

Spinal dural arteriovenous fistula presenting with subarachnoid hemorrhage

A case report

Jingzhe Han, MD^{a,*}, Duanhua Cao, MD^a, Hongmei Wang, MD^a, Ye Ji, MD^a, Zhilei Kang, MD^b, Jianguo Zhu, MD^a

Abstract

Rationale: Subarachnoid hemorrhage (SAH) is a common and serious disease and one of the most important differential diagnoses in the emergency department.

Patient concerns: A 39-year-old female patient with a 12 years' history of migraine, presented with a sudden headache combined with motor aphasia. Physical examination suggested probable positive neck resistance. SAH was confirmed by magnetic resonance imaging. In addition, spinal digital subtraction angiography and spinal vascular computed tomography angiography indicated spinal arteriovenous malformation (SAVM).

Diagnoses: The final diagnosis was spinal dural arteriovenous fistula presenting with SAH.

Interventions: Following diagnosis, appropriate drugs were administered, but the therapeutic effect was poor. Then the patient was then transferred to a superior hospital where she was treated with interventional embolization.

Outcomes: Through 2 years of follow-up, intermittent migraine was found in the patient, but the symptoms of occipital pain, nausea, and vomiting did not occur again.

Lessons: For patients who have been diagnosed with SAH but have no definite cause, we should pay attention to the etiological screening of SAH, and the possibility of the spinal cord SAH should be vigilant. The pain in the chest and back and the signs of spinal cord may be an important breakthrough in patients with spinal cord SAH, and medical history and physical examination are particularly important.

Abbreviations: CSF = cerebrovascular fluid, CTA = computed tomographic angiography, DSA = digital subtraction angiography, PMAVF = perimedullary arteriovenous fistula, SAH = subarachnoid hemorrhage, SAVMs = spinal arteriovenous malformations, SDAVFs = spinal dural arteriovenous fistulas, TCD = transcranial Doppler.

Keywords: migraine, spinal arteriovenous malformation, subarachnoid hemorrhage

1. Introduction

Subarachnoid hemorrhage (SAH) is one of the most important differential diagnoses in the emergency department.^[1] It is a clinical syndrome caused by bleeding from vascular arteriovenous malformations or aneurysms directly enter the subarachnoid space after rupture. Headache, intracranial hypertension,

and meningeal irritation are the most common clinical manifestations.^[2]

Spinal arteriovenous malformations (SAVMs) are rare congenital vascular diseases of the spinal cord, which accounting for about 2% to 4% of spinal diseases. Its natural history is still not fully defined.^[3,4] SAVMs can be divided into three types: spinal dural arteriovenous fistulas (SDAVFs), perimedullary arteriovenous fistula (PMAVF), and SAVMs.^[5]

The occurrence of SAVMs is rare and is sometimes misdiagnosed as a result of nonspecific clinical symptoms.^[6–8] If the best opportunity to treat acute aneurysmal SAH is missed, the high incidence of associated mortality and morbidity may result in devastating consequences.^[9,10] In the report of Vermeulen and Schull,^[11] 1507 patients were diagnosed with SAH, and 5.4% had missed the treatment.

In this study, we reported a patient with SDAVF presenting with spinal SAH, the difficulties and traps encountered in the course of the diagnosis were reviewed.

2. Case description

A 39-year-old female patient presented with a sudden headache and a motor aphasia for 4 hours within 2 days before admission. Pain in the parietal areas of head was a throbbing and swelling pain, accompanied by nausea but without vomiting. The patient

Editor: N/A.

The authors have no conflicts of interest to disclose.

^a Department of Neurology, ^b Department of Magnetic Resonance, Harrison International Peace Hospital, Hengshui, Hebei, China.

* Correspondence: Jingzhe Han, Department of Neurology, Harrison International Peace Hospital, 2 Renmin Zhong Road, Taocheng District, Hengshui 053000, Hebei, China (e-mail: hanjingzhe2017@sina.com).

Copyright © 2018 the Author(s). Published by Wolters Kluwer Health, Inc. This is an open access article distributed under the terms of the Creative Commons Attribution-Non Commercial-No Derivatives License 4.0 (CCBY-NC-ND), where it is permissible to download and share the work provided it is properly cited. The work cannot be changed in any way or used commercially without permission from the journal.

