during the observation period.

In summary, the presented cyst had three special features: one is the smallest lesion which the hemorrhage took place into the cyst; the second is its conservative management without operative interventions; the third is longest clinical and radiological follow-up period after the apoplexy.

Conflicts of Interest

The authors have no potential conflicts of interest.

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None

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