Cerebrospinal fluid (CSF) shunt related intracranial hypotension: Ultrasound dynamic test of the optic nerve sheath diameter (ONSD), a new technique for diagnostic support

Hipotensión intracraneal relacionada con la derivación de líquido cefalorraquídeo (LCR): Prueba dinámica por ultrasonido del diámetro de la vaina del nervio óptico (ONSD), una nueva técnica de apoyo diagnóstico

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Resumen

Introducción: El sobredrenaje de líquido cefalorraquídeo (LCR) es una complicación conocida de los procedimientos de derivación en hidrocefalia, mientras que su fisiopatología, diagnóstico y conceptos terapéuticos siguen siendo temas de discusión. En este contexto, la hipotensión intracraneal está bien documentada. La ecografía del diámetro de la vaina del nervio óptico (ONSD) es un procedimiento emergente validado para la hipertensión intracraneal. Recientemente, se ha propuesto su utilidad diagnóstica para pacientes con síndrome de hipotensión intracraneal espontánea. **Material y Método:** Presentamos tres casos de cefalea asociada a shunt de LCR donde se sospecha de hipotensión intracraneal. La prueba dinámica por ecografía del diámetro de la vaina del nervio óptico, una técnica novedosa y no invasiva, se presenta como un método complementario para el diagnóstico y seguimiento del paciente. En todos los casos se reemplazaron las válvulas del shunt de LCR, logrando así la resolución completa de los síntomas y la normalización de la prueba dinámica por ecografía. **Conclusión:** Aunque se trata de una serie de casos limitada, creemos que la prueba dinámica por ecografía del ONSD puede ser una técnica útil para el diagnóstico y seguimiento del paciente con cefalea relacionada con el shunt de LCR en la que se sospecha sobredrenaje. Esta serie de casos parece ser el primer informe que evalúa la utilidad de la prueba dinámica por ecografía del ONSD para la evaluación no invasiva de la cefalea relacionada con el shunt de LCR, dejando un interesante campo de investigación en este complejo contexto clínico.

Palabras clave: Sobredrenaje de líquido cefalorraquídeo, hipotensión intracraneal, ecografía dinámica, diámetro de la vaina del nervio óptico, cefalea relacionada con shunt, evaluación no invasiva.

Abstract

Introduction: CSF overdrainage is a known complication of hydrocephalus diversion procedures while its physiopathology, diagnosis and therapeutic concepts are still topic of discussion. In this setting, intracranial hypotension is well documented. ONSD ultrasound is an emerging procedure validated for intracranial hypertension. Recently, its diagnostic utility has been proposed for patients with spontaneous intracranial hypotension syndrome. **Method and Material:** We present three cases of CSF shunt associated headache where intracranial hypotension is suspected. Ultrasound dynamic test of the optic nerve

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sheath diameter, a novel, non invasive, technique is presented as a complementary method for diagnosis and patient follow-up. In all cases CSF shunt valves were replaced, thus achieving complete resolution of symptoms and dynamic ultrasound test normalization. **Conclusion:** While its a limited case series, we believe that the ultrasound dynamic test of the ONSD can be a useful technique for diagnosis and patient follow-up for CSF shunt related headache in which overdrainage is suspected. This case series seems to be the first report that evaluates the utility of the dynamic ultrasound test of the ONSD for non-invasive evaluation of CSF shunt related headache, leaving an interesting field of research in this complex clinical setting.

Key words: Cerebrospinal fluid overdrainage, intracranial hypotension, dynamic ultrasound, optic nerve sheath diameter, shunt-related headache, non-invasive evaluation.

Introduction

CSF overdrainage is a known complication of hydrocephalus diversion procedures[1]. Many theories have been proposed to explain this entity, which were recently revised by Cheok[2]. Nowadays, the most accepted concept is the one related to “CSF shunt related headache”[1]. In 2008 Rekate described the slit ventricle syndrome, which is defined as severe, life-modifying headache in patients with CSF shunts and normal or smaller than normal ventricles[3].

Overdrainage physiopathology is complex and not completely understood since it is not attributable solely to CSF drainage as seen in cases where intracranial pressure is oddly elevated[1]. This has resulted in multiple attempts for classification that are difficult to apply in clinical practice[3-5].

As a common factor, all of them consider CSF hypotension associated headache as a specific, demonstrable entity in the setting of dysfunctional CSF shunt valves.

Optic nerve sheath ultrasonography is an emergent non-invasive technique validated for intracranial hypertension[6-10] which is standardized and routinely used in our center[11]. Recently its utility has been proposed for spontaneous intracranial hypotension syndrome diagnosis and patient following[12-14].

Patients and Methods

We present three consecutive cases in which optic nerve sheath ultrasonography its used as a diagnostic and patient follow-up tool in the setting of CSF shunt related headache.