Medicine (2018) 97:16(e0513)

Received: 9 December 2017 / Received in final form: 28 March 2018 /

Accepted: 29 March 2018

<http://dx.doi.org/10.1097/MD.0000000000010513>

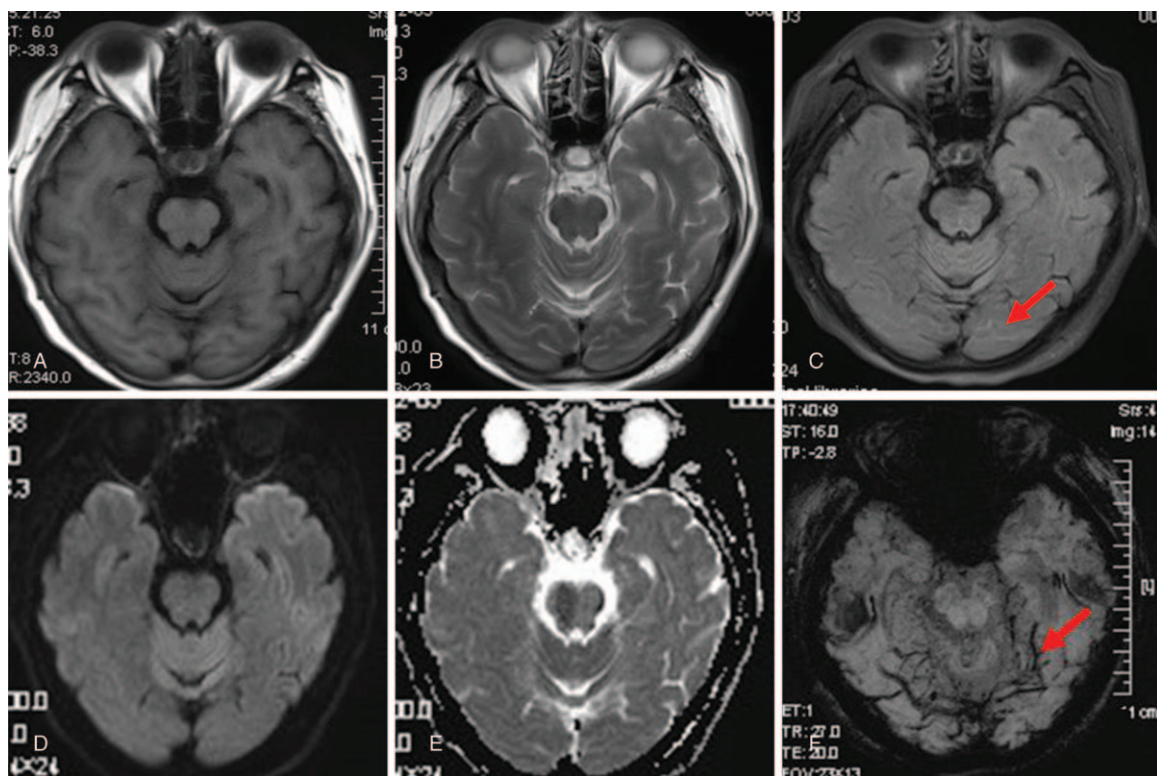


Figure 1. Head MRI showed bilateral occipital sulci lines with high FLAIR signal (C) and low SWI signal (F) (indicated by the red arrows). No characteristic changes were seen in the T1-weighted (A), T2-weighted (B), ADC (E), or DWI (D) sequences. SAH was considered.

has a 12 years history of migraine. Head computed tomography (CT) revealed no obvious abnormal changes. Transcranial Doppler (TCD) ultrasound showed that the velocity of blood flow was accelerated, then cerebrovascular spasm was suggested. Physical examination suggested resistance in the neck, but Kernig sign and Brudzinski sign were negative. The patient was initially diagnosed with migraine and suspected acute migraine attacks. Head magnetic resonance angiography (MRA) results was normal, however, SAH was considered because magnetic resonance imaging (MRI) showed abnormal signals in the bilateral occipital lobes (Fig. 1). Three days after admission, the lumbar puncture revealed uniform pink cerebrovascular fluid (CSF). Routine tests of the CSF showed that the RBC count was 8×10^6 cells/L (0–5 cells/visual field), then SAH was confirmed. Four days later, TCD reexamination showed that the blood flow of the middle cerebral artery return to normal, then cerebral vasospasm was reconfirmed. There was no obvious abnormality in whole-brain digital subtraction angiography (DSA). However, spinal DSA showed SAVM (Fig. 2). In addition, SAVM was also confirmed due to the CTA result of spinal vascular, and SDAVF was considered (Fig. 3). The final diagnosis was SDAVF presenting with SAH.

