SPONTANEOUS INTRATHECAL HYPOTENSION: CASE REPORT

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ABSTRACT

Introduction: Intracranial hypotension is a condition characterized by a lower than normal volume of cerebrospinal fluid (CSF) due to CSF leakage across the dural membrane in one or multiple locations. Symptoms include orthostatic headache and symptoms associated with stiff neck, tinnitus, photophobia, nausea, and evidence of low CSF pressure or leakage through at least one imaging modality. Case report: male patient, 46 years old, physician, with report of spontaneous CSF leak during physical activity. It evolved with the following symptoms: pulsatile holocranial headache that worsens when lying down and getting up. He underwent oral treatment without success, requiring an invasive procedure such as a blood patch to improve symptoms. After a few days he underwent new imaging tests with no changes. Discussion: The most common site of CSF leak is the cervicothoracic junction or upper thoracic region. Epidural blood patches are frequently performed for the treatment of post-dural puncture headache, which may vary between the previous site of cerebrospinal fluid leak or the lumbar location.

KEYWORDS: CEREBROSPINAL FLUID; INTRACRANIAL HYPOTENSION; EPIDURAL BLOOD PLAQUE.

INTRODUCTION

Intracranial hypotension is a condition characterized by a lower-than-normal volume of cerebrospinal fluid (CSF) due to CSF leakage through the dural membrane, at one or multiple locations. The loss of CSF results in the displacement of brain structures, causing headaches and other neurological symptoms[1]. Although the headache may present benign signs, if left untreated, it can lead to serious adverse events such as stroke and neurological sequelae[2,3].

Headache with low CSF pressure is almost always caused by CSF leakage, which can be iatrogenic, traumatic, or spontaneous in origin. The etiology of spontaneous CSF leakage is still unclear, and the site of the leak can be anywhere along the spinal column, but is usually at the level of the thoracic or low cervical region[4]. The most common presentation is orthostatic headache. Other features may include cranial nerve palsies, dizziness, tinnitus, photophobia, changes in hearing, or neck stiffness[5].

Data from large community-based studies on the epidemiology of spontaneous intracranial hypotension are not available, but an estimate of the annual incidence is 4 to 5 cases per 100,000 people[6], which is about half the incidence of aneurysmal subarachnoid hemorrhage. Including children and adolescents, spontaneous intracranial hypotension can affect patients of any age, particularly women aged 35 to 55 years[1,4].

The present study aims to present a case of spontaneous cerebrospinal fluid (CSF) fistula. Based on this case report, we seek to explore methods of diagnosis and treatment for this manifestation.

CASE REPORT

Male patient, 46 years old, residing in the city of Goiânia/Goiás, physician, with no underlying pathologies, regularly practicing physical exercises (running and swimming), and no allergies. He reports that during a few days of vacation in December 2021, he started experiencing back pain in the cervical region after more strenuous physical exercise than usual at the beach, associated with pulsatile holocranial headache that worsened with lying down/standing up, with a pain score of 3/10. He used regular painkillers with partial relief of the pain. After 4 days, already in Goiânia, he returned to work and started experiencing diplopia, blurred vision, and decreased visual acuity. He underwent cervical spine magnetic resonance imaging (MRI) with a diagnosis of pachymeningitis, represented by Figure 01 below, leading to further investigation, during which he underwent lumbar puncture and viral panel, all of which were normal.

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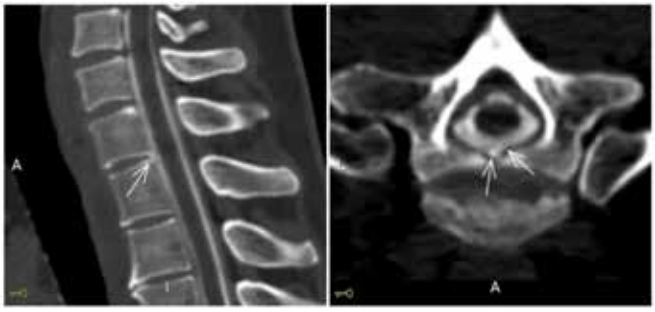

Figure 01: Computed tomography myelography of the cervical and thoracic spine showing epidural extravasation of contrast medium diffusely into the epidural space in the later acquisitions, from C5 to T5.

He underwent another MRI of the neuroaxis with contrast, which reported the presence of a collection in the C7-T1 region, leading to a diagnosis of cerebrospinal fluid hypotension. He rested at home for a week, with no improvement in mild pain symptoms. During this time, he was taking oral prednisolone for 5 days due to suspicion of viral meningitis. After the rest period, he returned to the hospital where an anesthesiologist performed a blood patch in the L3-L4 lumbar region with 20 ml, remaining at rest for another week without improvement. He returned to work with persistent symptoms, leading to another blood patch in the L2-L3 lumbar region with 20 ml. He remained at rest at home for another week, with no improvement.

