JCN Open Access LETTER TO THE EDITOR

pISSN 1738-6586 / eISSN 2005-5013 / J Clin Neurol 2022;18(5):594-596 / https://doi.org/10.3988/jcn.2022.18.5.594

Pineal Apoplexy Presenting With Recurrent Migraine-Like Headache and Transitory Neurological Dysfunction During Pregnancy

Gonçalo Cabral[a]
Marlene Saraiva[a]
Inês Freire[b]
Inês Gil[b]

Departments of [a]Neurology and [b]Neurradiology,
Hospital Egas Moniz,
Centro Hospitalar Lisboa Ocidental,
Lisboa, Portugal

Dear Editor,

Headache is a common complaint in pregnancy. Most cases are benign, but the prevalence of secondary headache reportedly ranges from 14.3% to 52.6% during pregnancy.[1,2]

Pineal cysts are clinically benign entities that are usually asymptomatic, and so normally found incidentally in neuroimaging studies.[3,4] Pineal apoplexy (PA) is a rare clinical condition that normally results from acute intracystic pineal hemorrhage, and it can manifest as headache or signs of increased intracranial pressure due to secondary hydrocephalus.[3-6] We report an unusual case of PA during pregnancy presenting with migraine-like headache along with focal neurological deficits.

A multiparous melanodermic 41-year-old female who was 32 weeks pregnant and had a relevant record of hypertension medicated with methyldopa (250 mg three times daily) and sickle-cell disease (last vaso-occlusive crisis was at the age of 12 years) presented to the emergency department with an acute right pulsatile hemicrania headache with a maximum intensity (pain scale score of 7 out of 10) at 3 hours after onset and associated with visual disturbance. She had no history of preeclampsia. She had a history of episodic migraine without aura, and the index headache had similar characteristics but was more intense. The headache did not worsen with recumbency or the Valsalva maneuver. She had no associated fever, chills, or neck pain. Her blood pressure was 146/91 mm Hg. The findings of a neurological examination were normal. Laboratory results were normal, including an sFlt-1/PlGF ratio of <38 (no indication of preeclampsia). She had mild proteinuria (30 mg/dL). She was admitted for surveillance and blood pressure control.

The headache and visual disturbance improved on the first day of surveillance following blood pressure reduction using methyldopa and nifedipine. However, the patient woke up on the second day with the same headache and blurred vision, and felt a heaviness in the left hemibody. A neurological examination indicated that the patient was alert and oriented but had slight psychomotor slowing. She had brisk reflexes in the left arm and an ipsilateral Babinski sign with a slight left pronator drift, but muscle strength was preserved. She also exhibited a decreased response to pinprick on the left side. There was no oculomotor nerve palsy or papilledema. MRI revealed a cystic pineal gland with a maximum diameter of 1.2 cm, which was hyperintense in T2-weighted/fluid-attenuated inversion recovery (FLAIR) images, without significant locoregional mass effect or associated acute hydrocephalus. This lesion had an internal blood–fluid level that was slightly hyperintense in T1-weighted (Fig. 1C) and hypointense in T2-weighted/FLAIR (Fig. 1A and B) MRI images, compatible with early subacute hemorrhage. In a susceptibility-weighted angiography (SWAN) image (Fig. 1D), the hematic component was demonstrated by an area of a signal void (blooming effect). These features were compatible with the diagnosis of PA in a previously cystic gland.

Received February 25, 2022
Revised May 3, 2022
Accepted May 4, 2022

Correspondence
Gonçalo Cabral, MD
Department of Neurology,
Hospital Egas Moniz,
Centro Hospitalar Lisboa Ocidental,
Rua da Junqueira 126,
1349-019 Lisboa, Portugal
Tel +351-927-376-804
Fax +351-210-431-589
E-mail goncalo2010286@gmail.com

