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UNILATERAL PSEUDOPAPILLEDEMA – CASE REPORT

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SUMMARY – Having sustained a head trauma, a 14-year-old boy presented with hematoma of the left upper eyelid and papilledema. Multi-slice computed scanning of the head and orbits showed a 2.6-mm oblique hyperdense mass behind the left optic disk. Ultrasonography of both eyes demonstrated unilateral buried disk drusen.

Key words: *Pseudopapilledema; Papilledema; Head trauma; Disk drusen*

Introduction

Unusual prominence and indistinct borders of the optic nerve head can be caused by a variety of diseases, but often, when not associated with any other abnormality, the pathogenesis remains unclear. In such cases of so called pseudopapilledema or pseudoneuritis, Rosenberg *et al*. described an unusual pattern with the optic disk subjectively reduced in size[1]. Pseudopapilledema is generally divided into cases caused by drusen of the optic disk and cases without optic disk drusen. In the majority of patients, pseudopapilledema occurs as an isolated phenomenon. In some patients, however, pseudopapilledema is associated with specific retinal disorder such as pigmented paravenous retinochoroidal atrophy[2], and in others it is part of a multisystem disorder such as Down's syndrome[3], Alagille syndrome (arteriohepatic dysplasia)[4], Kenny's syndrome (hypocalcemic dwarfism)[5], or linear sebaceous nevus syndrome[6]. Awan describes an association of pseudopapilledema and orbital hypotelorism[7]. Based on his clinical study in 3200 subjects, Lorentzen reports an incidence of optic disk drusen of 3.4 *per* 1000[8]. This prevalence increased by a factor of 10 in family members of patients with disk drusen, primarily because familial drusen are transmitted as an irregular autosomal dominant trait[9]. Men and women are equally affected, blacks less frequent than other races[10], and bilateral drusen occur in 67% to 85% of cases. Some drusen are easily visible with the ophthalmoscope; others are located anteriorly to the lamina cribrosa but are "buried" deep beneath the surface of the optic disk. Buried drusen may be detected by ultrasonography[11,12], computed tomography[13], or red-free photography[14]. Most patients with optic disk drusen are asymptomatic and remain so throughout life. Nevertheless, patients with optic disk drusen occasionally experience acute loss of central vision, peripheral vision, or both *via* a variety of different mechanisms[15].

Visual field defects are present or eventually develop in 71%-75% of eyes with disk drusen[16,17]. Loss of visual acuity is an extremely rare complication of optic disk drusen[18,19]. Prepapillary or peripapillary hemorrhages can develop in eyes with optic disk drusen[20]. Some patients with optic disk drusen develop serious maculopathy without hemorrhage. Drusen may also make the optic nerve more susceptible to other optic nerve diseases such as anterior ischemic optic neuropathy[21], glaucoma[22], and idiopathic intracranial hypertension[23]. There is no known treatment for the visual field loss associated with optic disk drusen.

The pathogenesis of optic disk drusen is unknown, but there are three classic theories. The first suggests that abnormal axonal metabolism leads to their formation[24]. The second attributes optic disk drusen to con-

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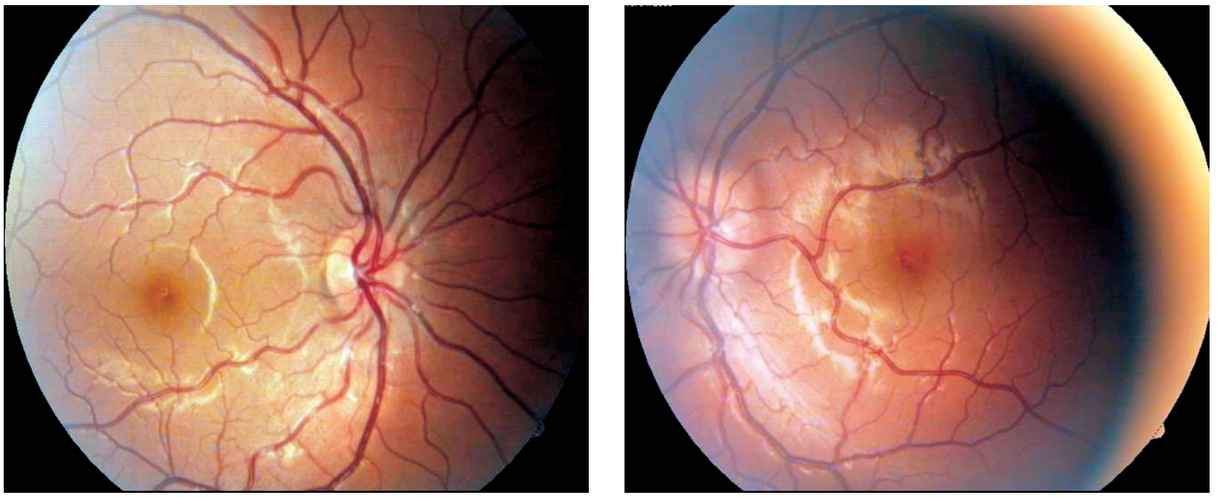


