

ial or syndrome association. There is a male-female predilection of 2:1. It has been described as freely mobile, well circumscribed, and subcuticular.

Although the clinical characteristics of fibrous hamartoma have been variably reported, it has unique microscopic features. All prior cases consistently report well-defined traversing bundles of dense fibrocollagenous tissue, immature loose-textured mesenchyme, and increased areas of interspersed mature adipose tissue.<sup>5</sup> It has been postulated that the mesenchymal tissue has vasoproliferative capability, which may explain the presence of an angioma involving the adjacent conjunctiva in this patient. In our case, the overall histopathologic pattern, with the focal basaloid budding overlying a circumscribed nodule, suggests the diagnosis of dermatofibroma.<sup>6</sup> However, the elements making up the tumor are much more characteristic of a fibrous hamartoma. Diffuse immunoreactivity for CD34 is suggestive of a solitary fibrous tumor. The negative reactivity to factor XIIIa<sup>7</sup> and muscle-specific actin rules out dermatofibroma and confirms the diagnosis of fibrous hamartoma of infancy. The basaloid hyperplasia overlying the fibrous hamartoma can be explained on the basis of activation of keratinocytes.<sup>8</sup> The presence of primitive mesenchymal cells in the underlying nodule may mediate the release of cytokines and growth factors that stimulate the keratinocytes. This leads to a cascade of events that may be responsible for the basaloid budding of the epidermis. A similar mechanism has been described in dermatofibroma.<sup>9</sup>

Other rare entities in the differential diagnosis include myofibroma, lipofibromatosis, and calcifying aponeurotic fibroma.<sup>10</sup> Myofibromas are found in the head and neck region and have light-staining areas and dark, more hemangiopericytoma-like staining areas histologically. The negative reactivity to muscle-specific actin in our specimen rules out myofibroma. Lipofibromatosis consists of abundant adipose tissue traversed by bundles of fibroblasts,

without immature mesenchyme. Calcifying aponeurotic fibroma is found interspersed with fat in infants and is composed of calcific areas surrounded by hyalinized collagen and fibroblasts.

The natural history of fibrous hamartoma suggests initial growth that slows with older age. No malignant degeneration or spontaneous regression has been documented. Local surgical excision is successful in most cases, with recurrent growth occasionally noted after incomplete excision.

Fibrous hamartoma is a rare, benign entity that occurs in infants and young children. It rarely involves the face. The lesion can be successfully excised, and its unique histopathologic characteristics are valuable in confirming the diagnosis.

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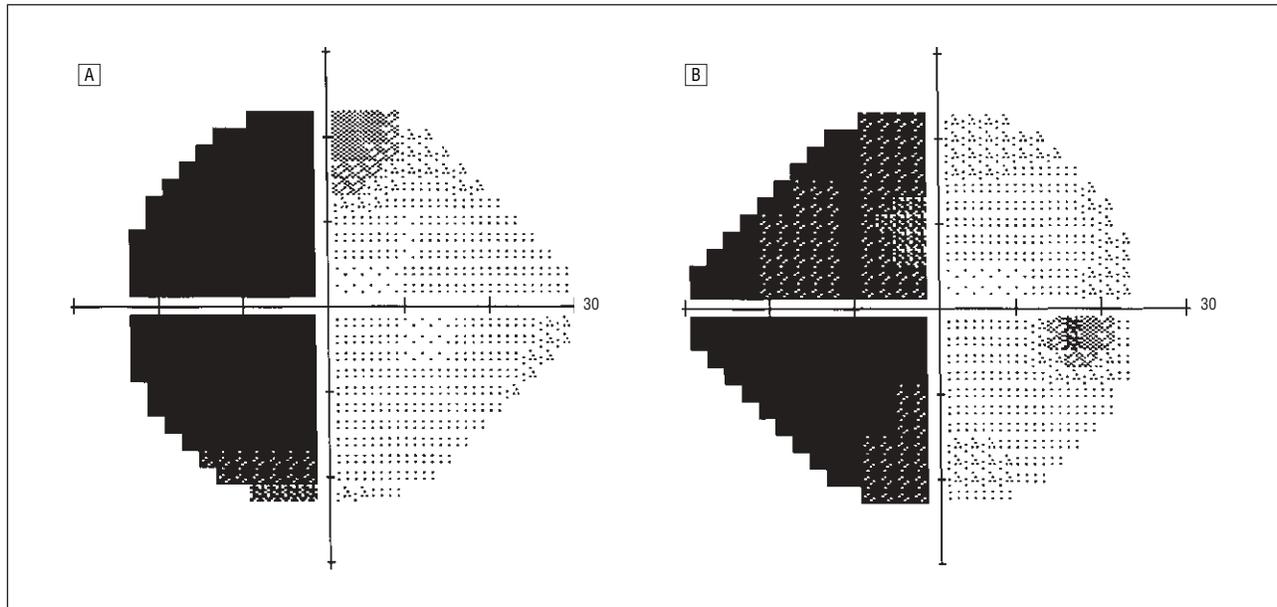
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## Transient Homonymous Hemianopia and Positive Visual Phenomena in Patients With Nonketotic Hyperglycemia