An antifibrinolytic agent (6-aminocaproic acid 12 g, infusion b.i.d.) was administrated, nimodipine liquid injection (5 mL/h, intravenous infusion) was administrated to prevent cerebral vasospasm, and rapid infusion (q8h) of mannitol (125 mL) was used for prevention of dehydration and reduction of intracranial pressure. The patient was then transferred to a superior hospital to be treated with interventional embolization.

Through 2 years of follow-up, discontinuous migraine attacks were found in patients. The headache was a mild, moderate,

temporal pulsatile headache that lasted for several hours, and can be relieve by itself. The explosive headache, posterior occipital pain, nausea, vomiting, and other associated symptoms did not occur again. At the time of headache, the head MRI was examined 2 times in a local hospital, with a interval of about half a year. No evidence of SAH was found. In addition, the patient had no recurrent pain on the chest and back, and no symptoms and signs of spinal cord involvement. However, it was regrettable that the patient did not receive the imaging examination of the spinal and spinal cord angiography.

3. Discussion

SAH has been reported to be attributed to the lumbar dural arteriovenous fistula,^[12] spinal perimedullary arteriovenous fistulas (SPAVFs) or SDAVFs,^[13] or cervical SDAVF.^[14,15] Perimesencephalic nonaneurysmal SAH also has been reported.^[16] Inamasu et al^[17] reported a rare case of dural arteriovenous fistula in the craniofacial junction showed cerebellar hemorrhage. In a recent study, transverse sinus thrombosis associated with peripheral arterial aneurysm SAH was reported.^[18] There are many reports about SAH caused by spinal vascular malformations, but the difference between our article and their reports is that our paper is not a simple case description. Instead, we summarized the “traps” in the process of the diagnosis and treatment of SAH by this case.

Previous history of headache might interfere with the diagnosis of SAH. The clinical manifestations of spinal SAH mainly include headaches, unconsciousness, back pain, and symptoms of nerve root irritation. It was reported that 80% of spinal SAH patients had cerebral symptoms, 70% of them had symptoms of

