Eyelid Fibrous Hamartoma With Conjunctival Angioma in an Infant

Eyelid lesions are frequently seen in healthy infants. Common diagnoses include viral papilloma, chalazion, molluscum contagiosum, epithelial inclusion cyst, dermoid cyst, and capillary hemangioma. Less frequently recognized eyelid tumors consist of pilomatrixoma, apocrine hidrocystoma, pseudoepitheliomatous hyperplasia, neurofibroma, and choristoma. Characteristics of specific lesions, such as the presence of inflammation, overlying skin discoloration, and mass depth, are often useful in determining a clinical diagnosis.

Fibrous hamartoma of infancy is a benign soft tissue tumor that usually develops in the first 2 years of life and is not distinctive clinically. It has been reported most commonly in the axilla, abdomen, buttock, chest, and shoulder. We provide the first reported case, to our knowledge, of an eyelid fibrous hamartoma.

Report of a Case. A healthy 7-week-old girl underwent evaluation of a presumed left corneal limbal dermoid and left upper eyelid lesion present since birth. She was born at full term, and the results of her mother’s prenatal TORCH (toxoplasmosis, rubella, cytomegalovirus, and herpes simplex) workup were negative. The patient’s parents had noted good visual response to light and toys, with normal eye movements.

At examination, she demonstrated intermittent fixation bilaterally with a reproducible eye-popping reflex. She had full motility without nystagmus. Bilateral levator function was normal. External examination revealed a 5 × 4-mm white, dome-shaped lesion with a reddish base, located along the left medial upper eyelid between the eyelid margin and crease (Figure 1). There was no associated inflammation or distortion of the eyelid margin. The surrounding eyelid structures appeared normal. Additionally, there was an elevated, fleshy, vascular mass involving the inferome-
immunoreactivity was present for CD34 (Figure 4A). However, no reactivity was detected for factor XIIIa (Figure 4B) or muscle-specific actin (Figure 4C), confirming the diagnosis of fibrous hamartoma of infancy.

The conjunctival specimen consisted of nonkeratinized stratified squamous epithelium overlying a substantia propria–containing meshwork of vascular lumina lined by a single layer of endothelium (Figure 5). This was suggestive of an angioma.

Comment. Fibrous hamartoma of infancy was first described by Reye in 1956. It is a benign soft tissue tumor that can develop at birth and occurs primarily on the trunk, axilla, or buttock. Rarely has it been found on the face and, to our knowledge, never involving the eye. The vast majority are benign solitary lesions and range from 0.5 cm to 4.0 cm in diameter, with no evidence of famil-
activation of keratinocytes. The can be explained on the basis of infancy. The basaloid hyperplasia muscle-specific actin rules out derm. The negative reactivity to muscle-specific actin in our specimen rules the diagnosis of dermatofibroma. 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 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. 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.

Other rare entities in the differential diagnosis include myofibroma, lipofibromatosis, and calcifying aponeurotic fibroma. 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.

Correspondence: Dr Bradfield, University of Wisconsin, Department of Ophthalmology and Visual Sciences, 2870 University Ave, Suite 206, Madison, WI 53705 (ysbradfield@ophth.wisc.edu).

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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 retrochiasmal 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 non-ketotic hyperglycemia (NKH).

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.