Case report

Diffuse subarachnoid and intraventricular hemorrhage as the presenting sign of a conus medullaris arteriovenous malformation: Case report

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Abstract

Spinal arteriovenous malformations (AVMs) are rare vascular lesions that usually present with progressive myelopathy or less frequently, with acute neurological deficit due to hematomyelia or spinal subarachnoid hemorrhage (SAH). There are few reports of concomitant cerebral SAH and intraventricular hemorrhage (IVH) following rupture of a spinal AVM. Herein, we present a rare case of conus medullaris arteriovenous malformation, out-breaking with loss of consciousness due to SAH and IVH.

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1. Introduction

Although cerebral aneurysms are the most common cause of non-traumatic subarachnoid hemorrhage (SAH), no intracranial vascular abnormalities is found in initial extensive work-up studies in 15–20% of patients. Except for second cerebral Digital Subtraction Angiography (DSA) [1], there are no uniform, accepted guidelines for performing further diagnostic studies. As the thoracolumbar spinal arteriovenous lesions (AVMs) are looked too far to cause diffuse SAH and intraventricular hemorrhage (IVH), the role of diagnostic imaging studies for lower spine including MRI or angiography is still uncertain in these patients.

Herein, we describe a rare case of angiogram-negative diffuse SAH and IVH due to a conus medullaris AVM.

2. Case report

A 48-year-old man was admitted with loss of consciousness (Glasgow coma scale 8) after experiencing a severe acute headache. On admission, head computed tomography (CT) revealed a diffuse SAH in basal cisterns and IVH (Figs. 1 and 2). Cranio cervical CT-angiogram (CTA) was negative for any vascular lesion, including aneurysm. The initial cranio cervical DSA failed to identify the bleeding source on day 3. A repeat cranio cervical DSA was normal 10 days later. When he gained...
consciousness few days later, he complained of low back pain and slight paraparesis. A whole spine MRI revealed prominent serpentine flow void ventral and dorsal to the spinal cord (Fig. 2). Hyperintense signals were shown at the conus medullaris in T2WI. Spinal angiography revealed a conus medullaris AVM at L1 that was supplied by medullary arteries from anterior spinal artery (ASA) and drained into medullary veins (Fig. 3).

Embolizations of AVM were performed through superselective catheterization of ASA and the medullary artery by 0.5 ml n-butyl-2-cyanoacrylate 75% (Glubrand 2, GEM, Italy) and 0.7 ml n-butyl-2-cyanoacrylate 33% (Glubrand 2, GEM, Italy) in two-consequence sessions within 3 months. He gained partial improvement after embolization then he was discharged to rehabilitation center.

3. Discussion

Initial CTA and cerebral DSA can reveal the cause of bleeding in the majority of cerebral SAHs. However, in 15% of cases, a cause cannot be readily identified. Rarely the origin is spinal pathologies [2]. Diffuse SAH with IVH is a rare presentation of thoracolumbar AVMs [3–5]. Our patient was admitted with coma due to both SAH and IVH. The initial work up did not show any cause. The development of symptoms in lower extremities lead us to explore the spine and to find a conus medullaris AVM. Lower AVMs seldom cause loss of consciousness [3]. It would require high suspicion not to miss the spinal origin of loss of consciousness following IVH and SAH.

A modified classification system for spinal arteriovenous lesions based on the anatomical and pathophysiological factors has been proposed by Spetzler et al. [6]. They are classified as: Extradural AVF, Intradural Dorsal AVF, Intradural Ventral AVF, Extradural-Intradural AVM, Intramedullary AVM (depending on the angio-architecture of nidus can be compact of diffuse), Conus Medullaris AVM. According to this system, our patient had a conus medullaris AVM lesion. Both upper and lower motor neuron symptoms can be observed due to vascular anatomical features of these lesions.

Spinal AVMs usually cause spinal SAH and/or hematomyelia [7] but cerebral SAH is rare [4]. Spinal aneurysms have greater trend to present with hemorrhage than the other AVMs and cranial SAH was reported in 30% of cases [8].

![Fig. 1](image1.png)

**Fig. 1** – (A and B) CT scan showed diffuse subarachnoid hemorrhage in basal cisterns and intraventricular hemorrhage.

![Fig. 2](image2.png)

**Fig. 2** – (A) T2 Weighted MRI depicting hyper-intensity changes in the lower spinal cord. (B) Spinal MRI (myelogram) revealing prominent flow and serpentine flow voids ventral and dorsal to the spinal cord consistent with dilated intrathecal vessels.
Conus medullaris AVM commonly becomes symptomatic with progressive myelopathy and radiculopathy, rarely with hemorrhage and cerebral SAH [9]. In children, there is little distance between conus and head, so hemorrhage from conus vascular malformation could easily produce cerebral SAH, as most of the reported cases are children. In adults, a large and continuous bleeding from conus AVM could result in cerebral SAH. The hemorrhage could be diffuse to cerebral subarachnoid space (SAS) when there is less space in the spinal SAS and there are large and rapid connections between cerebral and spinal SAS. The hemorrhage can enter easily to ventricular system through Lushka and Magendi when it arrives to cerebral SAS. It could also be assumed as a coincidence lesion.

Rebleeding from ruptured AVM is reported in 28–50% and occurs as soon as 24 h, to as late as 16 years [3,4,8,10]. Accordingly, not only the hemorrhagic presentation of AVM is associated with poor outcome but also the rehemorrhage and myelomalacia could worsen the outcome. Spinal MRI and angiography are consecutively the initial steps for imaging and the gold standard for diagnosis of spinal AVM.

This case and other reported cerebral SAHs associated with SVMs could create two questions: Is the current diagnostic work up sufficient for all cases of NASAH and would it be worth to add whole spinal MRI to those patients who also have paraparesis or long-standing lower back pain?

Conflict of interest

None declared.

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Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

REFERENCES


