Egyptian Journal of Neurosurgery

CASE REPORT Open Access

Cavernoma in a young adult: a case report

Hina Ali[1*], Samar Zaki[1], Ehsan Bari[2] and Sara Khowaja[3]

Abstract

Cavernomas or cavernous malformations of the central nervous system are acquired or hereditary vascular anomalies. Although they are present in 0.1–0.5% in the general population, they are usually asymptomatic and undetected until symptoms occur. We report a case of a healthy young adult, who presented with acute onset of dizziness, and intermittent episodes of loss of consciousness. MRI brain was suggestive of right insular cortex and right basal ganglia cavernoma. The patient later on developed seizures in the form of automatisms (continuous lip smacking). Surgical removal of the cavernoma was planned as in this case it was present in an eloquent area causing seizures. Seizure producing brain mapping and intraoperative electrocorticography was also performed which is rare in the region and offered by only few centres. The surgical resection of cavernoma was successfully performed, and patient reported improvement in symptoms during subsequent follow-ups.

Keywords Cavernoma, Cavernous malformation, Vascular malformation

Background

Cavernomas or cavernous malformations are large and deformed blood vessels that are gathered in clusters [1]. Cavernomas of the central nervous system are rare neurovascular malformations [2]. These can lead to intracerebral or intramedullary haemorrhage, causing significant morbidity [3]. They can present as an incidental finding or with a seizure or focal neurological deficit [3]. The symptoms can be attributed to mass effect or haemorrhage within or outside the lesion. With the development of imaging modalities such as magnetic resonance imaging (MRI), these are more frequently detected [2]. We describe a case of a young female with cavernoma who presented to primary care with the complaint of episodes of dizziness and loss of consciousness. In this report, the clinical, radiological features and treatment options of cavernoma are also discussed based on pertinent literature review.

Case presentation

A 19-year-old female with no known comorbids presented to the primary health care clinic with complaint of dizziness and two episodes of sudden loss of consciousness in the last few weeks. Each episode lasted for a few minutes. There were no associated jerky movements of the body, urine incontinence or up rolling of eyes during the episodes. No chest pain or palpitations were reported. She denied any motor or sensory weakness in the body. A review of systems was positive for fatigue.

She had no significant past medical, surgical, or family history, and she was not taking any regular medications. On examination, she was alert and well-oriented with no focal neurological deficits. The rest of the systemic examination was unremarkable. Relevant laboratory workup was done which showed iron deficiency anaemia (Hb 10.7 mg/dl and ferritin of 4.6). Echocardiogram and Holter monitoring were done which were unremarkable.

She was started on iron supplements and referred to neurologist for further evaluation of her symptoms. The neurology review was done, and patient was advised to have MRI brain and electroencephalogram (EEG). EEG was reported normal. MRI brain showed a lobulated

*Correspondence:
Hina Ali
hinaali.ahmed@aku.edu
[1] Department of Family Medicine, Aga Khan University Hospital, Karachi, Pakistan
[2] Section of Neurosurgery, Department of Surgery, Aga Khan University Hospital, Karachi, Pakistan
[3] School of Nursing & Midwifery, Aga Khan University Hospital, Karachi, Pakistan

abnormal signal intensity area in the right insular cortex and right basal ganglia (measuring approximately 24×12 mm) showing signal dropout on SWI sequence (as can be seen in Fig. 1). Findings were suggestive of cavernoma.

No acute infarct or cerebral venous sinus thrombosis was detected. After reviewing the MRI, the patient was started on antiepileptics by neurologist (lamotrigine and lacosamide). However, she had another episode of loss of consciousness, and she was brought to the emergency room. During the emergency visit, she was reviewed by neurosurgery. She was advised for watchful waiting vs. surgical resection of the cavernoma if her symptoms persisted.

After being discharged from emergency, she continued her antiepileptics and followed up regularly with her primary care physician and neurosurgeon. In the upcoming weeks, she developed complex focal seizures in the form of automatisms (continuous lip smacking) for which she followed neurosurgery. Sleep EEG was done which showed that syncopal episodes were drop attacks originating from cavernoma site.