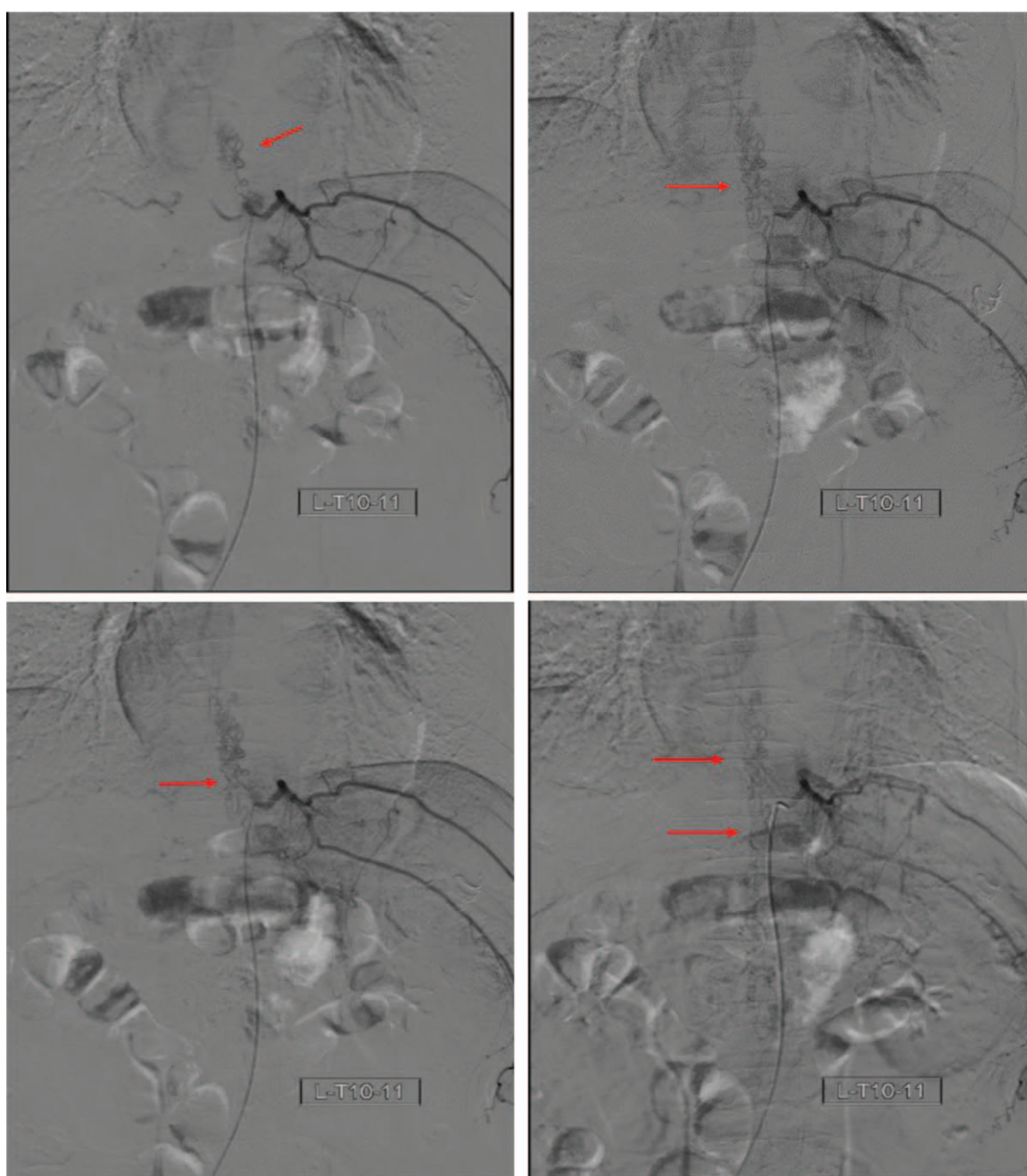


Figure 2. Spinal angiography revealed spinal cord arteriovenous malformation (indicated by the arrows) at the level of the T8, T9, and T10 vertebrae. The feeding artery was the T10 intercostal artery.

headache, and 22% of patients showed symptoms of consciousness change. Headaches may be the initial symptom of the spinal SAH, and the degree of headache may be as severe as that of intracranial SAH.^[19] The most serious headache can be regarded as a crucial symptom of SAH.^[20–22] The patient reported that her headache was the most serious headache in the past 12 years. Headache and other brain symptoms can be considered as the first symptom of spinal cord SAH, SAH, and intracranial headaches can be equally intense. SAH could be excluded basically based on CT and physical examination. However, the positive rate of SAH was about 90% only when the CT scan was performed on the first day after the onset of SAH. However, after 24 hours, this percentage decreased, negative CT results did not

exclude the possibility of SAH.^[23] Valle Alonso et al^[24] reported that CT scan within 6 hours of a sudden headache is sufficient to confirm or exclude SAH in patients with sudden headache but without nerve defects.

Lumbar puncture may be unnecessary when the cranial CT is normal, especially in low-risk SAH patients.^[24,25] But in this study, SAH was confirmed according to lumbar puncture result. It was prompted that lumbar puncture examination should be performed as early as possible to confirm the diagnosis for patient with negative head CT, but there was a sudden headache and meningeal irritation was positive.^[26]

Flair and susceptibility weighted imaging (SWI) magnetic resonance imaging can provide new basis for the diagnosis of

