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Confirmation of Elevated Intracranial Pressure in Arrested Hydrocephalus by Optic Nerve Ultrasonography

Arrest Hidrosefalide Artmış İntrakraniyel Basıncın Optik Sinir Ultrasonografisi ile Doğrulanması

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ABSTRACT

Introduction: Arrested hydrocephalus is a state of chronic hydrocephalus with mild dilatation of the ventricles in which the cerebrospinal fluid pressure has returned to normal, and there is no pressure gradient between the cerebral ventricles and brain parenchyma.

Case Report: In this case report, we present a 37-year-old female patient who was admitted to emergency department with complaints of headache, photophobia, and recurrent vomiting. She had been diagnosed with arrested hydrocephalus 5 years ago and was being followed radiologically since. Ocular ultrasonography was successfully utilized to assess and diagnose acute exacerbation of hydrocephalus.

Conclusion: Patients who are presumably assumed to have arrested hydrocephalus may experience an episode of hydrocephalus in any period of life. Ultrasonography can serve as a reliable tool in monitoring hydrocephalus; more specifically, sonographic visualization of the optic nerve sheath diameter may help to confirm increased intracranial pressure in such cases.

Keywords: Hydrocephalus, optic nerve sheath, ultrasonography
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ÖZET

Giriş: Arrest hidrosefali, beyin-omurilik sıvı basıncının normale döndüğü ve serebral ventriküller ile beyin parenkimi arasında basınç gradiyentinin olmadığı, hafif ventrikül genişlemesinin eşlik ettiği bir kronik hidrosefali durumudur.

Olgu Sunumu: Bu olgu sunumunda, acil servise başağrısı, fotofobi ve tekrarlayan kusma ile başvuran 37 yaşında bir kadın hastayı sunmaktayız. Hasta beş yıl önce arrest hidrosefali tanısı almış ve o zamandan beri radyolojik olarak takip edilmekteydi. Hidrosefalinin akut alevlenmesini değerlendirmek ve tanı koymak için göz ultrasonografisi başarılı bir şekilde kullanıldı.

Sonuç: Arrest hidrosefali farzedilen hastalar hayatlarının herhangi bir döneminde hidrosefali atağı geçirebilirler. Ultrasonografi, hidrosefalinin izlenmesinde güvenilir bir araç olarak görev alabilir, özellikle de, optik sinir kılıf çapının sonografik olarak görülmesi, bu vakalarda intrakraniyel basınç artışının doğrulanmasına yardımcı olabilir.

Anahtar Kelimeler: Hidrosefali, optik sinir kılıfı, ultrasonografi
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Introduction

In a sub-population of hydrocephalus patients, cerebrospinal fluid (CSF) pressure returns to normal, diminishing the pressure gradient between the cerebral ventricles and brain parenchyma to a chronic equilibrium. In this case, typically, ventricles are mildly dilated but without marked edema. This clinical entity is called 'compensated' or 'arrested' hydrocephalus (1). Such patients are challenging when presenting with a symptom or clinical sign of raised intracranial pressure (ICP). Common modalities to assess such patients may be limited. Computerized tomography (CT) of the cranium without contrast may not show significant changes from previous studies. In addition, radiation exposure associated with a lifetime of repeated CT scans may pose a

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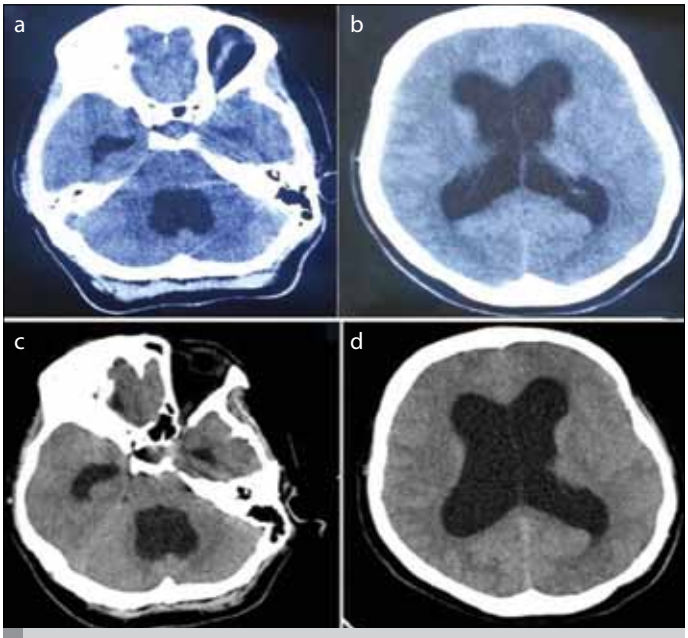