Ultrasound measurement of ONSD was performed as described in literature using a SonoSite® HFL50x Linear Probe connected to a SonoSite® Edge II ultrasound machine[11,24]. Patient in supine decubitus, preferably with head at 30° with closed eyes. Apply non-alcoholic ultrasound gel to the transducer and it is gently positioned on the temporal portion of the upper eyelid, oriented perpendicular to the vertical axis with an angulation of about 30°, in the horizontal plane. Placement and depth are adjusted to view the optic nerve entry into the globe.

The ONS is projected posteriorly from the optic disc, which is identified as an hyperechogenic band.

First measurement is made 3 mm behind the globe, perpendicular to the major axis of the ONS. A second measurement is made perpendicular to the first one, between the hyperechogenic limits of the ONS, configuring its diameter. The measurement is repeated 3 times for each eye, considering as final value the average of them (Figure 1).

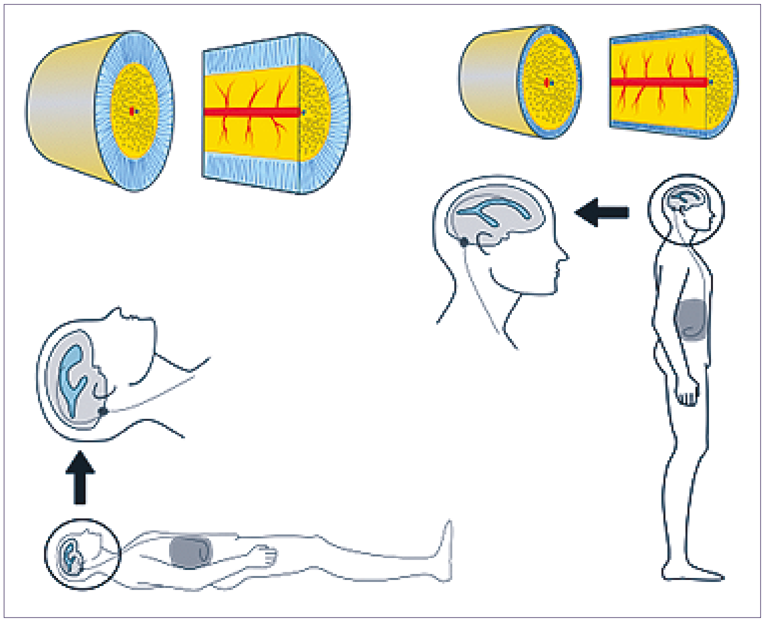

Figure 1. Dynamic test of ONSD ultrasound. Modified from Flitcher et al[13].

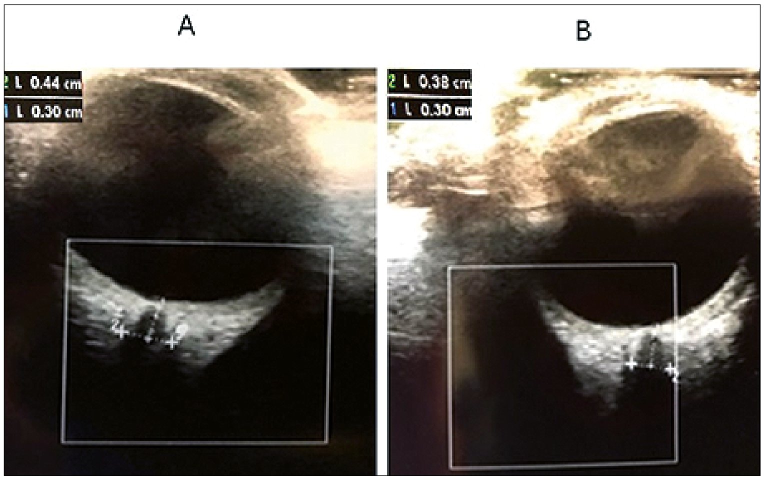

Figure 2. A) Average ONSD of 4.4 mm in supine decubitus, without headache. B) Average ONSD of 3.8 mm in standing position (14% collapse), after deambulation for 15 minutes, with intense headache.

