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Diagnostic value of the optic nerve sheath diameter in pseudotumor cerebri

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ABSTRACT

If persistent severe headache remains the only complaint of a patient, then the diagnosis of pseudotumor cerebri (PTC) can be delayed because in such cases practitioners are hesitant to immediately apply invasive intracranial pressure (ICP) measurement. Our purpose was to apply the technique of measuring diameters of the optic nerve sheath (ONSD) as a diagnostic tool in cases of PTC. Our aim was to provide practitioners with an additional sign to speed up their decision making about implementation of the lumbar puncture. In a retrospective study, CT scan data of 35 consecutive adult patients with PTC were collected and analyzed. ONSD were measured at the point where the ophthalmic artery crosses the optic nerve (anatomical landmark). The correlation analysis was performed with sex, age, and neuro-ophthalmological findings. We found that the ONSD was enlarged in 94.3% of patients with PTC. The enlarged ONSD were 6.2 ± 1.2 mm for the right and 6.3 ± 0.9 mm for the left (cut-off value >5.5 mm). The enlargement was bilateral, and no correlation with age or sex was found ($p = 0.67$ and $p = 0.76$, respectively). Presence of papilledema was detected in 91.4% of patients (32/35) presenting as a slightly less valuable diagnostic sign compared with ONSD. We conclude that in the majority of cases of PTC the ONSD is significantly enlarged, indicating elevated ICP even if CT scans are negative. Implementing this ONSD method as a diagnostic tool in cases of suspected PTC may help in early accurate diagnosis, avoiding misdiagnosis, and providing appropriate early treatment.

1. Introduction

Pseudotumor cerebri (PTC), also referred to as idiopathic intracranial hypertension or benign intracranial hypertension, is defined by the presence of pathologically elevated intracranial pressure (ICP) in the absence of dilated ventricles, intracranial mass lesion and other CT scan/MRI findings associated with elevated ICP, and normal composition of cerebrospinal fluid (CSF). The so-called Dandy criteria with further modifications add to this list headaches, nausea, vomiting, transient obscurations of vision that may evolve into mild vision loss, papilledema, and unilateral or bilateral abdecens nerve paresis [1,2]. PTC has often been considered a diagnosis of exclusion, especially if no neuro-ophthalmological pathology has been detected [2–5]. If a persistent and severe headache is the only complaint of a patient, then the diagnosis can be delayed because in such cases practitioners are hesitant to immediately apply invasive ICP measurement.

At the same time, if a patient is admitted to an emergency department complaining of a severe headache, as well as nausea and vomiting, then they are sent for a head CT scan by protocol. As a rule, these CT scans are negative, but we hypothesized that they can be additionally analyzed to detect raised ICP. Measurement of the optic nerve sheath diameter (ONSD) has been used as a non-invasive method of ICP monitoring since the mid-1990s. This method is based on the finding that the presence of enlarged ONSD indicates the elevated ICP [6,7]. This enlargement occurs because more CSF enters intraorbitally by filling subarachnoid space between the optic nerve and dura mater. Numerous reports indicate that ONSD is accurately measured by CT scan [8–11].

The introduction of an additional presenting sign might be a welcome addition to the above mentioned diagnostic criteria, because none of the signs are present in 100% of patients with PTC. Even the incidence of a headache varies from 80% to 90% of

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cases in various reports [2]. While papilledema was indicated as one of the leading signs in the 2013 revised diagnostic criteria for idiopathic intracranial hypertension, numerous reports have also indicated patients without this sign [12–15]. In addition to the distension of ONSD, the most recently proposed diagnostic criteria have included neuroimaging findings such as empty sella, flattening of the posterior globes, optic nerve head protrusion, tortuosity of the optic nerve, cerebellar tonsillar herniation, meningoceles, CSF leakage, and transverse venous sinus stenosis that could help in establishing the diagnosis of PTC [16,17]. At the same time, as the most recent review on the subject correctly concluded, "these findings are not diagnostic of idiopathic intracranial hypertension, and their presence is not required for the diagnosis of definite idiopathic intracranial hypertension" [16]. While the cerebellar tonsillar herniation can be diagnosed and the Chiari malformation and the CSF leakage can lead to the intracranial hypotension rather than hypertension, the distension of ONSD is indicative for the elevated ICP.

Lumbar puncture is used both for diagnostics and for treatment of PTC. However, in an emergency department, practitioners readily perform it in cases with suspected meningitis or subarachnoid hemorrhage. In cases when a patient does not present with fever, altered mental status, meningeal signs, or symptoms of hemorrhagic stroke and when headache remains the main complaint, practitioners tend to postpone this invasive technique until more presenting symptoms are collected by an ophthalmologist and neurologist. A signed informed consent should be obtained from the patient and a practitioner needs to present sound arguments to obtain the signature. As a rule, lumbar puncture is performed if neuro-ophthalmological pathology has been detected (by papilledema and changes to the visual field), if a headache has lasted for more than 3 months, and in the presence of additional signs such as nausea or vomiting.

