

SPONTAN İNTRAKRANYAL HİPOTANSİYON

Defne ALİKILIÇ¹
Bülent KARA²

GİRİŞ

İntrakranyal basınç dinamiklerini beyin omurilik sıvısının (BOS) üretimi, emilimi ve akışı belirler.¹ BOS basıncı ve volümündeki azalma intrakranyal hipotansiyon olarak tanımlanır ve başta ortostatik karakterde baş ağrısı olmak üzere çeşitli nörolojik semptomlara neden olabilir.

Spontan intrakranyal hipotansiyon (SIH) nadir olmakla birlikte, giderek daha fazla tannılmaktadır. Bu sendromun başlıca özellikleri ortostatik baş ağrısı, BOS basıncında azalma ve manyetik rezonans görüntülemede yaygın meningeal kontrastlanmadır.

SIH yerine geçmişse spontan veya idyopatik düşük BOS basınçlı baş ağrısı, düşük BOS hacimli baş ağrısı, BOS azalması baş ağrısı, BOS yokluğu baş ağrısı, BOS kaçağına bağlı baş ağrısı, BOS hipovolemisi, BOS hacminde azalma gibi terimler kullanılmıştır.¹ Bazı yazarlar BOS kaçağıyla ilişkili semptomlar olduğunda 'BOS hipovolemi sendromu' terimini kullanmanın daha doğru olduğunu önermektedir.²

ETYOLOJİ VE PATOFİZYOLOJİ

Intrakranyal hipotansiyon spontan, dural ponksiyon sonrasında veya travmayla ilişkili

olarak gelişebilir. Spontan BOS kaçağı gelişen olgularda kalitsal bağ dokusu hastalıkları ve vasküler anomalilerin dışlanması gereklidir. SIH etyolojisinde yer alan hastalıklar Tablo 1'de gösterilmiştir.

Tablo 1: Spontan intrakranyal hipotansiyon etyolojisi^{9,10,11,27}

Kalitsal Bağ dokusu Hastalıkları

Marfan sendromu
Ehler-Danlos sendromu (özellikle tip II)
Otozomal dominant polikistik böbrek hastlığı
İzole eklem hipermobilitesi
Spontan retina ayrışması sendromu
Sınıflanamayan bağ dokusu hastalıkları

Diger nedenler

Meningeal divertikül
Kalsifiye disk protrüzyonu veya osteofitlere bağlı ventral dural yırtıklar
BOS-paraspinal venöz fistüller
Gizli/minör travmalar

Nedeni saptanamayanlar

Normal kranyal-spinal anatominin yapıda beyin BOS tarafından desteklenir. Beynin ağırlığı normalde yaklaşık 1,500 g iken, BOS içinde 48'ę denk gelir.³ BOS basıncı azaldıkça beyin parenkimini destekleyen kaldırma kuvvetinde

¹ Uzm. Dr., Kocaeli Üniversitesi Tip Fakültesi, Çocuk Sağlığı ve Hastalıkları AD., Çocuk Nörolojisi BD., (Yandal araştırma görevlisi) defnekocaoglu@hotmail.com

² Prof. Dr., Kocaeli Üniversitesi Tip Fakültesi, Çocuk Sağlığı ve Hastalıkları AD., Çocuk Nörolojisi BD., bkuskudar@gmail.com