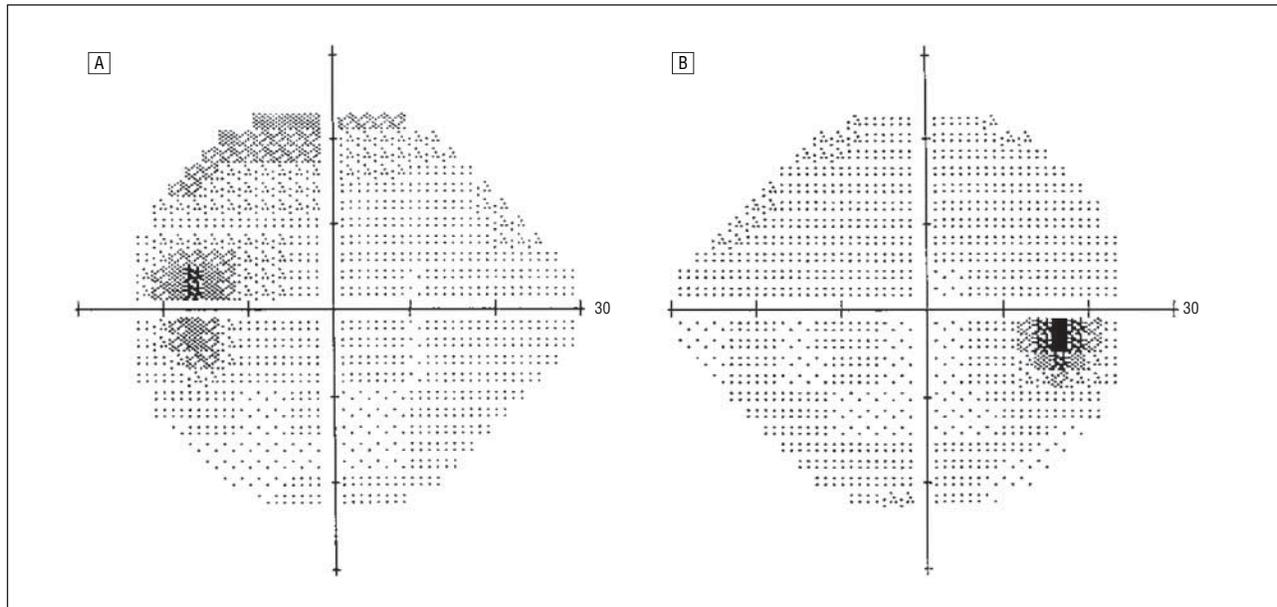
Homonymous hemianopic visual field defects usually result from structural processes affecting retinohypothalamic visual pathways. Cranial magnetic resonance imaging typically identifies the responsible lesions. Etiologies of homonymous hemianopias and normal neuroimaging include the Heidenhain variant of Creutzfeldt-Jakob disease, the visual variant of Alzheimer disease, occipital or global ischemia/hypoxia, MELAS (mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes), anemia, migraine, occipital seizures, functional illness, and nonketotic hyperglycemia (NKH).<sup>1</sup> Herein, we report a case of transient homonymous hemianopia and positive visual symptoms caused by NKH and review the literature on this rare phenomenon.

**Report of a Case.** A 68-year-old man had well-controlled type 2 diabetes mellitus (blood glucose levels consistently 90-130 mg/dL [5.00-7.22 mmol/L]). His physician changed his medication to insulin glargine in early December 2004, which resulted in poorly controlled blood glucose levels that were consistently more than 600 mg/dL (33.31 mmol/L) until early January. He developed intermittent photopsias, visual hallucinations, and "distorted" vision OU in the middle of December 2004. He denied having any other visual or neurologic symptoms.

Visual acuities were 20/50 OD and 20/40 OS. Automated perimetry revealed a complete left homonymous hemianopia (**Figure 1**). The rest of his neuro-ophthalmic examination findings were unremarkable except for nuclear sclerosis in the right eye and scleral buckle in the left.



**Figure 1.** Thirty degree Humphrey visual fields of left eye (A) and right eye (B). Note a dense left homonymous hemianopia is present during the period of uncontrolled hyperglycemia.



**Figure 2.** Thirty degree Humphrey visual fields of left eye (A) and right eye (B). Note resolution of the visual field defect after correction of hyperglycemia.

Cranial magnetic resonance imaging (including diffusion-weighted imaging, fluid attenuated inversion recovery, and gadolinium) and single-photon emission computed tomography scans showed normal results. Neuropsychological test results were unremarkable. His blood glucose levels improved to 100 to 150 mg/dL (5.55-8.33 mmol/L) and the hallucinations resolved 1 week later. At 2-week follow-up, automated perimetry showed resolution of the homonymous hemianopia (**Figure 2**). Electroencephalogram

was not obtained given the prompt resolution of symptoms after blood glucose levels improved.

