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Collagenoma of the Eyelid

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Abstract

We present the rare case of an eyelid lesion comprised of hamartomatous dermal collagen, known as a collagenoma. Collagenomas may be sporadically acquired, or inherited as part of numerous autosomal dominant syndromes. In the appropriate clinical context, their diagnosis should prompt a thorough review of systems, systemic examination, and inquiry into family history, to assess for underlying autosomal dominant syndromes. Recognition of collagenomas may thus allow diagnosis of inherited syndromes, allowing patients to obtain appropriate genetic counseling, as well as screening and treatment of associated systemic pathology.

The differential diagnosis for eyelid lesions is broad as lesions may derive from any tissue layer of the eyelid, from skin to conjunctiva. Lesions of the skin may derive from epidermal elements, with examples including squamous papillomas and carcinomas. Lesions may also arise from dermal elements. Connective tissue nevi, for example, are benign, circumscribed hamartomatous lesions classified according to the predominant dermal connective tissue component comprising them: collagenoma (collagen), elastoma (elastin), and nevus mucinosis (proteoglycan).¹ Here, we present the rare case of a collagenoma manifesting on the upper eyelid of a 77 year old woman. This report is compliant with the Health Information Portability and Accountability Act and the tenets of the Declaration of Helsinki.

Case Report

A 77 year old Caucasian female presented for evaluation of a painless right upper eyelid lesion present for eight years, as well as bilateral upper eyelid dermatochalasis limiting her superior field of vision. The patient did not notice any recent growth of the lesion, and it measured 4 mm in diameter during a dermatology examination eight years prior to presentation.

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The patient's ocular history was notable for dry eyes, treated with cyclosporine 0.05% ophthalmic solution bilaterally twice daily, a single episode of right eye optic neuritis three decades previously, and uncomplicated cataract extractions of both eyes eight years prior to presentation.

Her medical history was notable for endometrial carcinoma ten years prior to presentation, complicated by liver metastases treated with chemoradiation, and maintained on anastrozole. The patient also had a history of multiple lower extremity deep venous thromboses for which she took apixaban. She was a non-smoker and consumed 1 alcoholic drink daily. The patient's family history was notable for a brother with multiple sclerosis and a father with bladder cancer.

On examination, best corrected visual acuity was 20/25 in the right eye and 20/20 in the left eye. Intraocular pressure was 11 mmHg in the right eye and 13 mmHg in the left eye. Pupils were equal and reactive. External examination was notable for a 5 mm × 5 mm firm, non-erythematous, non-tender nodular lesion of the central right upper eyelid (Fig. 1A). Clinically, the lesion was only slightly mobile, and was thought to reside subcutaneously with dermal attachments. The patient also had dermatochalasis of both upper eyelids and bilateral brow ptosis. Anterior slit lamp examination was unremarkable. Posterior examination was remarkable only for stable trace optic nerve pallor of the right eye related to the patient's past episode of optic neuritis, a small stable chorioretinal scar of the right eye, and small drusen in both eyes.

A surgical plan was made to excise the right upper eyelid lesion via an internal approach, and perform bilateral upper blepharoplasty. Risks, benefits, and alternatives were discussed with the patient, who elected to proceed.

Intraoperatively, the right upper eyelid lesion was everted over a chalazion clamp. As conjunctiva and tarsus were incised, it became apparent that the lesion was circumscribed and mildly adherent to anterior tarsus. Dissection proceeded along anterior tarsus to sharply dissect the lesion from tarsus, with eventual delivery of a 5 mm firm, tan, intact mass. Delivery required sharp dissection of the lesion from tarsus, but the lesion did not invade tarsus, and it was removed completely without the removal of any tarsus. Bilateral upper blepharoplasty was then performed in standard fashion.

Hematoxylin and eosin staining of the lesion showed an unencapsulated dermal nodule composed of thickened collagen bundles in a storiform pattern with prominent intervening clefts, promoting a diagnosis of a storiform collagenoma (Fig. 1B–C).

Discussion

Connective tissue nevi represent benign, circumscribed hamartomatous lesions of the dermis. They are typically classified as a collagenoma, elastoma, or nevus mucinosis based on the predominant comprising connective tissue component (collagen, elastin, or proteoglycan, respectively).¹

Collagenomas, also known as sclerotic fibromas, present as asymptomatic, well-circumscribed pink, white, or flesh-colored papules or nodules that histopathologically demonstrate prominent hypocellular storiform collagen bundles separated by clefts.² The differential diagnosis of collagenomas includes other well-circumscribed skin lesions such as dermatofibromas, fibrolipomas, and benign fibrous histiocytomas.³ There is still debate as to whether collagenomas may merely represent involutional sclerotic stages of these and other cellular skin lesions, rather than a unique class of proliferating neoplasm.⁴

Collagenomas may develop as part of various autosomal dominant disorders.¹ Familial cutaneous collagenoma typically presents in young adulthood with development of multiple collagenomas on the torso, and is associated with cardiac structural and conduction abnormalities.⁵ Buschke-Ollendorff syndrome is characterized by collagenomas, typically located on the extremities, as well as osteopoikilosis.⁶ Collagenomas are also present as the leathery, flesh-colored, typically lumbosacral, shagreen patches seen in tuberous sclerosis.⁷ Collagenomas have also been associated with multiple endocrine neoplasia type 1, in which they are present in approximately two-thirds of patients and are typically located on the torso and neck.⁸ Finally, collagenomas are characteristic of Cowden syndrome which in addition to mucocutaneous lesions, carries an increased risk of breast, thyroid, gastrointestinal, and endometrial neoplasia.⁹ Our patient had no history or physical findings consistent with an inherited disorder.

Collagenomas can also present in isolation and sporadically. Isolated collagenomas typically present in middle age and involve the extremities or face,² but can rarely involve the palm, sole, or oral mucosa.¹⁰ Eruptive collagenoma is an acquired condition that presents with the sudden symmetrical appearance of many small collagenomas, typically during adolescence and young adulthood.¹¹

The collagenoma in this case was likely an isolated collagenoma given the patient's advanced age at onset, and her lack of signs, symptoms, or family history of collagenoma-associated autosomal dominant disease. To our knowledge, isolated collagenoma of the eyelid has been reported only once previously.² Because collagenomas may be a part of an underlying inherited disease, it is important to characterize their clinical and surgical appearance. Tissue diagnosis in the appropriate clinical context may prompt further work up, screening, and genetic counseling for patients and family members.

This case of an eyelid collagenoma expands upon our current knowledge by describing the external appearance, examination characteristics, and intraoperative behavior of this very rare eyelid neoplasm.

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Fig.1.

A, External photograph of the right upper eyelid lesion, in which the elevated contour of the lesion (arrow) can be appreciated even with obscuring dermatochalasis. Hematoxylin and eosin stain of the eyelid lesion (**B**, 40x magnification. **C**, 200x magnification) demonstrated an unencapsulated dermal nodule composed of thickened collagen bundles in a storiform pattern with prominent intervening clefts.