Considering her young age and persistent symptoms despite being on antiepileptics, the patient and her family made an informed decision to undergo surgery. Neuronavigational-guided right temporal craniotomy and lesion resection was performed. Seizure producing brain mapping and intraoperative electrocorticography was performed which is offered in only few centres in the region. (Postoperative MRI shown in Fig. 2).

Over nine months of observation, the patient showed improvement in her symptoms, and there were no episodes of loss of consciousness or automatisms. She does however report occasional episodes of vertigo, generalized fatigue and headaches. She was started on oral lamotrigine 50 mg once daily preoperatively; however, she developed rash and itching, and it was switched to oral lacosamide preoperatively which was continued postoperatively. Currently, the patient is on lacosamide 100 mg twice daily. She was also started on tablet sertraline due to her mood swings which she developed after the diagnosis of cavernoma. She does report improvement in her mood symptoms after taking it. The patient is currently being followed by primary health care and the neurosurgeon with 6 monthly follow-up brain scans.

Discussion

Cavernomas are neurovascular malformations, affecting up to 0.1–0.5% of the population [2, 10]. They are closely arranged abnormal blood vessels without muscular and elastic layers and contain blood at different levels of thrombosis and organization [3]. There is typically no nervous tissue within the lesion. Men and women are equally affected, and most patients present in the third or fourth decade of life [1, 3]. Most of the lesions occur in the supratentorial region (80–92%) but they can be found in the entire central nervous system [6]. Cavernomas occur in two forms, a sporadic form usually having an isolated lesion and a familial form (20%) characterized by an autosomal mode of inheritance and multiple lesions [4, 10].

They often have a benign natural history (only requiring regular clinic and radiological follow-ups), but can sometimes lead to intracerebral and intramedullary

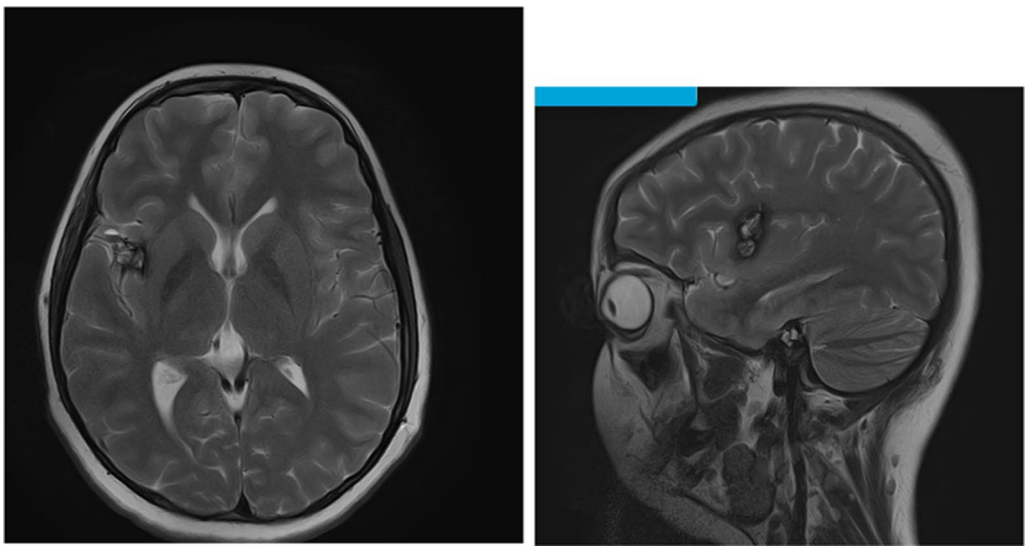

Fig. 1 Preoperative MRI brain-lobulated abnormal signal intensity area in the right insular cortex and right basal ganglia (measuring approximately 24×12 mm) No acute infarct or cerebral venous sinus thrombosis is detected