He underwent a third blood patch with a higher location using 15 ml, without improvement. He decided to seek other means to continue the investigation, where he underwent a myelotomography with contrast, with a report of epidural extravasation of the contrast medium from T1-T2, spreading throughout the epidural space in the later acquisitions, from C5 to T5.

He returned to Goiânia, where a fourth blood patch was performed in the C7-T1 region guided by tomography with 10 ml, and after 15 days, the patient denied any symptoms. After five months, another MRI was performed, which was reported as normal.

DISCUSSION

The diagnostic criteria for spontaneous intracranial hypotension, as established by the International Classification of Headache Disorders, require the presence of (1) orthostatic headache, (2) associated symptoms of >=1 (neck stiffness, tinnitus, photophobia, nausea), and (3) evidence of low CSF pressure or leakage through at least one modality (brain MRI detecting indirect signs of low CSF pressure, lumbar puncture measuring CSF pressure less than 6 cm-H2O, CT myelography, or radionuclide cisternography RC)[7].

The most common site of CSF leakage is the cervicothoracic junction or upper thoracic region. This usually occurs spontaneously or after minor trauma to the spinal cord. As a result of CSF leakage, the brain sinks, causing traction on the bridging veins, pain-sensitive meningeal structures, and cranial nerves, leading to symptoms[8].

Epidural blood patches are commonly used for headaches attributed to CSF leakage. They are used to treat patients who experience headaches after a known dural injury. CSF leakage was first reported in 1955[9]. The etiology of this leakage is variable, with the most common cause being lumbar puncture. In a recent Cochrane Review, the blood patch was considered superior to conservative management for the treatment of post-dural puncture headache10. Most blood patches, however, are performed at the lumbar level. Cervical blood patching is feared for several reasons. Commonly cited complications include cranial nerve palsy[11], altered mental status[12,13], subdural hematoma[14], seizures1[5], and transient bradycardia[16]. Serious adverse events also include compression of nerve roots[17] and chemical meningitis[18]. At the cervical level, spinal cord compression becomes the most feared complication. However, no systematic study has been done to elucidate the incidence of these complications.

There are several reports indicating that the lumbar blood patch can permanently relieve headache, regardless of whether the site of leakage is identified or not[19,20,21]. However, other reports demonstrate that the lumbar blood patch does not always result in permanent relief[23,23,24]. A study by Diaz[25] suggests that the site of leakage should be identified by radionuclide cisternography and treated with a blood patch targeted at the levels of the CSF leak. A report by Kantor and Silberstein[26] also suggests that cervical blood patch may be useful after failure of lumbar blood patch when the site of leakage is not identified.

Our case report exemplifies two unsuccessful lumbar epidural blood patch (EBP) attempts for spontaneous CSF leakage. Subsequently, when the site of the leakage was identified, a blood patch was performed at the C7-T1 level, resulting in symptom improvement.

A case report by Ferrante et al. indicated that the blood patch successfully treated spontaneous CSF leakage in the cervical region. This report also showed the spread of blood from the lumbar to the cervical region. Cousins et al.[28] suggested that placing the blood patch close to the site of CSF leakage is important. The proposed mechanism is that the injected blood seals the dural defect and stops the leakage. Another theory is that the blood injection causes an epidural tamponade effect on the leakage. It would seem sensible to target the treatment to the site of leakage to maximize the chances of success, but there is no clear evidence to support targeted blood patching, and randomized clinical trials are likely not feasible given the low incidence of the disease.

In anesthesia practice, a blood patch at the site of leakage is the treatment of choice if conservative measures fail. Other treatments have been suggested with varying success rates, including intrathecal fluid infusion, epidural sa-

line infusion, epidural dextran infusion, epidural fibrin glue injection, CSF diversion, and surgical repair of the leak[29]. The procedure then becomes more technically challenging and higher risk.

REFERENCES

tigation findings and treatment outcomes of spontaneous intracranial hypotension syndrome: a systematic review and metaanalysis. JAMA Neurol 2021;78:329-37. DOI:10.1001/jamaneurol.2020.4799

2. Schievink W. Stroke and death due to spontaneous intracranial hypotension. Neurocrit Care. 2013;18:248–251. DOI: 10.1007/s12028-012-9800-3

3. Zakaria R, Wilby M, Fletcher N. Spontaneous csf collection in the cervical spine may cause neurological deficit and intracranial. DOI: 10.2174/1874205X01307010001

4.Schievink WI, Michael LM II, Maya M, Klimo P Jr, Elijovich L. Spontaneous intracranial hypotension due to skull-base CSF leak. Ann Neurol 2021;90:514-6. DOI: 10.1002/ana.26175

5. Panya N, Suyin GT. High thoracic/Cervical epidural blood patch for spontaneous cerebrospinal fluid leak: a new challenge for anesthesiologists. Anesth Analg 2011;113:1476 –9. DOI: 10.1213/ANE.0b013e31822e5665

6. Schievink WI, Maya M, Moser FG, Simon PA, Nuño M. Incidence of spontaneous intracranial hypotension in a community. Beverly Hills, California, 2006- 2020. Cephalalgia 2021 September 23. https://doi.org/10.1177/0333102421104

7.Headache Classification Subcommittee of the International Headache Society. The International Classification of Headache Disorders: 2nd Ed. Cephalalgia. 2004;24(S1):9–160.