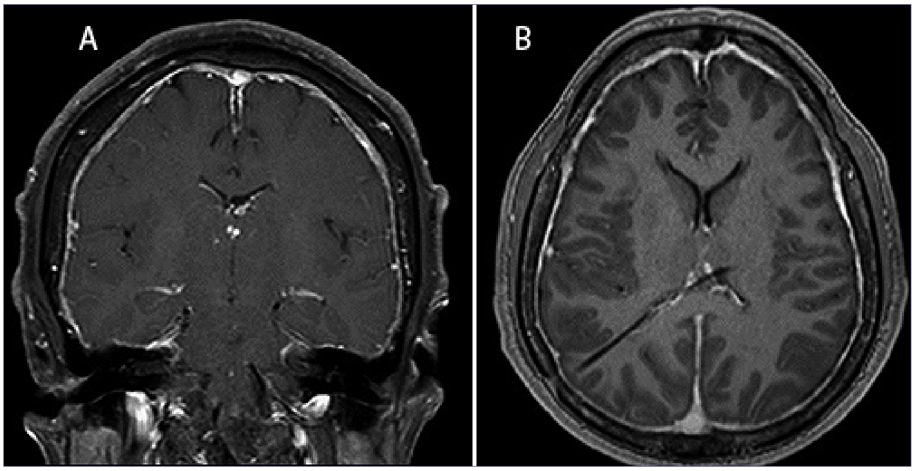

Figure 3. T1 weighted sequence that shows slit ventricles and pachymeningeal enhancement in shunt presence A) Coronal plane y B) Axial plane.

First measurement is done with the patient in supine decubitus position for at least 30 minutes and in absence of headache. In the next step the patient stands up and begins deambulation accompanied by a staff member. Intermediate measurements in standing position were performed in the presence of mild headache (VAS < 5) or associated symptoms (dizziness, photopsia, and others). A final measurement was done after 60 minutes or earlier in case of moderate to severe headache (VAS > o = 5).

Case Reports

Case 1

A 13 year old female with a history of tuberculous meningitis associated hydrocephalus treated 5 years ago with ventriculo-peritoneal (VP) shunt. Evolves with a 2 year recurrent progressive headache that interferes with daily living activities, including social and sports.

She has presented to the emergency department multiple times in which image study has shown no active hydrocephalus.

The headache is preceded with photopsia as a prodrome symptom and begins 10 to 30 minutes after physical activities, currently even after just walking. Initially, the pain is localized in the occipital area then migrates to bifrontal and resolves completely at supine position. No nausea/vomiting nor other neurological symptoms are reported by the patient. Physical examination is normal, valve refilling is slowed, ocular fundus shows no papilledema, CT imaging and MRI show slit ventricles, no active hydrocephalus signs, yuxtadural collections nor meningeal enhancement.

Ultrasound dynamic test of the ONSD was performed: ONSD is measured in supine position in which the patient is asymptomatic. When adopting an upright position, photopsia are referred after 5 minutes and headache starts progressively after deambulation. Ultrasound shows ONSD decrease greater than 10% at 15 minutes (Figure 2).

Shunt valve is replaced and ONSD ultrasound postural test is repeated showing no decrease on the fifth postoperative day. At 24 months follow-up, the patient is asymptomatic, resuming school and sport activities.

Case 2

A 47 year old male with hydrocephalus treated with VP shunt 20 years ago. Begins 10 years ago with progressive history of bifrontal headache preceded by dizziness starting 5 minutes after standing up. The headache starts gradually 30 minutes after deambulation. Physical examination is normal, shunt valve refilling is slowed, ocular fundus shows no papilledema. Brain CT imaging and MRI shows no active hydrocephalus, slit ventricles and small subdural bilateral fluid collections and significant pachymeningeal enhancement (Figure 3), suggesting intracranial hypotension. Ultrasound dynamic test shows decrease of the ONSD greater than 10% 35 minutes after adopting upright position, associated with intense headache.

Shunt valve is replaced with complete resolution of symptoms and normal ONDS postural test. At 24 month follow-up, the patient shows no headache but persists with occasional vertigo.

Case 3

A 33 year old male, history of myelomeningocele and congenital hydrocephalus treated with surgery during perinatal period. Last VP shunt replacement 18 years ago. Presents a 10 year duration headache in the occipital area preceded by vertigo and tinnitus when standing upright. The headache has been progressing significantly over the last 2 years to the point of being disabling after 60 - 90 minutes of standing upright. Multiple hospitalizations because of headache, chronic use of NSAIDS and opioids for pain control.

Neurologic exam shows spastic paraparesis (myelomeningocele sequelae), preserved sphincter control. Ocular fundus shows no papilledema, shunt valve refilling can't be evaluated because it's an old device system. Brain CT and MRI shows no significant change in ventricular size from previous ones, no active hydrocephalus, significant thickening of Skull and pachymeningeal enhancement that suggest intracranial hypotension. Ultrasound dynamic test shows decrease of the ONSD greater than 10% at 45 minutes with headache added.

Shunt valve system is replaced achieving complete postural symptom resolution with persisting mild, non postural, headache which its probably a rebound effect due to analgesic abuse. Late postoperative ultrasound test shows no sheath collapse. At 12 months, the patient shows complete resolution of postural symptoms, persisting in controls due to a residual component of headache associated with analgesic abuse.