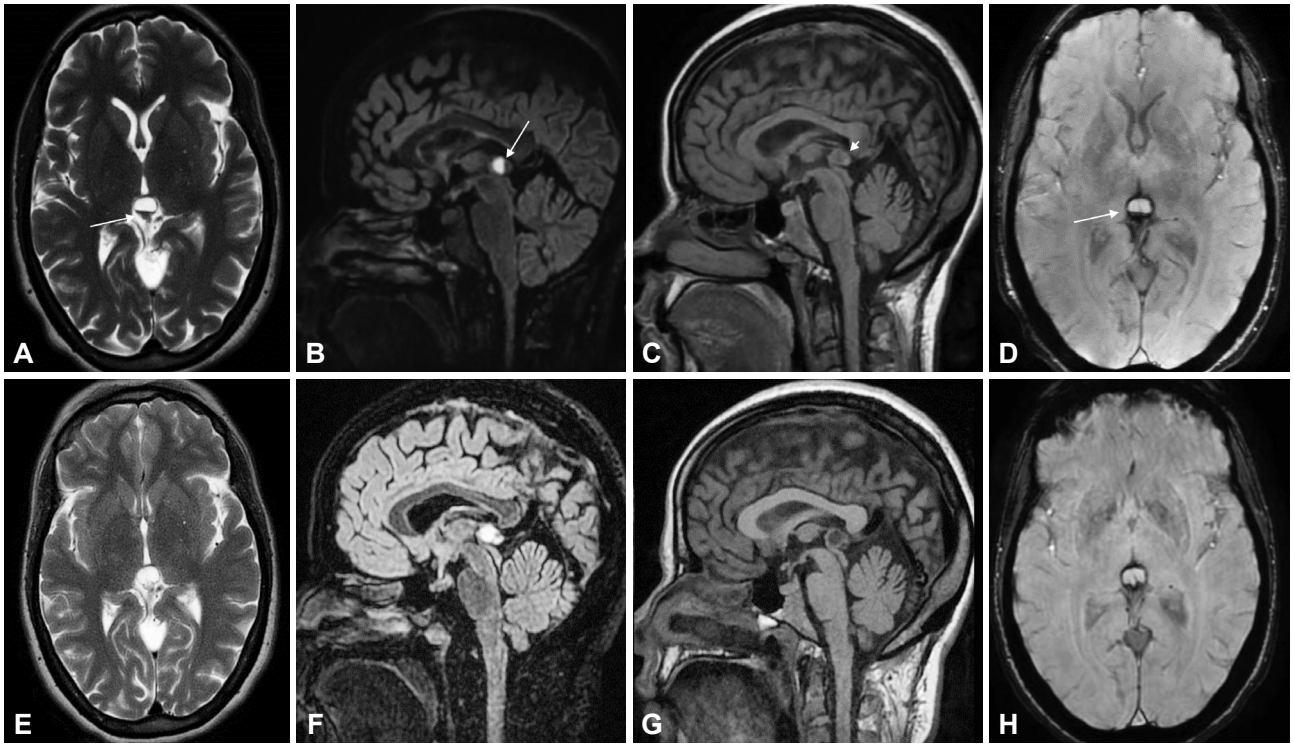

Fig. 1. Initial MRI revealed a blood–fluid level in the pineal cyst that was hypointense in T2-weighted/FLAIR images (A, B; arrows) and slightly hyperintense in a T1-weighted image (C, short arrow). There was a blooming hemorrhage in the cyst wall and a blood–fluid level in the pineal cyst in a magnetic susceptibility image (D, arrow). Follow-up MRI performed 9 months later revealed reabsorption of the previously visualized blood–fluid level in the cystic pineal gland, which was hyperintense in T2-weighted and FLAIR images (E and F) and hypointense in a T1-weighted image (G). A susceptibility-weighted angiography image (H) shows voids persisting in the cyst wall without evidence of a blood–fluid level. FLAIR, fluid-attenuated inversion recovery.

Conservative treatment was applied, which resulted in an improvement in symptoms within 24 h. She was discharged on methyldopa and nifedipine. The patient remained asymptomatic after 1 year of follow-up. Follow-up MRI performed at a 9-month follow-up revealed a cystic pineal gland of identical dimensions and benign characteristics with total resorption of the previously identified hematic content (Fig. 1E-G). In a SWAN image, hemosiderin deposits persisted in the cyst wall (Fig. 1H).

We have described a case of a pregnant female who had recurrent migraine-like headache with visual disturbance and transitory focal neurological deficits. The main conditions considered were preeclampsia, posterior reversible leukoencephalopathy syndrome, and reversible cerebral vasoconstriction syndrome. It was also important to rule out cerebral venous thrombosis, subarachnoid hemorrhage, ischemic stroke, and pituitary apoplexy.[1,2] The absence of headache with cervical irradiation or cervical trauma essentially excluded arterial dissection, and also made idiopathic intracranial hypertension less likely.[1,2]