Our aim was to implement the ONSD method for initial detection of elevated ICP in patients with PTC. While patients with this diagnosis usually have negative CT scans, we hypothesized that using the same initial emergency department CT scans for measuring ONSD might speed up the process of detecting elevated ICP and establishing the diagnosis.

2. Materials and methods

2.1. Study design and setting

In a retrospective study, we collected and analyzed the CT scan data of 35 adult patients (aged 18+ years) who were admitted to the Department of Radiology at our Medical Center from January 2011 to July 2015. The study protocol conformed to the ethical guidelines of the 1975 Declaration of Helsinki (amended 2000) as reflected *a priori* after approval by the institution's Helsinki committee. We examined CT scans of the patients who were admitted to the emergency department with complaints that included headache, nausea, vomiting, disturbance of vision, diplopia and dizziness in various combinations, and these were referred to the CT scan investigation of the cranial region, and appeared to have negative CT scans.

2.2. Inclusion and exclusion criteria

Severe headache of unknown origin and negative CT scans were the primary inclusion criteria; from these we selected patients that were diagnosed with PTC (*International Classification of Diseases Ninth revision Clinical Modicfication* 348.2 Benign intracranial hypertension). Patients with known ophthalmological or neuro-ophthalmological disorders were excluded.

During history taking, complaints which were usual for raised ICP (headache, nausea, vomiting, etc.) were detected and all patients were examined by a neurologist. By the Medical Center protocol, all patients with such complaints underwent initial non-contrast head CT scan. All patients with suspected PTC were examined by an ophthalmologist, even if they did not complain of disturbance of vision, obscuration of vision, or diplopia. Patients who complained of headache associated with tinnitus and dizziness were also examined by an otorhinolaryngologist.

2.3. Data sources and measurements

We collected and analyzed data on the following variables. (1) ONSD in the middle third of the intraorbital path (the point where the ophthalmic artery crosses the optic nerve served as an anatomical landmark); (2) presence/absence of papilledema; (3) sex, and (4) age of participants.

We analyzed CT scans obtained by the 256-slice CT scanner (Brilliance iCT, Philips Healthcare, Andover, MA, USA), with an initial single slice section of 3 mm and an area of interest slice section of 0.6 mm. The initial step in radiological differential diagnosis was to exclude any hemorrhagic or nonhemorrhagic lesions, contusions, brain swelling and space occupied lesions. If no pathology was found, then the ONSD were measured bilaterally by the computer program at the same CT scan (Fig. 1). Window parameters were spine window, middle third; window width 60 and window length 360 (also abbreviated as C:60,0. W:360,0 spine); and one pixel accuracy. All analyzed measurements were made using the same window, contrast and brightness. The error margin was expressed by means of the technical error of measurement (TEM) to calculate the intra-evaluator variability and inter-evaluator variability between two evaluators. The same equipment and methodological procedures for measurements were adopted by both evaluators (I.B. and T.S.).

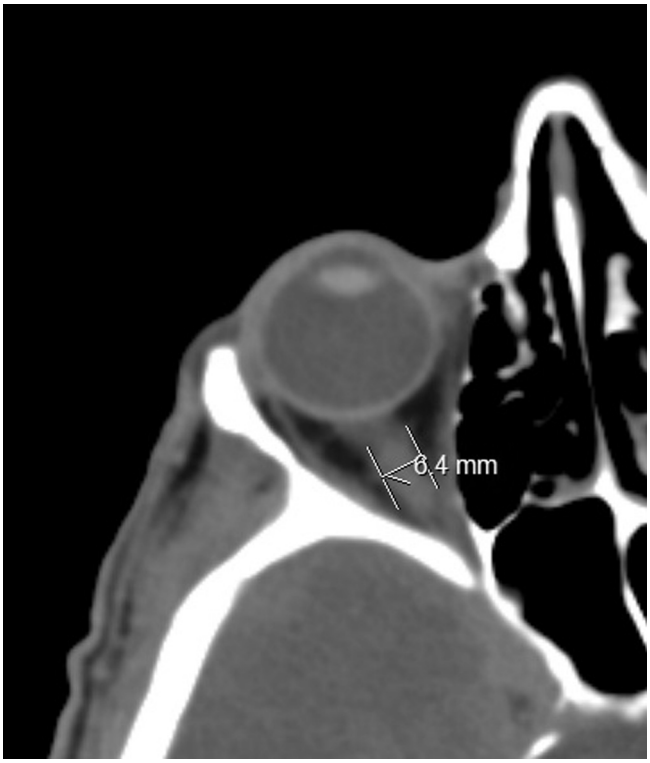

Fig. 1. The measurement of the optic nerve sheath diameter at a point where the ophthalmic artery crosses the optic nerve (the anatomical landmark) on transverse plain CT scan.