Figure 1. a-d. a, b) Dilated 3rd and lateral ventricles in the previous computed tomography obtained 2 years ago. c, d) Same dilatation of the 3rd and lateral ventricles is seen in the computed tomography obtained at the emergency department presentation

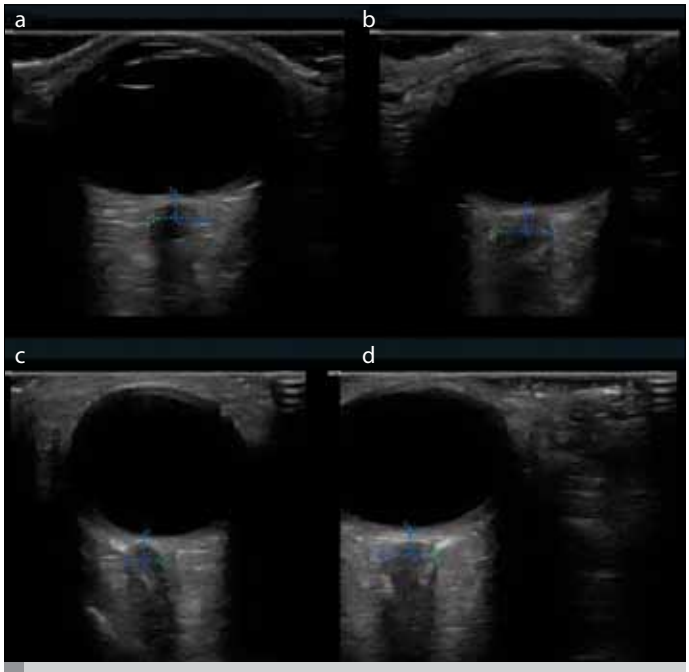

Figure 2. a-d. a, b) Bilaterally increased optic nerve sheath diameters measured sonographically. c, d) Bilaterally decreased optic nerve sheath diameters after the therapy

danger to the patient over the long run. In this case report, we aimed to show that measurement of optic nerve sheath diameter (ONSD) with ocular ultrasonography, which can be performed easily at the bedside by emergency physicians, can serve as a useful radiological adjunct in confirming elevated ICP in patients with arrested hydrocephalus.

Case Report

A 37-year-old female presented to the emergency department with complaints of headache, photophobia, and recurrent vomiting. Her symptoms had progressively worsened over the last few days previous to the presentation, such that on arrival, she reported the most severe headache of her life. She denied any history of trauma. The patient was being treated with oral ampicillin-sulbactam over the previous 10 days for an upper respiratory tract infection. Five years prior, she had been diagnosed with arrested hydrocephalus and was regularly followed radiologically without a CSF shunt placement.

On the physical examination, she was alert with a Glasgow Coma Scale score of 15. Her vital signs were as follows: blood pressure, 125/65 mm Hg; heart rate, 68 beats per minute; respiratory rate, 14 breaths per minute; and body temperature, 36.8°C. The rest of her examination was unremarkable. Of note, she was not found to have neck stiffness or lateralizing findings in the neurological examination.

In the emergency department, a cranial CT scan was obtained, which revealed enlarged ventricles but no increase when compared to the previous imaging studies (Figure 1). To detect if there was an elevated ICP, ONSD was measured with a 7.5 MHz transducer by bedside ultrasonography (Fazone CB, Fujifilm, USA), which revealed a mean ONSD of 6.5 mm (5.7 mm on the right and 7.4 mm on the left), which was compatible with elevated ICP (Figure 2 a, b). The optic disc was then evaluated by ophthalmoscopy, which showed papilledema with the optic disc borders obscured bilaterally. The patient's laboratory results were notable for an erythrocyte sedimentation rate of 90 mm/h, C-reactive protein of 4.8 mg/dL, and white blood cell count of 7.8 K/uL.