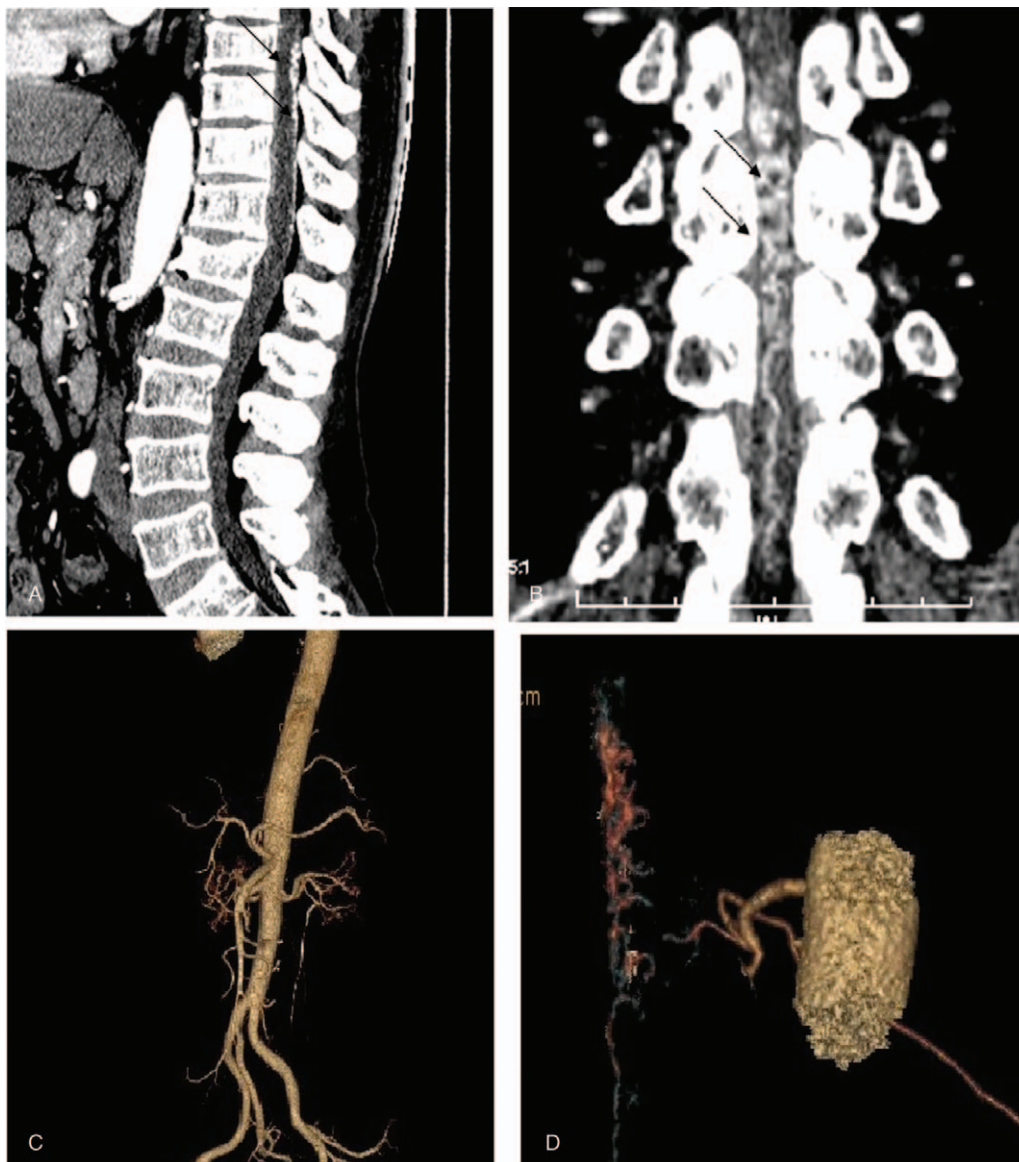


Figure 3. Spinal vascular CTA showed spinal dural arteriovenous fistula. The arrow in the figure A and B refers to the varicose vein mass in the surface of the spinal cord. (C) Spinal vascular CTA result showed that the posterior spinal vein of the horizontal direction of the thoracic spine in thoracic vertebrae 9 to 11 appeared earlier, winding into a group, and a slender posterior spinal vein downward drainage can be seen. (D) In the horizontal direction of right thoracic vertebrae 9 and 10, a fine vessel connected to the intercostal artery and laterally entered the venous mass.