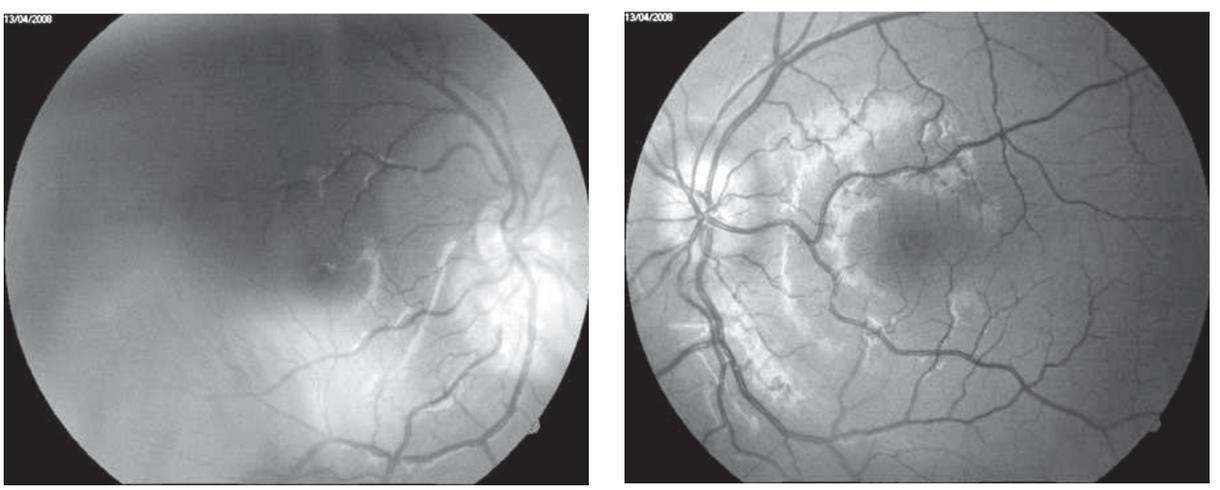
Fig. 1. Normal right fundus and blurred disk margins of the left fundus.

genitally dysplastic disks[25,26]. According to the last theory, patients with optic disk drusen have smaller-than-normal scleral canal[27,28]. Rigid edge of the scleral canal physically compresses optic nerve, blocking axoplasmic flow, leading to ganglion cell axonal damage and ganglion cell death. As the axons die, they extrude their mitochondria, which then serve as nidi for calcification.

Floyd *et al*. measured scleral canal using optical coherence tomography in patients with optic nerve drusen and concluded that the scleral canal area of unaffected eyes of subjects with unilateral drusen was not significantly different from the control[29]. The eyes of the first-degree relatives and eyes with optic nerve drusen both had scleral canal areas significantly larger than the control[29].

Case Report

A 14-year-old patient crushed the left side of his face against the back window of an automobile. He did not lose consciousness after the trauma. He was examined by a neuropediatrician on the same day, and had normal neurological status. The patient presented for ophthalmologic examination six days after the trauma because of a hematoma of his left upper eyelid. Visual acuity, intraocular pressure, bulbar motility, pupillary reaction and slit-lamp biomicroscopy were normal. Ophthalmoscopic examination of the right eye fundus was normal, but the left optic disk was elevated with blurred disk margins (Fig. 1).



Red-free right fundus; *red-free left fundus.*

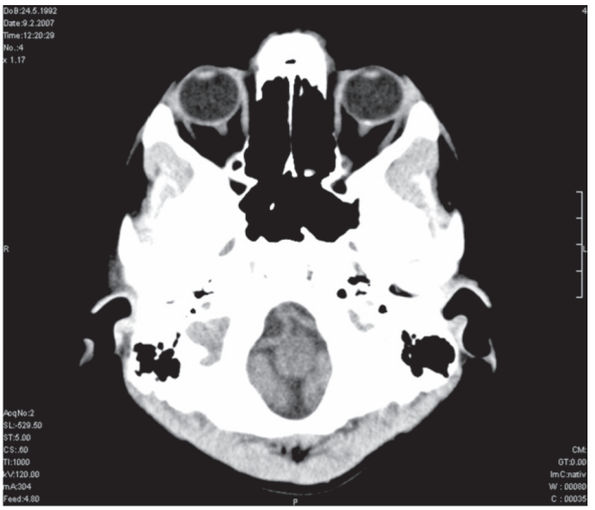


Fig. 2. Multi-slice computed tomography of the brain and orbits: oblique hyperdense mass behind the left optic disk.