It was essential to perform brain imaging this patient, which revealed PA in an intrapineal cyst. PA is a rare complication that it always diagnosed by exclusion. The symptoms can range from mild manifestations to sudden death.[7] The most-common symptom of PA is headache, although it has a highly nonspecific and variable presentation, and can manifest as a worsened headache in people with previous primary headaches,[4] as in our case report. Karthik et al.[8] similarly reported a PA case presenting with migraine-like headache. Visual deficits and gaze paresis are often identified, which are either due to direct compression of the midbrain or secondary to obstructive hydrocephalus.[3,6] Although rare, transient neurological deficits have been reported. Additional manifestations include nausea, vomiting, ataxia, dizziness, hemiparesis, and seizures.[4]

Strictly unilateral headaches and lateralized neurological deficits are not typical for midline lesions.[9] Similar clinical symptoms have been described previously in a case of pituitary apoplexy, also without neurosurgical intervention.[10] In the present report, we hypothesized that the acute headache and transient neurological dysfunction resulted from an acute bleeding/apoplectic event without signs of hydrocephalus or venous hypertension. Apoplexy could result from an increase in the size of the pineal cyst due to hormonal effects associated with pregnancy. In addition, the presence of hypertension could increase the risk of hemorrhage. Since the symptoms of

PA are often indicative of hydrocephalus or venous intracranial hypertension, neurosurgical intervention is essential.[3] The evidence for conservative management of PA is insufficient, but neurologically intact patients without signs of compression or hydrocephalus should not be treated surgically, with instead regular neurological and radiological follow-up applied to those patients.[3,7]

To our knowledge, this is the first case of PA reported in pregnancy presenting with unilateral recurrent headache and neurological focal deficits with spontaneous resolution, and which was managed successfully using conservative treatment. This case highlights that PA should be considered as a possible cause of headache and neurological transitory dysfunction during pregnancy.

Ethics Statement

Informed consent was obtained from the patient.

Availability of Data and Material

Data sharing does not apply to this article as no data sets were generated or analyzed during the study.

ORCID iDs

Gonçalo Cabral https://orcid.org/0000-0002-3517-8365
Marlene Saraiva https://orcid.org/0000-0003-2804-4112
Inês Gil https://orcid.org/0000-0003-4935-5921

Author Contributions

Conceptualization: Gonçalo Cabral. Data curation: Gonçalo Cabral. Formal analysis: Gonçalo Cabral. Funding acquisition: Gonçalo Cabral, Inês Freire. Investigation: all authors. Methodology: Gonçalo Cabral, Marlene Saraiva. Project administration: Gonçalo Cabral. Resources: Gonçalo Cabral, Inês Freire. Software: Gonçalo Cabral, Inês Freire. Supervision: Marlene Saraiva, Inês Gil. Validation: Marlene Saraiva, Inês Gil. Visualization: Marlene Saraiva, Inês Gil. Writing—original draft: all authors. Writing—review & editing: all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Funding Statement

None

REFERENCES

1. Robbins MS, Farmakidis C, Dayal AK, Lipton RB. Acute headache diagnosis in pregnant women: a hospital-based study. *Neurology* 2015;85:1024-1030.
2. Negro A, Delaruelle Z, Ivanova TA, Khan S, Ornello R, Raffaelli B, et al. Headache and pregnancy: a systematic review. *J Headache Pain* 2017;18:106.
3. van Oosterhout W, Haan J. Atypical presentation of an apoplexy in a pineal cyst. *J Clin Case Rep* 2015;S3:1-3.
4. Kim E, Kwon SM. Pineal cyst apoplexy: a rare complication of common entity. *Brain Tumor Res Treat* 2020;8:66-70.
5. Peres MF, Valença MM, Amaral FG, Cipolla-Neto J. Current understanding of pineal gland structure and function in headache. *Cephalalgia* 2019;39:1700-1709.
6. Patel AJ, Fuller GN, Wildrick DM, Sawaya R. Pineal cyst apoplexy: case report and review of the literature. *Neurosurgery* 2005;57:E1066.
7. Asundi A, Tampieri D, Melançon D, Del Maestro R, Petrecca K, Cortes MD. Pineal apoplexy: imaging diagnosis and follow-up of three new cases. *Can J Neurol Sci* 2011;38:931-933.
8. Karthik DK, Khardenavis V, Kulkarni S, Deshpande A. Pineal gland apoplexy mimicking as migraine-like headache. *BMJ Case Rep* 2018;2018:bcr2018225187.
9. Peres MF, Zukerman E, Porto PP, Brandt RA. Headaches and pineal cyst: a (more than) coincidental relationship? *Headache* 2004;44:929-930.
10. Schoen JC, Campbell RL, Sadosty AT. Headache in pregnancy: an approach to emergency department evaluation and management. *West J Emerg Med* 2015;16:291-301.