It's worth noting that in all patients, a decrease in optic nerve diameter was observed minutes before the headache started, coinciding with other associated symptoms, but this decrease was less than 5%. In all 3 patients, a change to an adjustable valve system (Strata adjustable pressure valve by medtronic®) was performed. All patients had an acute episode of headache during the first 24 hrs post-surgery, which completely resolved when the valve drainage was regulated. In 2 of the 3 patients there was a 4% and 6% increase in ONSD in decubitus, as well as a discrete increase in ventricular size compared to preoperative findings. In all patients, the main symptom of postural headache was resolved.

Discussion

It is estimated that approximately 10% of patients with CSF shunt systems will present with overdrainage symptoms. Through invasive monitoring, Rekate showed intracranial pressures (ICP) of -25 to -30mmhg in symptomatic patients during the study of "slit ventricle syndrome"[16], confirming the presence of intracranial hypotension. Its clinical behavior is analogous to post lumbar puncture headache, characterized by headache that progresses during the day, worsens while the patient stands or performs activities, and improves rapidly when lying down in supine position[1-3]. It has been associated with the diagnostic triad: 1) postural headache; 2) small ventricles on imaging and 3) slow valve filling on palpation, not being specific for headache due to shunt related CSF hypotension[1,2].

The optic nerve ultrasonography is an emergent non-invasive technique whose clinical utility has been proven for intracranial hypertension diagnosis[6-11]. Hansen proved immediate correlation between ICP and ONSD with an intrathecal infusion test[17]. Maissan confirmed this correlation in traumatic brain injury[18]. In the same way, there are reports about real time response during lumbar punctures and ONSD[19-21], thus successively expanding the clinical scenarios for its use[26].

Although there are previous reports of optic nerve sheath collapse in MRI, and its usefulness in the diagnosis of spontaneous intracranial hypotension (SIH)[22,23,28], it was until 2011 that Baürele was the first to report the usefulness of ultrasound of ONSD in the diagnosis and follow-up of a case with SIH[12].

Fichtner proved the usefulness of the ultrasound of ONSD in a cohort with three groups of patients diagnosed with SIH: symptomatic, asymptomatic and a control group. It was shown that patients with SIH and active postural headache presented a decrease (collapse) of 10% of the EVNO value, when passing from horizontal to vertical position, with no significant difference between the other groups, marking a precedent for the test.

Continuing his research, Fichtner published in 2018 an excellent series of patients with SIH syndrome and objectified active CSF fistula refractory to conservative management protocol and requiring surgical resolution, confirming the usefulness of ultrasound measurement of ONSD for the diagnosis and follow-up of these patients[14]. As remarkable findings of this study it is worth mentioning: 1) In the post-surgical follow-up, the ONSD in horizontal position increased significantly (6%) with respect to the preoperative, which had also been noted by Dubost in patients treated with blood patch for post puncture headache[24]; 2) The "collapse" or postural decrease of the ONSD value when standing up, disappears in patients with resolution of symptoms. Both findings are in agreement with those found in our patients. This suggests that the presence of an active CSF fistula is necessary for the appearance of clinical symptoms[14]. There are studies based on continuous ICP monitoring that showed decreases up to -25 - -30 mmHg[3,29], which can be interpreted as an anti siphoning system failure. This can be compared to the presence of an active CSF fistula. In the same line, we propose that an active overdrainage mechanism in valvular systems is necessary, which we interpret in 3 phases during the performance of the ONSD dynamic ultrasonography test (Figure 1). First, in the horizontal phase, the patient is asymptomatic in the absence of activities that trigger CSF drainage. In a second or prodromal phase, the patient adopts a vertical position and there is an increase of drainage, which manifests by photopsia, dizziness or tinnitus, which may be transitory. At this stage, even without reaching severe levels of intracranial hypotension, it is possible to observe a partial decrease in ONSD, which generally does not exceed 5%. Finally, the third or symptomatic stage, persistent CSF drainage, causes a sufficiently low ICP decrease, characteristically defined as less than 6 cm H_2O[25], to generate symptoms of intracranial hypotension, which may manifest with different intensity. We believe that it is at this stage where a significant decrease in ICP can be objectified with the use of ONSD ultrasonography, manifested by a collapse greater than 10% with respect to baseline in the horizontal position. In the author's opinion, ONSD ultrasonography could play a relevant role in patients with CSF shunt systems, allowing to evaluate the presence of intracranial hypertension or hypotension. Although there are reports of its usefulness in obstructive shunt dysfunctions[27], this is the first report of the usefulness of the EVNO postural test in the noninvasive evaluation of CSF shunt related headache, opening an interesting field of development in the management of this complex clinical scenario.

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