Bilateral optic disc drusen in a patient misdiagnosed as papilledema.

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Abstract

Optic disc drusen (ODD) are benign calcified deposits located at the head of the optic disc and are often visible within the optic nerve head. Most ODD patients are asymptomatic. In affected patients, the number and size of disc drusen are highly variable, and the drusen may be visible near the disc surface or buried within the disc, causing them to appear as pseudopapilledema. I report a case referred to our clinic with papilledema and a headache. She was previously misdiagnosed with papilledema and treated accordingly. However, she was still uncomfortable after treatment. After ophthalmologic examination and B-scan ultrasonography, she was diagnosed with ODD. We confirmed our diagnosis based on autofluorescence and spectral domain optical coherence tomography (SD-OCT), and she was subsequently diagnosed with pseudopapilledema and migraine.

Keywords: Optic disc drusen, papilledema, auto fluorescence.

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Introduction

Optic Disc Drusens (ODD) are asymptomatic benign a cellular calcified deposits located at the head of the optic disc and are often visible within the optic nerve head [1-2]. Previous studies have reported prevalence's of 0.34-2% [3-4]. ODD is more common in women, bilateral in 75-86% of cases and is generally asymmetric [5]. The pathophysiology of ODD remains unclear however, most investigators suggest that they are the final products of metabolic abnormalities of intra-axonal mitochondrial damage caused by disrupted cell axonal transport [6-7]. ODD can cause visual field defects, vascular complications (ischemic optic neuropathy, hemorrhage), and retinal detachment, and can mimic papilledema [8-10]. In some cases its often difficult to correctly diagnose buried optic nerve head and distinguish it from true papilledema. ODD usually follow a benign natural period but papilledema needs an emergent neurologic effort for potentially life-threatening conditions. Buried optic nerve head can be determined using various diagnostic tools including fundus auto fluorescence, fluorescein angiography, B- scan ultrasonography computed tomography and optical coherence tomography.

Case Report

A 35-year-old woman diagnosed with papilledema was referred to our ophthalmology department from a small-town state hospital's neurology clinic. She was also suffering from a history of headaches for three months. In the neurology department, a lumbar puncture was performed and the cerebrospinal fluid pressure was 174 mm H₂O. The patient was diagnosed with idiopathic intracranial hypertension and treated with oral carbonic anhydrase enzyme inhibitors. There was no pathology based on her cranial magnetic resonance imaging. After two weeks, she was still uncomfortable and referred to our hospital's neurology department.

During the ophthalmologic examination, her best corrected visual acuity was 20/20 in both eyes. Her pupils were isochoric and reacted equally, and there was no relative afferent pupillary defect. Anterior segment examination was unremarkable and intraocular pressures were within normal limits bilaterally. Based on fundoscopy, we noticed that both optic discs were swollen with a blurred margin but there were no cotton wool spots, multiple hemorrhages or swelling around the disc, hyperemia and venous congestion (Figure 1). To differentiate papilledema from ODD, ocular ultrasonography was performed calcifications at the optic nerve heads were observed, which is diagnostic of ODD (Figure 2). We also investigated the patient's ODD based on auto-fluorescence (Figure 3) and spectral domain optical coherence tomography (SD-OCT) (Figure 4). There were auto florescent deposits in both optic disc heads, and we observed hyaline calcific deposits in the disc line images based on SD-OCT testing. The patient was then diagnosed with pseudo papilledema due to ODD. The patient was also diagnosed with and treated for migraines by the neurologist [11].

Discussion

ODD are a cellular calcific deposits first described by Müller in 1858 [12]. Patients with ODD are commonly asymptomatic, and the drusen are generally discovered incidentally during ophthalmologic examination. ODD can be associated with ocular or systemic disorders, such as retinitis pimentos, pseudoxanthoma elasticum, angioid streaks, and Alagille syndrome [13]. Diagnosing ODD is often challenging based on clinical examination alone because its appearance can mimic other more common and concerning conditions, such as papilledema from elevated intracranial pressure caused by a space-occupying lesion, ischemic optic neuropathy, and optic nerve head tumors [14]. The primary clinical importance of ODD is that they can mimic true optic disc edema [15-16]. Misdiagnosing ODD as true papilledema may result in extensive, invasive and unnecessary workup for elevated intracranial pressure, including neuroimaging and lumbar puncture.



Figure 1: Fundus photo of the patient. Bilateral blurred margin of optic discs.



Figure 2: Hyper reflectivity of drusens in B-scan ultrasonography.



Figure 3: Autofluroscence of ODD.



Figure 4: Spectral domain optical coherence tomography section of drusens showing shallow subretinal hyporeclective space.

Patients with papilledema due to elevated intracranial pressure have hyperemic optic discs and swollen per papillary retina. Patients with soft exudates and per papillary hemorrhage more likely suffer from papilledema. Furthermore, spontaneous venous pulsations are absent in papilledema. Visual acuity is affected during the later stages of papilledema but is very rare for ODD.

In addition to ophthalmoscopy, ultrasonography (USG), computed tomography, fluorescein angiography, fundus auto fluorescence, and OCT are required for the correct diagnosis of ODD [17-19]. Our patient was misdiagnosed as papilledema because only ophthalmoscopy was performed by the neurologist. After fundus examination, we performed B-scan USG and investigated the hyper echoic region over the optic disc heads. This finding was suggestive of ODD. To confirm our diagnosis, we performed auto fluorescence testing and identified an auto fluorescent mass at the optic disc head. The OCT revealed the borders of the ODD. Savini et al. noticed that a structure called the Sub-Retinal Hypo Reflective space (SHYPS), which is located between the retinal pigment epithelium and the choriocapillaris, may be useful for differentiating between ODD and optic disc edema [20]. Johnson et al. suggested that the SHYPS thickness is greater in eyes with disc edema than those with ODD [21]. They noticed that the extravagated fluid from the optic nerve head, percolating into and elevating the sub retinal space, as the most reasonable cause for the increased SHYPS thickness. These authors characterized the OCT appearance of ODE as an elevated optic nerve head with a smooth internal contour and a SHYPS thickness under the optic nerve head with a gradient taper away from the disc.

Conclusion

In conclusion, ODD should be considered in all patients with blurred or elevated discs before being treated for papilledema. This article was prepared in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. From the experimental results it is found that the proposed method outperforms the state of art algorithm in terms of objective & subjective visual quality for Speckle noise in Optic al tomography images with Reasonable computational cost compared to BM3D for retinal images that is due to additional pre-filtering step at initial stage.

Conflict of Interest

The author declares no conflict of interest.

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