KAYNAKLAR

1. Sun-Edelstein C, Lay CL. Spontaneous intracranial hypotension: Pathophysiology, clinical features and diagnosis. In, Swanson J (ed). Last updated in Apr 08, 2021.
2. Miyazawa K, Shiga Y, Hasegawa T, et al. CSF hypovolemia vs intracranial hypotension in “spontaneous intracranial hypotension syndrome”. Neurology 2003; 60:941-7.
3. Horton JC, Fishman RA. Neurovisual findings in the syndrome of spontaneous intracranial hypotension from dural cerebrospinal fluid leak. Ophthalmology 1994; 101:244-51.
4. Schievink WI, Reimer R, Folger WN. Surgical treatment of spontaneous intracranial hypotension associated with a spinal arachnoid diverticulum. Case report. J Neurosurg 1994; 80:736-9.
5. Swanson JW, Dodick DW, Capobianco DJ. Headache and other craniofacial pain. In: Neurology in Clinical Practice, Bradley WG, Daroff RB, Fenichel GM, Marsden CD (Eds), Butterworth, Boston 2000. p.1832.
6. Marcelis J, Silberstein SD. Spontaneous low cerebrospinal fluid pressure headache. Headache 1990; 30:192-6.
7. Mokri B. Spontaneous cerebrospinal fluid leaks: from intracranial hypotension to cerebrospinal fluid hypovolemia--evolution of a concept. Mayo Clin Proc 1999; 74:1113-23.
8. Levine DN, Rapalino O. The pathophysiology of lumbar puncture headache. J Neurol Sci 2001; 192:1-8.
9. Ferrante E, Citterio A, Savino A, Santalucia P. Postural headache in a patient with Marfan's syndrome. Cephalgia 2003; 23:552-5.
10. Kumar N, Diehn FE, Carr CM, et al. Spinal CSF venous fistula: A treatable etiology for CSF leaks in craniospinal hypovolemia. Neurology 2016; 86:2310-2.
11. Schievink WI, Moser FG, Maya MM. CSF-venous fistula in spontaneous intracranial hypotension. Neurology 2014; 83:472-3.
12. Franzini A, Messina G, Nazzi V, et al. Spontaneous intracranial hypotension syndrome: a novel speculative physiopathological hypothesis and a novel patch method in a series of 28 consecutive patients. J Neurosurg 2010; 112:300-6.
13. Schievink WI, Maya MM, Jean-Pierre S, et al. A classification system of spontaneous spinal CSF leaks. Neurology 2016; 87:673-9.
14. Schievink WI, Maya MM, Moser FG, Tourje J. Spectrum of subdural fluid collections in spontaneous intracranial hypotension. J Neurosurg. 2005;103:608-13.
15. Schievink WI, Morreale MV, Atkinson JLD, Meyer FB, Piegras DG, Ebersold MJ. Surgical treatment of spontaneous spinal cerebrospinal fluid leaks. J Neurosurg. 1998;88:243-46.
16. Mea E, Chiapparini L, Savoardo M, et al. Headache attributed to spontaneous intracranial hypotension. Neurol Sci. 2008;29:164-165.
17. Schievink WI, Maya MM, Tourje J. False localizing sign of C1-2 cerebrospinal fluid leak in spontaneous intracranial hypotension. J Neurosurg 2004; 100:639-44.
18. Rando TA, Fishman RA. Spontaneous intracranial hypotension: report of two cases and review of the literature. Neurology 1992; 42:481-7.
19. Amorim JA, Gomes de Barros MV, Valenca MM. Post-dural (post-lumbar) puncture headache: Risk factors and clinical features. Cephalgia. 2012;32:916-23.
20. Ozge A, Bolay H. Intracranial hypotension and hypertension in children and adolescents. Curr Pain Headache Rep. 2014 Jul;18(7):430.
21. Syed NA, Mirza FA, Pabaney AH, Rameez-ul-Hassan. Pathophysiology and management of spontaneous intracranial hypotension- a review. J Pak Med Assoc. 2012;62:51-5.
22. Good DC, Ghobrial M. Pathologic changes associated with intracranial hypotension and meningeal enhancement on MRI. Neurology 1988;43:2698-700.
23. Krans PG, Gray L, Amrhein TJ. Spontaneous intracranial hypotension: 10 myths and misperceptions. Headache. 2018;58:948-59.
24. Mokri B. Spontaneous intracranial hypotension. Continuum (Minneapolis Minn) 2015;21:1086-108.
25. Luetmer PH, Mokri B. Dynamic CT myelography: a technique for localizing high-flow spinal cerebrospinal fluid leaks. AJNR Am J Neuroradiol. 2003;24:1711-4.
26. Wang YF, Lirng JF, Fuh JL, et al. Heavily T2-weighted MR myelography vs CT myelography in spontaneous intracranial hypotension. Neurology 2009;73:1892-8.
27. D'Antona L, Jaime Merchan MA, Vassilou A, et al. Clinical presentation, investigation findings and treatment outcomes of spontaneous intracranial hypotension syndrome: A systematic review and meta-analysis. JAMA Neurol 2021;78:329-37.
28. Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalgia 2018; 38:1-211.
29. Schievink WI. Spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension. JAMA. 2006 May 17;295(19):2286-96.
30. Lipman IJ. Primary intracranial hypotension:the syndrome of spontaneous low cerebrospinal fluid pressure with traction headache. Dis Nerv Syst. 1997;38:212-3.
31. Sun-Edelstein C, Lay CL. Spontaneous intracranial hypotension: Treatment and prognosis. In, Swanson JW (ed). Last updated in Jul 08, 2021.
32. Hoffmann J, Goadsby PJ. Update on intracranial hypertension and hypotension. Curr Opin Neurol 2013;26:240-7.

33. Amoozegar F, Guglielmin D, Hu W, et al. Spontaneous intracranial hypotension: recommendations for management. *Can J Neurol Sci* 2013;40:144-57.
34. Marcelis J, Silberstein SD. Spontaneous low cerebrospinal fluid pressure headache. *Headache* 1990;30:192-6.
35. Wilmott RW. Pediatric spontaneous intracranial hypotension. *J Pediatrics*. 2013;163:309-11.
36. Bateman BT, Cole N, Sun-Edelstein C, Lay CL. Post dural puncture headache. In, Hepner DL, Swanson JW (eds). UpToDate, last updated Feb 03, 2021.
37. Bakshi R, Mechtler LL, Kamran S, et al. MRI findings in lumbar puncture headache syndrome: abnormal dural-meningeal and dural venous sinüs enhancement. *Clin Imaging*. 1999;23:73-6.
38. Ljubisavljevic S, Trajkovic JZ, Ignjatovic A, Stojanov A. Parameters related to lumbar puncture do not affect occurrence of postdural puncture headache but might influence its clinical phenotype. *World Neurosurg*. 2020;133:e540.
39. Smith JH, Mac Grory B, Butterfield RJ, et al. CSF Pressure, Volume, and Post-Dural Puncture Headache: A Case-Control Study and Systematic Review. *Headache*. 2019; 59:1324-38.
40. Kokki H, Saloavaara M, Herrgård E, Onen P. Post-dural puncture headache is not an age-related symptom in children: a prospective, open-randomized, parallel group study comparing a 22-gauge Quincke with a 22-gauge Whitacre needle. *Paediatr Anaesth*. 1999; 9:429-34.
41. Kokki H, Heikkinen M, Turunen M, et al. Needle design does not affect the success rate of spinal anaesthesia or the incidence of postpuncture complications in children. *Acta Anaesthesiol Scand*. 2000; 44:210-3.
42. Nath S, Koziarz A, Badhiwala JH, et al. Atraumatic versus conventional lumbar puncture needles: a systematic review and meta-analysis. *Lancet* 2018;391:1197-204.
43. Strupp M, Brandt T, Müller A. Incidence of post-lumbar puncture syndrome reduced by reinserting the stylet: a randomized prospective study of 600 patients. *J Neurol*. 1998; 245:589-92.