The patient was admitted to the hospital by a neurosurgeon for lumbar puncture and a ventriculoperitoneal shunt. She received dexamethasone 3 x 2 mg iv, furosemide 2 x 20 mg iv, and acetazolamide 2 x 250 mg po and was also given aminophylline 2 x 240 mg iv for headache. The patient was not administered mannitol, as she did not have radiological edema. Her symptoms dramatically resolved on the second day of hospitalization; so, the plan for lumbar puncture was abandoned. Her optic disc borders became clear at the end of the third day. As her headache and vomiting completely resolved, a ventriculoperitoneal shunt was not placed. There was no radiological difference on her repeat CT, which was obtained on the fifth day. She was subsequently discharged for out-patient follow-up. On her follow-up examination 1 month later, her repeat ONSD measurements were decreased compared to previous measurements (Figure 2 c, d).

Discussion

Arrested hydrocephalus is commonly seen in adult and geriatric patients but is rare in childhood and is most likely to occur in communicating hydrocephalus (1,2); in most cases, CSF shunt

placement is not indicated. In a study conducted on 46 children and adolescents with a diagnosis of arrested hydrocephalus, Whittle et al. showed that 88% of the patients had episodic or persistent intracranial hypertension (3). Our patient had not experienced elevated ICP before presenting to our emergency department.

When there is a clinical sign of elevated ICP in a patient with radiologically confirmed hydrocephalus and no difference between the previous imaging studies, normal pressure hydrocephalus, which is characterized clinically by a classic triad of dementia, gait disturbance, and urinary incontinence, in combination with an enlarged ventricular system and normal CSF pressure, should be accurately diagnosed and differentiated from arrested hydrocephalus. Because this clinical syndrome is potentially reversible by the placement of a ventriculoperitoneal shunt (4).

Various techniques have been proposed to estimate ICP, from clinical examination to invasive intracranial catheter (5). Sonographic measurement of ONSD has been proposed as a safe and useful predictor of raised ICP when compared to CT findings (6, 7, 8). The optic nerve sheath is a continuation of the dura; a significant increase in ICP is transmitted through the subarachnoid space to the optic nerve sheath and the optic nerve head. The resulting distension of the optic nerve head results in papilledema. Papilledema may lead to diagnostic difficulties in its different stages. It should be differentiated from optic neuropathies and structural abnormalities of the optic disc, known as pseudopapilledema (9). Papilledema can take several days to develop; therefore, it is not a sensitive marker of raised ICP in the acute setting. However, the retrobulbar aspect of the optic nerve sheath may undergo sonographically detectable distension seconds after an increase in ICP; thus, ONSD distension occurs faster than any other ophthalmoscopic finding. As a result, ocular fundus examination is not reliable for the evaluation of these changes during the initial hours (7).

There is no current standardized cut-off value of ONSD in diagnosing elevated ICP. However, the proposed values for predicting elevated ICP have been suggested, as follows: a mean ONSD of 4.5 mm in older children with hydrocephalus; 5.0 mm in adult patients with head injury, and 5.5 mm in critically ill patients due to varying causes (6, 7, 8). In comparison, our patient had a relatively large ONSD, with a mean of 6.5 mm, which subsequently decreased to 5.3 mm 1 month later after appropriate treatment. Despite the ONSD decreasing after treatment, it should be noted that it remained elevated above the normal range. This may be due to a number of factors. First, we measured ONSD only in the horizontal plane, not in the vertical plane as described in other studies. Second, our patient was not a trauma patient and can not be compared to the normal reference values of hydrocephalic children. Third, this may be an anatomic variation.

It should also be noted that upper respiratory tract infections, such as mastoiditis, otitis media or paranasal sinus infections, may disrupt CSF absorption by slowing blood flow in the cerebral venous system. Such patients may be predisposed to benign intracranial hypertension, also known as pseudotumor cerebri (10). Our patient had many characteristics typically seen with pseudotumor cerebri: her being female, demonstrating signs and symptoms of increased ICP following simple upper respiratory tract infection, and rapid resolution of symptoms. However, ventricular enlargement is not an expected finding of this entity. Increased resistance to CSF absorption or delayed absorption may minimally increase the pressure of CSF, with the occurrence of resulting symptoms. Such processes can often be seen without accompanying obstruction in major venous sinuses. Thus, the inflammatory process following upper respiratory tract infections is thought to have played a role in triggering the raised intracranial hypertension in this patient with arrested hydrocephalus.

Conclusion

Patients who are presumably assumed to have arrested hydrocephalus may experience an episode of hydrocephalus in any period of life. Since other methods either involve radiation or are invasive, they may not be undertaken as a routine procedure in the emergency departments. Thus, measurement of ONSD with ultrasonography may serve as a simple method to confirm elevated ICP in this patient population.

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