

laser therapy for oral lichen planus. *Lasers Med Sci.* 2014;29:185–90.

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## Blepharochalasis: A Rare Presentation of Cutis Laxa<sup>☆</sup>



### Blefarocalasia: una presentación rara de cutis laxo

To the Editor:

Cutis laxa is a congenital or acquired connective tissue disorder characterized by extracellular matrix and elastic fiber defects. It manifests clinically as sagging skin, giving an appearance of premature aging. Blepharochalasis is an acquired form of cutis laxa that affects the periorbital region. It normally starts in childhood or adolescence and is characterized by recurrent episodes of painless eyelid swelling. Repeated acute attacks lead to elastin fiber degradation and sagging skin in the periorbital region that can interfere with vision. The pathogenesis of blepharochalasis or blepharoptosis is still unclear, although a role has been proposed for immunopathogenic mechanisms that result in elastin fiber degradation.<sup>1,2</sup> Grassegger et al.<sup>1</sup> and Schaeppi et al.<sup>2</sup> reported immunoglobulin A (IgA) deposits around blood vessels and sweat glands and in the papillary dermis. The second group of authors also found IgA deposits in elastic fibers around hair follicles. Histopathology shows disperse fragmented, granular elastic fibres in the reticular dermis.

The differential diagnosis in blepharochalasis should include inflammatory diseases (contact dermatitis, angioedema, blepharitis, and hereditary angioedema), tumors (retrobulbar tumor or lacrimal gland tumor/cyst), systemic diseases (thyroid and kidney disease), other extracellular matrix diseases (Ehlers-Danlos syndrome, elastic pseudoxanthoma, mid-dermal elastolysis, anetoderma, and postinflammatory elastolysis), and even physiologic aging. The condition can also form part of syndromes, such as Ascher syndrome, which is characterized by blepharochalasis, double lip, and nontoxic goiter.

The treatment for blepharochalasis is reconstructive surgery with cosmetic and therapeutic intent, but recurrence is common.<sup>3</sup>

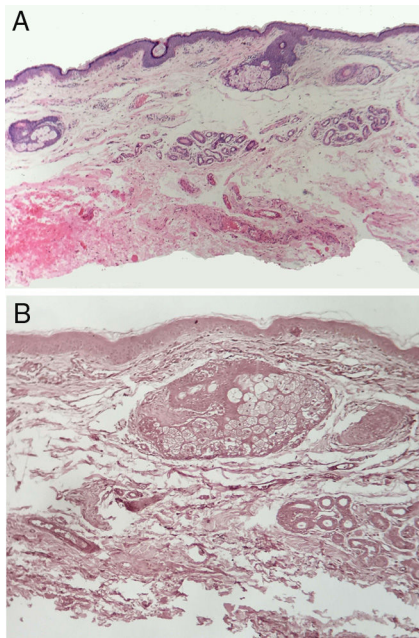
In this article, we describe the case of a young man with blepharochalasis as a clinical presentation of acquired cutis laxa.

A 24-year-old man presented with looseness of the upper eyelids that had become progressively worse over 8 years. The sagging was such that it had caused considerable cosmetic damage. The patient reported that he had experienced several episodes of eyelid swelling over a period of more than 2 years before his eyelids began to sag. He had not received any treatment and reported no other problems. Skin examination showed loose skin on both upper eyelids (more pronounced on the right) and partial occlusion of the eyelashes. The skin in the upper eyelid region was atrophic and had a violaceous color (Fig. 1). Vision was not affected by the blepharoptosis. The patient confirmed that the condition had not been treated medically. Biopsy of the eyelids (with hematoxylin-eosin staining) showed just slight swelling of the upper dermis. Orcein staining showed absence of elastic fibers (Fig. 2). IgA was positive in the dermal vessel walls. Computed tomography of the orbits, a chest X-ray, and laboratory tests were all normal. The patient underwent upper blepharoplasty (Fig. 3) and the cosmetic and func-



Figure 1 Eyelid redundancy.

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**Figure 2** Histopathologic study of eyelid skin. A, The only remarkable finding was mild edema in the upper dermis (hematoxylin-eosin, original magnification  $\times 40$ ). B, Absence of elastic fibers in the eyelid dermis (orcein, original magnification  $\times 100$ ).



**Figure 3** Excellent results after blepharoplasty.

tional results during follow-up were excellent. He reported that his field of vision had improved, particularly the upper field as his eyes were no longer covered by the sagging eyelids.