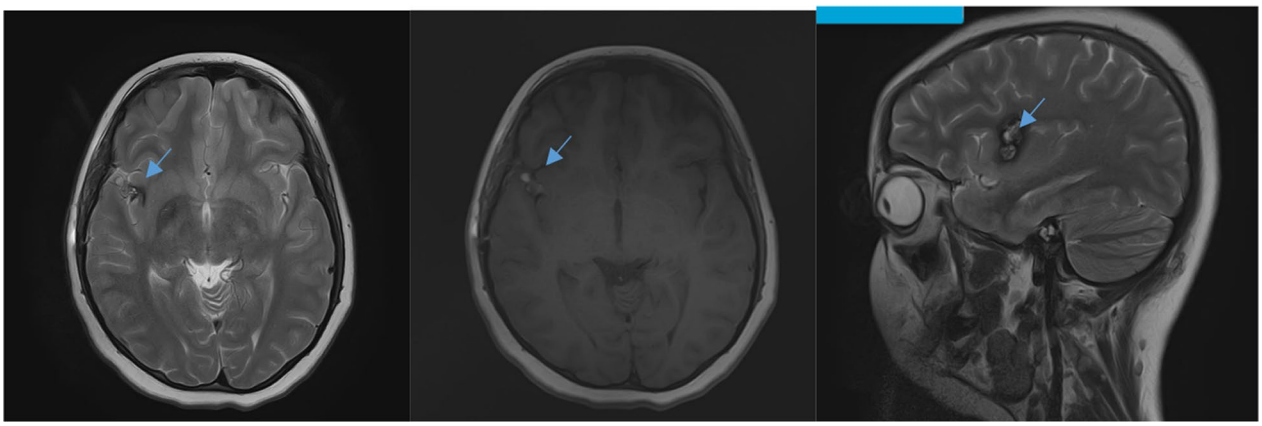

Fig. 2 Postoperative MRI: Status post-craniotomy with interval development of resection cavity in the right insular region. *Arrow showing redemonstration of lobulated abnormal signal intensity area medial to the surgical cavity representing residual cavernoma

haemorrhage, causing significant disability and morbidity [3, 5]. Cavernoma-related haemorrhages can cause significant morbidity with seizures and / or focal neurologic deficits. If left untreated, the probability of recurrent haemorrhages likely increases over time, specifically in cases with bleeding at presentation or localization in spinal cord [3]. In our case, the patient was symptomatic at presentation.

Diagnostic modalities

According to Nobukata et al. magnetic resonance imaging (MRI) is more sensitive in comparison with computed tomography (CT) and angiography in detecting cavernous malformations [4]. MRI is considered the most reliable imaging modality for not only the identification, but also for the follow-up of cavernous malformation [4].

On MRI, cavernomas are better demonstrated utilizing special techniques such as gradient echo (GRE) or susceptibility weighted imaging (SWI) [6]. The MRI appearance of cavernoma has been described as a mixed signal lesion on all sequences enclosed by a haemosiderin rim as it contains blood at various stages of evolution. Its appearance is often described as resembling mulberry or popcorn on imaging [4, 6].

Subacute haemorrhage and degraded blood products in the lesion create a halo of signal hyperintensity surrounding the lesion on T-1 weighted images. This is particularly useful in differentiating cavernomas from other intracranial haemorrhages [6].

Management

Management and follow-up of cavernomas depend on multiple factors such as clinical presentation, comorbid conditions, location of lesion, and haemorrhagic events [8, 9]. Total surgical resection is considered the best treatment option for patients who are symptomatic and have recurrent haemorrhages, intractable seizures, and progressive neurological defects [9]. Stereotactic radiosurgery has been utilized for treating deeply seated critical locations with some success [6, 7, 9].

Conclusion

Cavernoma is a rare neurovascular malformation, which can be detected incidentally or presents with seizures or focal neurological deficits. It can affect any part of the central nervous system. MRI is the imaging modality of choice, typically showing a lobulated lesion. Our case presented with episodes of loss of consciousness and automatisms that responded significantly to neuronavigational-guided lesion resection. Early referral, timely identification and management of the lesion reduced morbidity in our case.

Acknowledgements

Not applicable.

Author contributions

HA, SZ, EB and SK were contributors in writing the manuscript. All authors read and approved the final manuscript. EB provided his expert opinion and review being a senior consultant neurosurgeon.

Funding

No funding was received.

Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

Ethical approval was obtained from ethics review committee of Aga Khan University Hospital (ERC Number: 2023-9475-27191). Consent was obtained from the participant in this study.

Consent for publication
Consent for publication was obtained from the study participant.

Competing interests
The authors declare that they have no competing interests.

Received: 17 February 2024 Accepted: 10 April 2024
Published online: 22 May 2024

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