**Comment.** “Stroke mimics” are unusual manifestations of nonvascular conditions that may resemble acute stroke symptoms and may result from metabolic, psychiatric, and central nervous system disorders (seizure, complicated migraine, tumor, myasthenia, and multiple sclerosis). Metabolic disorders represent a small subgroup (0.7% of mimics) of potentially treatable

causes of acute focal neurological deficits.<sup>2</sup> They include hypoglycemia, hyperglycemia, hyponatremia, hypoxia, uremia, and hepatic encephalopathy.

Seizures have been reported in 25% of patients with NKH; published reports emphasize partial motor seizures almost exclusively.<sup>3</sup> However, other clinical manifestations include hallucinations, myotonic twitches, nystagmus, tonic eye deviations, hemiparesis, hemisensory defect, aphasia, and homonymous hemianopia.<sup>3,4</sup>

**Table. Patients With Transient Homonymous Hemianopia and Positive Visual Phenomena Secondary to Nonketotic Hyperglycemia**

Source	Sex/Age, y	Blood Glucose, mg/dL	Symptoms	Imaging of Brain	EEG	SPECT of Brain	Duration	Resolution*
Taban et al (present study)	M/68	>600	Visual hallucinations, illusions	MRI: microischemic deep white matter changes	NA	Normal	6 weeks	Days
Freedman and Polepalle (2004) <sup>5</sup>	F/72	343	Visual hallucinations	MRI: normal	NA	NA	Several weeks	Several days
Brazis et al (2000) <sup>1</sup>	M/75	702	Visual hallucinations	CT: normal	Diffuse slowing	NA	1 week	1 day
Harden et al (1991) <sup>6</sup>	M/28	371	Visual hallucinations, focal seizures (left gaze deviation)	CT: normal	Ictal rhythmic waves in right posterior quadrant (during hallucinations)	NA	2 weeks	Several days
	F/67	452	Photopsias in hemianopic defect	CT: old lacunar infarcts	Ictal irregular waves in right occipital and parietal regions	NA	2 weeks	Several days
Johnson and Loge (1988) <sup>7</sup>	M/57	609	Visual hallucinations, palinopsia, motor seizures	CT: normal	Partial seizure from left occipital lobe (during hallucinations)	NA	5 days	2 days
Berkovic et al (1984) <sup>2</sup>	F/80	479	Expressive dysphagia, right hand weakness, finger agnosia, acalculia, agraphia	CT: normal; angiography: left carotid stenosis	Continuous high-voltage slow waves in left temporal region	NA	10 days	2-4 days
Maccario (1968) <sup>4</sup>	M/73	750	Visual hallucinations, right hemiparesis, stupor, seizures	Unknown	NA	NA	>1 week	With recurrence

Abbreviations: CT, computed tomography; EEG, electroencephalogram; MRI, magnetic resonance imaging; NA, not applicable; SPECT, single-photon emission computed tomography.

SI conversion factors: To convert glucose to mmol/L, multiply by 0.05551.

\*Resolution refers to time to normalization of homonymous hemianopia field defect after blood glucose control.

The patient herein developed transient homonymous hemianopia and positive visual phenomena secondary to NKH. We found only 7 other cases in the literature (**Table**). The average blood glucose level was 538 mg/dL (29.86 mmol/L) with a minimum of 343 mg/dL (19.04 mmol/L). There were 5 men and 3 women. All but 1 were older than 55 years of age. Half of the cases experienced visual symptoms only. The symptoms all resolved within days of controlling the blood glucose level. Although not completely understood, the pathophysiologic mechanism in some cases may occur from underlying seizure activity, as evidenced by electroencephalogram abnormalities.

When glucose concentration rises, water is osmotically attracted from the intracellular fluid space, resulting in cellular dehydration, alteration of enzyme activity, and subsequent neuronal dysfunction.<sup>3,4</sup> Additionally, most patients with focal seizures in NKH are elderly and

may have cerebral areas with borderline vascular supply. Given the stresses of hyperosmolality, incipient vascular ischemia in an area of borderline perfusion leads to cellular anoxia, clinically manifested in focal neurological deficits and focal seizures. This interaction between an area of potential dysfunction and the epileptogenic effect of hypertonic glucose solutions has been experimentally demonstrated.<sup>3,4</sup> In keeping with this hypothesis, Maccario<sup>4</sup> reported a recurrence of stereotypic neurological symptoms with hyperglycemia.

In conclusion, NKH should be considered in patients with homonymous hemianopia, positive visual phenomena, and negative neuroimaging studies. The mechanism is not completely understood but may represent ictal or postictal inhibition.

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