Because of the trauma and the finding of the left-sided papilledema, the patient was referred for repeat neuropediatric examination and perimetry. These results were normal, so the patient was referred for multi-slice computed tomography (MSCT) scanning of the brain and orbits. MSCT showed a 2.6-mm oblique hyperdense mass behind the left optic disk, which was described as probably buried disk drusen (Fig. 2).

Ultrasonography confirmed deeply buried optic disk drusen of the left optic disk, which could not be seen on ophthalmologic examination (Fig. 3). The diagnosis of unilateral pseudopapilledema was made, however, coinciding with head trauma, thus implying some diagnostic uncertainty.

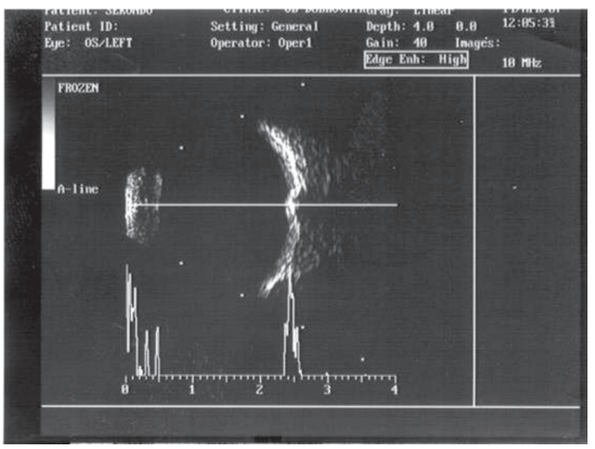


Fig. 3. Ultrasonography of buried optic disc drusen.

Discussion

The finding of elevated disk in a patient usually makes a diagnostic alarm. In our case, we encountered such a problem. We had a child with head trauma and elevated optic disk with blurred disk margins. Clinically, our patient was in good condition and had no signs of increased intracranial pressure but the fundus finding implied diagnostic uncertainty. Because his drusen were deeply buried we could not differentiate it by ophthalmoscopy as papilledema or pseudopapilledema.

Our next step was to recommend MSCT. In spite of the associated radiation exposure, it appeared justified because MSCT enables the brain and optic nerve fiber to analyze simultaneously. Having obtained the MSCT finding, we performed ultrasonographic examination and substantiated our diagnosis. Our diagnostic work-up yielded good results in this case.

In the literature there are different directions for the diagnosis of pseudopapilledema. Most authors differ as for the imaging technique preferable, i.e. CT or ultrasonography, because both can readily detect both visible and buried optic disk drusen[12,13]. However, the overall sensitivity of these noninvasive diagnostic studies in detecting buried optic disk drusen is almost 100%, so there is no reason to subject the patient with one or both optic disks elevated to lumbar puncture or other invasive tests until it is clear that the optic disks are truly swollen. In addition, some special photographic techniques such as red-free photography and fluorescein angiography can also be useful in detecting superficial or buried optic disk drusen[14].

Although unilateral pseudopapilledema caused by optic disk drusen is a rare condition, it should be considered on differential diagnosis even when it coincides with head trauma and has not been diagnosed before. However, one should not forget that patients with pseudopapilledema are not immune to neurologic and ophthalmologic disorders of the general population.

Conclusion

We present a patient with unilateral pseudopapilledema caused by deeply buried drusen and head trauma unassociated with the disease. History data are crucial in making the diagnosis; however, in the case presented history data led to diagnostic uncertainty because pseudopapilledema coincided with head trauma. Deeply buried drusen can be diagnosed in various ways, by use

of ophthalmoscopy, computed tomography, ultrasonography, magnetic resonance, fluorescein angiography, or optic coherent tomography. In most cases, papilledema can be differentiated from pseudopapilledema by ophthalmoscopy because in pseudopapilledema the physiologic cup is usually absent, the disk is not hyperemic, disk capillaries are not dilated, disk vessels are clearly seen, center is the most elevated portion of the disk, elevation is confined to the optic disk, anomalous vascular patterns are often present on the disk including an increased number of otherwise normal vessels, abnormal arterial and venous branchings, increased tortuosity, vascular loops, vascular shunts and cilioretinal arteries, and venous congestion is not present. Analyzing these details in a disk with indistinct borders, unnecessary tests can be avoided and definitive diagnosis made.

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Sažetak

JEDNOSTRANI PSEUDOPAPILOEDEM – PRIKAZ SLUČAJA

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Četrnaestogodišnji dječak je primljen s hematomom lijeve gornje vjeđe i papiloedemom nakon traume glave. Višeslojna kompjutorizirana tomografija glave i orbita pokazala je nagnutu masu od 2,6 mm, visoke gustoće, iza lijevog očnog diska. Ultrazvučna pretraga oba oka pokazala je jednostrane ukopane kvržice (*drusen*) diska.

Ključne riječi: *Pseudopapiloedem; Papiloedem; Trauma glave; Kvržice (drusen) diska*

timalen

timolol - maleat

kapi za oči

u koncentracijama

0,25% i 0,5%

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