Blepharochalasis is an uncommon clinical presentation of cutis laxa characterized by painless episodes of bilateral edema.<sup>4</sup> The condition is generally self-limiting and does not respond to antihistamines or corticosteroids. During the quiescent stage of disease and following several acute attacks, it progresses to cause bilateral loss of periorbital skin.<sup>5</sup> There may be violaceous coloring on the upper eyelids.<sup>6</sup> In our patient, this coloring led to consideration of dermatomyositis in the differential diagnosis.

The pathophysiology of blepharochalasis remains elusive. Inflammatory episodes are believed to give rise to elastic fiber degradation due to elastolysis triggered by increased elastase activity or impaired elastase inhibitor function.<sup>3</sup> Systemic manifestations have not been observed in the vast majority of cases.<sup>7</sup> In line with previously described findings, IgA deposits, which probably have a pathogenic role, were observed in the blood vessel walls.<sup>1</sup>

Proposed triggers include stress, fever, and upper respiratory tract infections, among others.<sup>3</sup> A more specific immune mechanism associated with elastin and collagen degradation by metalloproteases may also be involved.<sup>1</sup>

The recommended treatment is blepharoplasty with cosmetic or therapeutic intent. Ideally, surgery should be performed during the quiescent disease stage.<sup>8</sup> Unlike most connective tissue diseases, cutis laxa does not usually interfere with postoperative recovery.<sup>9</sup> Patient follow-up is necessary, however, given the risk of recurrence.<sup>10</sup>

Blepharochalasis must be contemplated in the differential diagnosis of skin disorders that affect the periorbital region. Correct diagnosis is important for ensuring adequate treatment and follow-up.

### Conflicts of Interest

The authors declare that they have no conflicts of interest.

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## Osteonevus of Nanta: A Rare Skin Condition <sup>☆</sup>



### Osteonevus de Nanta, un fenómeno cutáneo poco habitual

*To the Editor:*

Osteonevus of Nanta is a rare condition characterized by osseous metaplasia in an intradermal nevus.<sup>1</sup> The lesion was first described by Heidesfield in 1908, and in 1911, it was reported in a publication by French dermatologist André Nanta.<sup>2</sup>

Bone formation in the skin is uncommon and can be primary (when there is no evidence of a pre-existing lesion) or secondary to an inflammatory and/or neoplastic process.<sup>3</sup> Secondary bone formation has been reported in a range of lesions, including pilomatrixoma, basal cell carcinoma, acne, pyogenic granuloma, and dermatofibroma. Ossification of an intradermal nevus, however, is very rare.<sup>1,4</sup>

A 38-year-old man with no remarkable history presented with a hyperpigmented lesion on his left cheek that had grown and become progressively harder with time. Physical examination showed a hard, black nodule that measured 1.5 cm in diameter and was not painful on palpation (Fig. 1). The lesion was fully excised and a sample sent for evaluation. Histologic examination showed nests of nevus cells with appropriate maturation in the superficial dermis and, underneath, in the deep dermis, bone marrow trabeculae containing osteocytes (Fig. 2). Signs of intramedullary hematopoiesis and mature adipocytes were observed in the center (Fig. 3). The lesion was diagnosed as osteonevus of Nanta and the patient was scheduled for regular follow-up.

Primary cutaneous bone formation has been described in Albright hereditary osteodystrophy, progressive osseous heteroplasia, myositis ossificans progressiva, and osteoma cutis.<sup>4</sup> Secondary cases, in turn, have associated with scars, pyogenic granuloma, epidermal cyst, fibroxanthoma, and lipoma, and there have also been reports at the site of trauma or injection.<sup>3,4</sup> Rarer associations include burns, dermabrasion, stasis dermatitis, and

cutaneous metastases from breast, bladder, or bronchial cancer.<sup>5</sup> Benign tumors, and melanocytic nevi in particular, are the most common causes of secondary osteoma formation.<sup>3</sup>

Clinically, osteonevus of Nanta resembles an intradermal nevus, is more common in women,<sup>6</sup> and tends to be located in the upper part of the body, in particular the face, suggesting a potential pathogenic role for repeated hair follicle trauma and chronic inflammation.<sup>3,7</sup> Lesions with necrosis, bleeding, and tissue regeneration could affect physical and chemical factors, such as calcium and phosphorous ion concentrations, pH, oxygen levels, and enzyme activity.<sup>3</sup> These factors could induce a granulomatous reaction, triggering the transformation of mesenchymal cells into osteoblasts and resulting in



**Figure 1** Black nodule with a diameter of 2 cm on the left cheek. Note the ill-defined borders and hair follicles.

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