SAH patients with negative CT, but lumbar puncture was refused. In this patient, head MRI showed bilateral occipital sulci lines with a high FLAIR signal and low SWI signal, which was considered to be the impact of SAH; this result was consistent with that of Verma et al.^[27]

Although negative DSA results could basically exclude most of bleeding etiology, such as aneurysmal and vascular malformation, many possible causes of SAH still remained. It is difficult to screen the entire etiology of SAH. Head DSA was normal, then negative SAH or spontaneous SAH may be easy to consider. Although SAH was confirmed in this patient, but no particular cause was found in cerebral screening, then the possibility of spinal SAH was kept vigilant. A large quantity of research has shown that TCD has a high rate of accurately detecting of cerebral vasospasm, and also has a high rate of agreement with DSA and computed tomographic angiography (CTA).^[28,29]

Combined the result of spinal vascular CTA with spinal DSA, SAVMs were identified, and SDAVF was considered. The occurrence of SDAVF with SAH as the initial presentation was relatively rare, which seemed to be related to the locations of the dural arteriovenous fistulas. The etiology of SAH is related to venous hypertension. Increased venous pressure caused by movement, certain postures, pregnancy, and valsalva movement, resulting in venous rupture and then leading to SAH. The patient reported that she had transient back pain during the valsalva exercise. The possibility of spinal SAH was considered.

4. Conclusion

For patients who have been diagnosed with SAH but have no definite cause, we should pay attention to the etiological screening of SAH, and the possibility of the spinal cord SAH

should be vigilant. The pain in the chest and back and the signs of spinal cord may be an important breakthrough in patients with spinal cord SAH, and medical history and physical examination are particularly important.

Acknowledgments

This study was approved by the institutional review board of Harrison International Peace Hospital. Written informed consent was obtained from the members of the patient's family for publication of this report.

Author contributions

Conceptualization: Duanhua Cao, Zhilei Kang, Jianguo Zhu.

Data curation: Hongmei Wang.

Investigation: Jingzhe Han, Hongmei Wang.

Methodology: Ye Ji.

Resources: Duanhua Cao.

Software: Zhilei Kang.

Writing – original draft: Jingzhe Han, Ye Ji, Jianguo Zhu.

Writing – review & editing: Jingzhe Han.

