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### Case Report

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#### ABSTRACT

CSF hypotension arises in the context of a leak of CSF which causes negative intracranial pressure. Sacral fractures result from high-energy trauma which are frequently underdiagnosed. A ten-year-old boy presented with hip pain, after a fall. He mobilized both lower limbs, reported no leg pain, irradiation nor lack of sphincter control. The neurological examination was normal. When asked to stand, he began biparietal headache, nausea and vomiting, which improved laying down. CT scan showed an occult intrasacral meningocele; the MRI revealed collections of CSF along the spine, a S3 fracture with potential laceration of the meningocele and opening of a CSF fistula. Our diagnosis was the CSF hypotension, secondary to the fistula opening. The diagnosis was challenging. The child first presented with symptoms of CSF hypotension without evident cause. The discovery of the meningocele led us to hypothesize the opening of a fistula, a rare diagnosis, later confirmed by MRI.

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#### Hipotensión del líquido cefalorraquídeo tras caída en un niño: caso clínico

#### RESUMEN

La hipotensión del LCR surge en el contexto de una fuga de dicho líquido que causa presión intracraneal negativa. Las fracturas del sacro son originadas por traumatismos de alta energía que a menudo no se diagnostican. Un niño de diez años acudió con dolor de cadera tras una caída. Podía mover las piernas, no reportando ningún dolor en las mismas, ni irradiación o falta de control del esfínter. El examen neurológico resultó normal. Cuando se le pidió que se pusiera de pie comenzó a sufrir cefalea biparietal, náuseas y vómitos, que

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mejoraban al tumbarse. La TC mostró un meningocele intrasacro oculto. La RM reveló rastros de LCR a lo largo de la columna, fractura en S3 con laceración potencial del meningocele y apertura de una fístula de LCR. Nuestro diagnóstico fue hipotensión de LCR, secundaria a la apertura de la fístula. El diagnóstico resultó difícil. El niño presentó síntomas de hipotensión del LCR sin causa evidente. El descubrimiento del meningocele nos llevó a conjeturar la apertura de una fístula, un diagnóstico infrecuente que fue confirmado posteriormente mediante RM.

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#### Introduction

The brain is supported by the cerebrospinal fluid (CSF), in such fashion that the brain's weight of 1500 g corresponds to only about 50 g "floating" in CSF.<sup>1</sup> CSF hypotension usually arises in the context of known or suspected leak of CSF which causes abnormal intracranial (negative) pressure lowering.<sup>2</sup> This leak may occur spontaneously or be due to trauma or iatrogenesis.<sup>3</sup> For the past years, there has been an increasing number of reports and also reviews on spontaneous intracranial hypotension showing the relevance of its diagnosis and management. There is no clear explanation about aetiology and its physiopathology in under discussion.<sup>4</sup> It is increasingly a recognized cause of atypical headache, as well.<sup>5</sup>

Our goal is to report a case of intracranial hypotension after a fall and trauma on the sacral and coccygeal region, in a ten-year-old boy with an unknown occult dysraphism and highlight the importance of a careful clinical history and examination.

#### **Case report**

A healthy ten-year-old boy presented with bilateral hip pain, after falling on the previous day, clashing the sacral region against the ground. Later he reported hip and lower limb pain, while trying to move, which led him to seek medical attention. On examination, he was again able to mobilize both lower limbs, reporting no leg pain or irradiation, no lack of sphincter control, and no sensitive deficits. The neurological examination was normal and there were no cutaneous stigmata of occult spinal dysraphism. However, when asked to stand up or with head elevation, he began biparietal headache, nausea and several vomiting episodes, which improved when laying down.

Computed tomography (CT) scan showed an occult intrasacral meningocele (Fig. 1) but was otherwise unremarkable. Due to persisting symptoms, he was admitted to the ward to pursue investigation. Magnetic resonance imaging (MRI) of the head and spine revealed slight haemorrhagic content along the cerebellum, the interhemispheric fissure and cerebral convexity, possibly a subarachnoid haemorrhage and meningeal irritation. Also, it showed collections of CSF anterior to the posterior arches of C1 and C2, C7-D1, D1-D11, and along the anterior sacral surface. It confirmed a cystic intracanal lesion between S2-S4: an occult intrasacral meningocele Fig. 1 – MRI showing an occult intrasacral meningocele.

with probable spinal dysraphism associated, and a recent fracture of S3 with potential laceration of the meningocele creating a CSF fistula (Figs. 2 and 3).

Our diagnosis was CSF hypotension, secondary to the fistula opening. The child was in bedrest for two weeks, under intravenous and oral hydration therapy and watchful waiting in neurosurgical care. No surgical nor invasive treatments were done. Subsequent control MRI showed significant CSF absorption with meningocele refilling and haemorrhage resolution. The child started to ambulate gradually with progressive and complete resolution of symptoms. Currently, he is asymptomatic and able to maintain normal physical activity for his age, with follow-up yearly neurosurgical consult.

#### Discussion

Occult spinal dysraphisms are rare and are often associated with cutaneous stigmata, which were absent in our case. There was also no history of autonomic or sphincteric dysfunction, sensorimotor deficits in the lower limbs, or back pain, prior to the fall.





Fig. 2 – MRI T2 with hyperintensity in S3 suggesting recent fracture.

