Pineal cyst apoplexy: report of an unusual case managed conservatively

Correspondence address: Selim Ayhan, MD, Hacettepe University School of Medicine, Department of Neurosurgery, 06100, Sihhiye, Altindag, Ankara, Turkey, phone +90 312 305 1715, fax +90 312 311 1131, e-mail: selim_ayhan@yahoo.com

Received: 15.02.2011; accepted: 31.05.2011

Abstract

Pineal cyst apoplexy is a very rare entity with previously reported symptoms of severe frontal or occipital headache, gaze paresis and visual field defects, nausea or vomiting, syncope, ataxia, hearing loss and sudden death. The treatment options for symptomatic pineal cysts are observation, shunting, aspiration via stereotactic guidance or endoscopy, third ventriculostomy, ventriculocysternostomy, and/or surgical resection by craniotomy and microsurgery.

Here, the authors report an unusual case of a 28-year-old male patient with pineal cyst apoplexy, presenting with headache, insomnia, and sexual dysfunction symptoms who is being managed conservatively and observed for two years by an academic tertiary care unit.

Key words: apoplexy, melatonin, observation, pineal cyst, pineal gland.

Introduction

Pineal cysts are benign intracranial lesions. They may be congenital, degenerative in nature or may represent a normal anatomical variant. Its prevalence in magnetic resonance imaging (MRI) studies is reported to range from 1.1% to 4.3%, and they can be found in up to 40% of individuals on autopsy studies [1-5].

Although almost asymptomatic, these cysts rarely cause symptoms as a result of increased intracranial pressure and/or compression of the tectal plate [2]. According to Wisoff and Epstein, paroxysmal headache with gaze paresis, chronic headache, gaze paresis, papilloedema and hydrocephalus, and pineal apoplexy with acute hydrocephalus can occur [6]. Of note, pineal apoplexy, first reported as haemorrhage into a pineal cyst by Apuzzo...
et al. in 1976, is the least common but most dangerous symptom, which could be a medical emergency [2,3,7]. Here, the authors report an unusual case of a patient with pineal cyst apoplexy, presenting with headache, insomnia, and sexual dysfunction who is being managed conservatively and followed up for two years by an academic tertiary care unit.

**Case report**

A previously healthy 28-year-old male patient was admitted with complaints of worsening of ongoing headache which had been increased over a period of a month, insomnia and sexual dysfunction. Neurological examination revealed no focal abnormality and laboratory investigations were within normal limits. Cranial MRI showed an ovoid-shaped cystic lesion in the pineal region, measuring 16 mm × 12 mm, without compression of the aqueduct (Fig. 1A). There was a fluid-fluid level inside the cyst which was an indicator of acute haemorrhage (Figs. 1B-D). Gadolinium-enhanced studies demonstrated contrast enhancement of the cyst wall. There was not any abnormal vessel around the lesion. Upon the MRI findings, the diagnosis was consistent with pineal cyst apoplexy. Because of lack of any evidence of hydrocephalus, the health status of the patient (young, neurologically intact, no significant medical history), and radiological appearance of the cyst (intact wall, without any vascular pathology), the decision was made to follow the patient conservatively. Regular neurological and radiological follow-up examinations using cranial MRI scans were performed every 6 months. After 2 years, the patient was doing well without any complaint or neurological deficit. The last MRI scans revealed stable cyst size with resolution of the intracystic haemorrhage (Fig. 1E).

**Discussion**

Starting from the MRI era, the identification and hence frequency of pineal cysts have been increasing and the prevalence rates range from 1.1 to 4.3% [1-3]. These lesions, however, rarely cause symptoms which require

![Fig. 1](image-url)
The symptoms of pineal apoplexy can be classified as paroxysmal or chronic, ranging from a few days to months or even years [2,3]. The most frequent symptoms are severe frontal or occipital headache, gaze paresis and visual field defects, nausea or vomiting, syncope and ataxia, which are due to accompanying hydrocephalus [2,3]. Hearing loss or sudden death have also been reported [13,14].

Radiological appearance requires a fluid-fluid level on the MRI scan, representing the layering of blood products caused by acute haemorrhage, which is known as a hallmark of pineal cyst apoplexy with intracystic bleeding [2,3]. Especially after intracystic haemorrhage or in the occurrence of obstructive hydrocephalus, the radiological diagnosis of the pineal cysts get more complicated and one should differentiate these lesions from pineal region tumours, such as pineoblastomas, teratomas or pilocytic astrocytomas, which could also present as large cysts [3,15]. It was shown that a thin, smooth cyst wall less than 2 mm thick was specific to pineal cyst [3,16].

The treatment options for symptomatic pineal cysts are observation, shunting, aspiration via stereotactic guidance or endoscopy, third ventriculostomy, ventriculocysternostomy, and/or surgical resection by craniotomy and microsurgery [2,3,12,17,18]. The evidence for conservative management of pineal apoplexy is insufficient but neurologically intact patients should not be surgically treated. Possible symptoms should be explained and one should apply to the medical specialist in the event of development of such symptoms during the observation period [3]. Imaging studies should be done for decision making at admission time. An MRI scan is essential for both diagnosis and follow-up [2]. In terms of surgical treatment, because of the inadequate follow-up times, it is hard to determine the best approach for managing pineal cyst apoplexy, although it is suggested that microsurgical resection or endoscopic treatment is superior to shunting or third ventriculostomy [2,3]. Of note, close and lengthy follow-up should be done in any case [2].

Here, the pineal cyst apoplexy case that the authors present has two unique features: one is the accompanying symptoms, i.e. insomnia and sexual dysfunction; the second is its conservative treatment, being the third of its kind in the literature [17,18]. Due to the lack of any evidence of hydrocephalus in the radiological work-up, the authors considered the headache to be the result of the apoplectic event. However, the other accompanying symptoms, insomnia and sexual dysfunction, have not been previously reported along with pineal cyst apoplexy. This may be due to either the camouflage of the symptoms by acute hydrocephalus, removal of the symptoms by emergency surgery, or other reasons yet to be identified. Hypothetically, the aforementioned symptoms could be because of pineal gland dysfunction due to the apoplectic event, resembling endogenous melatonin level decline. Melatonin is known as one of the potential substances affecting initiation and maintenance of sleep [19]. Although there are several ways to determine the endogenous melatonin level, the authors were not able to measure it in this patient due to technical insufficiency at the time of diagnosis. Secondly, this is the third case in the English literature which was managed conservatively without any neurological intervention [17,18]. Because of the patient's characteristics, symptomatology and the radiological appearance of the cyst, the decision was made to follow up the patient conservatively instead of taking the risk of a major neurosurgical procedure [20].

In conclusion, although pineal cysts are quite commonly encountered lesions, pineal apoplexy is very rare. The patient with pineal cyst apoplexy presented in this report had exclusive symptoms of insomnia and sexual dysfunction and was treated conservatively. As the symptoms and consequences of pineal cysts vary, each patient should be assessed individually and treated accordingly.
Disclosure

Authors report no conflict of interest.

References