References

- [1] Ferro JM, Canhão P, Peralta R. Update on subarachnoid haemorrhage. *J Neurol* 2008;255:465–79.
- [2] Feigin VL, Lawes CM, Bennett DA, et al. Stroke epidemiology: a review of population-based studies of incidence, prevalence, and case fatality in the late 20th century. *Lancet Neurol* 2003;2:43–53.
- [3] Ducruet AF, Crowley RW, McDougall CG, et al. Endovascular management of spinal arteriovenous malformations. *J Neurointerv Surg* 2013;5:605–11.
- [4] Berenstein A. Endovascular treatment of spinal cord AVMs. *Neuroradiology* 1991;33:187–93.
- [5] Cho WS, Kim KJ, Kwon OK, et al. Clinical features and treatment outcomes of the spinal arteriovenous fistulas and malformation: clinical article. *J Neurosurg Spine* 2013;19:207–16.
- [6] Brinjikji W, Nasr DM, Morris JM, et al. Clinical outcomes of patients with delayed diagnosis of spinal dural arteriovenous fistulas. *AJNR Am J Neuroradiol* 2016;37:380–6.
- [7] Krings T, Geibprasert S. Spinal dural arteriovenous fistulas. *AJNR Am J Neuroradiol* 2009;30:639–48.
- [8] Hurst RW, Kenyon LC, Lavi E, et al. Spinal dural arteriovenous fistula: the pathology of venous hypertensive myelopathy. *Neurology* 1995;45:1309–13.
- [9] Roos YB, de Haan RJ, Beenen LF, et al. Complications and outcome in patients with aneurysmal subarachnoid haemorrhage: a prospective hospital based cohort study in the Netherlands. *J Neurol Neurosurg Psychiatr* 2000;68:337–41.
- [10] Connolly ES Jr, Rabinstein AA, Carhuapoma JR, et al. Guidelines for the management of aneurysmal subarachnoid hemorrhage: a guideline for healthcare professionals from the American Heart Association/American Stroke Association. *Stroke* 2012;43:1711–37.
- [11] Vermeulen MJ, Schull MJ. Missed diagnosis of subarachnoid hemorrhage in the emergency department. *Stroke* 2007;38:1216–21.
- [12] Koch C, Gottschalk S, Giese A. Dural arteriovenous fistula of the lumbar spine presenting with subarachnoid hemorrhage. Case report and review of the literature. *J Neurosurg* 2004;100(4 suppl Spine):385–91.
- [13] Kai Y, Hamada J, Morioka M, et al. Arteriovenous fistulas at the cervicomedullary junction presenting with subarachnoid hemorrhage: six case reports with special reference to the angiographic pattern of venous drainage. *AJNR Am J Neuroradiol* 2005;26:1949–54.
- [14] Aviv RI, Shad A, Tomlinson G, et al. Cervical dural arteriovenous fistulae manifesting as subarachnoid hemorrhage: report of two cases and literature review. *AJNR Am J Neuroradiol* 2004;25:854–8.
- [15] Fassett DR, Rammos SK, Patel P, et al. Intracranial subarachnoid hemorrhage resulting from cervical spine dural arteriovenous fistulas: literature review and case presentation. *Neurosurg Focus* 2009;26:E4.
- [16] Hong Y, Fang Y, Chen T, et al. Recurrent perimesencephalic nonaneurysmal subarachnoid hemorrhage: case report and review of the literature. *World Neurosurg* 2017;107:877–80.
- [17] Inamasu J, Tanaka R, Nakahara I, et al. Dural arteriovenous fistula of the craniocervical junction manifesting as cerebellar haemorrhage. *Neuroradiol J* 2016;29:356–60.
- [18] Fu FW, Rao J, Zheng YY, et al. Perimesencephalic nonaneurysmal subarachnoid hemorrhage caused by transverse sinus thrombosis: a case report and review of literature. *Medicine (Baltimore)* 2017;96:e7374.
- [19] Hankey GJ, Nelson MR. Practice easily missed? Subarachnoid haemorrhage. *BMJ* 2009;339:b2874.
- [20] Evans RW. Diagnostic testing for the evaluation of headaches. *Neurol Clin* 1996;14:1–26.
- [21] Rico M, Benavente L, Para M, et al. Headache as a crucial symptom in the etiology of convexal subarachnoid hemorrhage. *Headache* 2014;54:545–50.
- [22] Agostoni E, Zagaria M, Longoni M. Headache in subarachnoid hemorrhage and headache attributed to intracranial endovascular procedures. *Neurol Sci* 2015;36(suppl 1):67–70.
- [23] Mark DG, Kene MV, Udaltsova N, et al. Sensitivity of a clinical decision rule and early computed tomography in aneurysmal subarachnoid hemorrhage. *West J Emerg Med* 2015;16:671–6.
- [24] Valle Alonso J, Fonseca Del Pozo FJ, Vaquero Álvarez M, et al. Sudden headache, lumbar puncture, and the diagnosis of subarachnoid hemorrhage in patients with a normal computed tomography scans. *Emergencias* 2018;30:50–3.
- [25] Migdal VL, Wu WK, Long D, et al. Risk-benefit analysis of lumbar puncture to evaluate for nontraumatic subarachnoid hemorrhage in adult ED patients. *Am J Emerg Med* 2015;33:1597–601.
- [26] Carpenter CR, Hussain AM, Ward MJ, et al. Spontaneous subarachnoid hemorrhage: a systematic review and meta-analysis describing the diagnostic accuracy of history, physical examination, imaging, and lumbar puncture with an exploration of test thresholds. *Acad Emerg Med* 2016;23:963–1003.
- [27] Verma RK, Kottke R, Andereggen L, et al. Detecting subarachnoid hemorrhage: comparison of combined FLAIR/SWI versus CT. *Eur J Radiol* 2013;82:1539–45.
- [28] Matula C. Subarachnoid hemorrhage and cerebral vasospasm—facts and perspectives. *Wien Klin Wochenschr* 2000;112:519–22.
- [29] Dietrich HH, Dacey RG Jr. Molecular keys to the problems of cerebral vasospasm. *Neurosurgery* 2000;46:517–30.