Sacral fractures are uncommon injuries resulting from high energy trauma. They are frequently underdiagnosed and mistreated, and so are their complications. Rarely, they occur isolated, being frequently associated with neurological symptoms.<sup>5</sup> In our case, the presence of a meningocele adjacent to the sacral fracture helped creating an optimal condition to the resulting fistula and consequent CSF leakage. When there is opening of a CSF fistula, both CSF volume and pressure decrease, leading to a reduction in the resistance of the brain's supportive cushion.<sup>1</sup> The brain then falls in the cranial cavity, causing traction on its anchoring and supporting structures.<sup>2,3</sup> This traction on pain-sensitive and meningeal structure, namely sensory nerves and bridging veins, is the probable cause for the headache. Since this traction is stronger when standing, the headache has a postural component.<sup>1,2</sup> Furthermore, due to the fixed intracranial space, compensatory meningeal venodilation and blood volume expansion, possibly contribute to the headache as well. The vessel engorgement affects both the intracranial and spinal veins that may be enough to shear the bridging veins leading to the development of subdural haematoma or hygroma.<sup>1,2</sup>

CSF leakage usually develops because of unknown minor trauma, such as sneezing or stretching, due to structural weakness of meninges, but it can also be secondary to a lumbar punction or high energy trauma, such as sacral fractures.<sup>1,6</sup>

In clinical practice, CSF hypotension should be considered when confronted with a patient presenting with an orthostatic headache that relieves in the supine position. Other symptoms include neck pain, nausea, vomiting, double vision or cognitive impairment.<sup>6</sup> In our case, the boy presented with orthostatic headache, vomiting and nausea, symptoms that led us to perform a CT scan and later an MRI. The previously unknown meningocele and the fracture of S3 lead us to hypothesize that a supposed fistula would be located there.

According to Schievink criteria, the MRI demonstration of extrathecal CSF supports the diagnostic of CSF hypotension.<sup>7</sup> It also fulfils the "International Classification of Headache Disorders third edition" for headache secondary to CSF hypotension as it led to discovery of CSF pressure leak.<sup>8</sup> Since



Fig. 3 - Extra-canal CSF collections along the dorsal spine.

the diagnosis was supported with clinical and imaging criteria and the patient had resolution of symptoms with non-invasive approach, there was no need for further investigation.

The non-invasive conservative approach can include bedrest in the Trendelenburg position, steroids, hydration and abdominal binder. Additionally, intravenous or oral caffeine may be beneficial, due to the vasoconstrictive effect and as a stimulator of the CSF production.<sup>9</sup>

#### Conclusion

In conclusion, intracranial CSF hypotension is a rare condition with no clear aetiology or physiopathology, being spontaneous or secondary to trauma or iatrogenesis. The final diagnosis is challenging as the authors showed in this case. The child presented with symptoms of CSF hypotension but with no evident cause. The discovery of the meningocele led us to hypothesize the possible opening of a fistula, which is a rare diagnosis, later confirmed by MRI.

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### **Conflict of interests**

The authors declare that there were no conflicts of interest in conducting this work.

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#### REFERENCES

- Christina Sun-Edelstein C, Christine L Lay. Spontaneous intracranial hypotension: pathophysiology, clinical features, and diagnosis – UpToDate<sup>®</sup>. Available from: https://www. uptodate.com/contents/spontaneous-intracranialhypotensionpathophysiology-clinical-features-and-diagnosis [updated April 2018; accessed May 2019].
- Díaz-Cabanas L L, Prieto-González A, Frieiro-Dantas C, Vázquez-Herrero F, Aneiros-Peña F. Síndrome de hipotensión intracraneal espontánea: presentación de dos casos. Rev Neurol. 2013;56:283–8.
- Pattichis A, Slee M. CSF hypotension: a review of its manifestations, investigation and management. J Clin Neurosci. 2016;34:39–43, http://dx.doi.org/10.1016/j.jocn.2016.07.002.
- Peter G, Kranz PG, Gray L, Timothy J, Amrhein TJMD. Spontaneous intracranial hypotension: 10 myths and misperceptions – an editorial comment. Headache. 2019;59:977–8, http://dx.doi.org/10.1111/head.13328.
- Cultrera F, Lofrese G, Nasi MT. Spontaneous intracranial hypotension due to sacral diverticula: two-case history and a pocket-sized review. Neurocirugía. 2019;30:228–32, http://dx.doi.org/10.1016/j.neucir.2018.06.003.
- Rodrigues-Pinto R, Kurd MF, Schroeder GD, Kepler CK, Krieg JC, Holstein JH, et al. Sacral fractures and associated injuries. Global Spine J. 2017;7:609–16, http://dx.doi.org/10.1177/2192568.
- Schievink W, Maya M, Louy C, Moser F, Tourje J. Diagnostic criteria for spontaneous spinal CSF leaks and intracranial hypotension. Am J Neuroradiol. 2008;29:853–6, http://dx.doi.org/10.3174/ajnr.A0956.
- Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018;38:1–211.
- Bahram Mokri BMD. Spontaneous low pressure, low CSF volume headaches: spontaneous CSF leaks. Headache. 2013;53:1034–53, http://dx.doi.org/10.1111/head.12149.