8.Sriganesh K, JS, VB. Targeted cervical epidural blood patch for the management of refractory headache in a patient with spontaneous intracranial hypotension. J Neurosurg Anesthesiol. 2015 Jan;27(1):68-9. DOI: 10.1097/ANA.0000000000000059

9. Nosik W. Intracranial hypotension secondary to lumbar nerve sleeve tear. J Am Med Assoc 1955;157: 1110–11.

10. Boonmak P, Boonmak S. Epidural blood patching for preventing and treating post-dural puncture headache. Cochrane Database Syst Rev 2010; 1: CD001791.

11. Perez M, Olmos M, Garrido FJ. Facial nerve paralysis after epidural blood patch. Reg Anesth 1993;18: 196–8.

12. Beers RA, Cambareri JJ, Rodziewicz GS. Acute deterioration of mental status following epidural blood patch. Anesth Analg 1993;76:1147–9.

13. Sperry RJ, Gartrell A, Johnson JO. Epidural blood patch can cause acute neurologic deterioration. Anesthesiology 1995;82:303–5.

14. Tekkok IH, Carter DA, Brinker R. Spinal subdural haematoma as a complication of immediate epidural blood patch. Can J Anaesth 1996;43:306–9.

15. Kardash K, Morrow F, Beique F. Seizures after epidural blood patch with undiagnosed subdural hematoma. Reg Anesth Pain Med 2002;27:433–6.

16. Andrews PJD, Anaes FRC, Ackerman WE, et al. Transient bradycardia associated with extradural blood patch after inadvertent dural puncture in parturients. Br J Anaesth 1992;69:401–3.

17. Waguri N, Tomita M, Hayatsu K, Okamoto K, Shimoji K. Epidural blood patch for treatment of spontaneous intracranial hypotension. Acta Anaesthesiol Scand 2002;46:747–50

18. Oh J, Camann W. Severe, acute meningeal irritative reaction after epidural blood patch. Anesth Analg 1998;87:1139–40

19. Ferrante E, Arpino I, Citterio A. Is it a rational choice to treat with lumbar epidural bloodpatch headache caused by spontaneous cervical CSF leak? Cephalalgia 2006;26:1245– 6

20. Terjima K, Oi Y, Ogura A, Sakai N, Takei M, Tamaki T, Ogawa R. Blood patch therapy for spontaneous intracranial hypotension: safe performance after epidurography in an unconscious patient. Anesth Anal 2002;94:959 – 61

21. Benzon HT, Nemickas R, Molloy RE, Ahmad S, Melen O, Cohen B. Lumbar and thoracic epidural blood injections to treat spontaneous intracranial hypotension. Anesthesiology 1996;85:920 –2

22. Berroir S, Loisel B, Ducros A, Boukobza M, Tzourio C, Valade D, Bousser M-G. Early epidural blood patch in spontaneous intracranial hypotension. Neurology 2004;63:1950 –1

23. Hannerz J, Dahlgren G, Irestedt L, Meyerson B, Ericson K. Treatment of idiopathic intracranial hypotension: cervicothoracic and lumbar blood patch and peroral steroid treatment. Headache 2006;46:508 –11

24. Nowak DA, Takano B, Topka H. Spontaneous cerebrospinal fluid hypovolaemia: a therapeutic dilemma? Eur J Neurol 2006;13:1128 –38

25. Diaz JH. Epidemiology and outcome of postural headache management in spontaneous intracranial hypotension. Reg Anesth Pain Med 2001;26:582–7

26. Kantor D, Silberstein SD. Cervical epidural blood patch for low CSF pressure headaches. Neurology 2005;65:1138

27. Ferrante E, Arpino I, Citterio A. Is it a rational choice to treat with lumbar epidural bloodpatch headache caused by spontaneous cervical CSF leak? Cephalalgia 2006;26:1245– 6

28. Cousins MJ, Brazier D, Cook R. Intracranial hypotension caused by cervical cerebrospinal fluid leak: treatment with epidural blood patch. Anesth Anal 2004;98:1794 –7

29.. Mokri B. Headaches caused by decreased intracranial pressure: diagnosis and management. Curr Opin Neurol 2003;16:319 –26