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Unilateral blepharochalasis

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SUMMARY. Unilateral blepharochalasis is an extremely rare disorder with an unknown etiology and pathogenesis. The authors present a 22-year old patient in whom a right-sided skin overhang of the upper eyelid caused visual field impairment. The condition was corrected by a standard blepharoplasty. The histological examination suggested a localised anomaly of the lymphatic system and an almost complete absence of elastic fibres as the causative agent. © 2003 The British Association of Plastic Surgeons. Published by Elsevier Science Ltd. All rights reserved.

Keywords: blepharochalasis, unilateral blepharoplasty eyelids.

Blepharochalasis, a term introduced by Fuchs[1] in 1892, is a rare idiopathic disorder affecting the upper eyelids, but additionally the lower eyelids may be involved in more severe cases. The disorder usually presents bilaterally, although a few unilateral cases have been reported.[2–5] The condition typically manifests itself in adolescence and young adulthood with intermittent, recurrent bouts of a painless oedema and redness of the lids. As a result of these repeated attacks the eyelid skin becomes thinned, stretched and often there is lacrimal gland prolapse and orbital fat herniation. Ultimately, the skin hangs down over the lid margin and may lead to a mechanical visual obstruction accompanied by physiological dysfunction. We present here the extremely rare case of a young man with unilateral blepharochalasis, a condition that was caused by a localized lymphatic dysfunction.

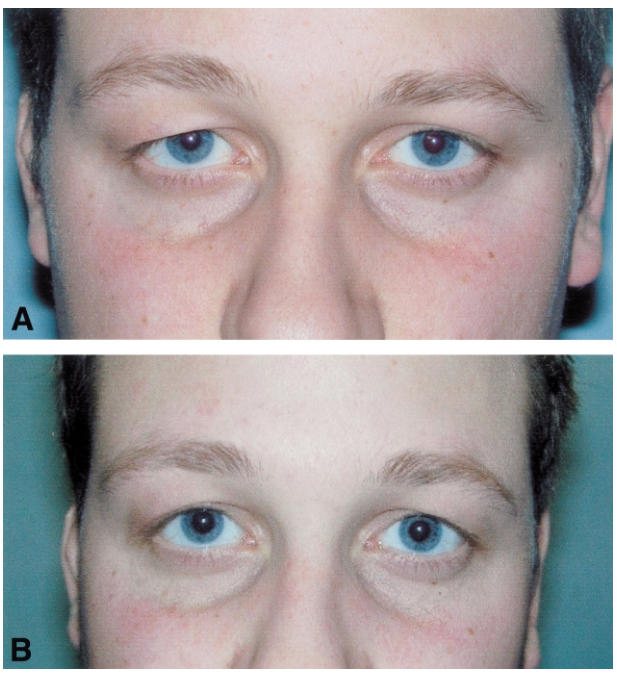

Figure 1—(A) Preoperative appearance of the patient with marked right-sided, unilateral blepharochalasis. (B) Postoperative result with good symmetry at ten weeks.

Case report

A 22-year-old male presented to the plastic surgery department with a moderate blepharochalasis of the right upper eyelid, but with involvement neither of the right lower eyelid nor the lids of the left eye (Fig. 1(A)).

The patient reported that since about the age of 12 he had been suffering from recurrent episodes of eyelid oedema and to his knowledge there was no known inciting agent or a direct or indirect trauma, which would explain a causal relationship. Interestingly, the patient experienced an aggravation of the swelling after physical exhaustion, in particular sporting activities.

He has no known allergies and never had any evidence of urticaria. In fact, allergy tests such as a routine screening for seasonal and perennial allergens proved negative. There is also no family history of angioneurotic oedema or swelling of the lids. Initially he was not bothered by this condition but the

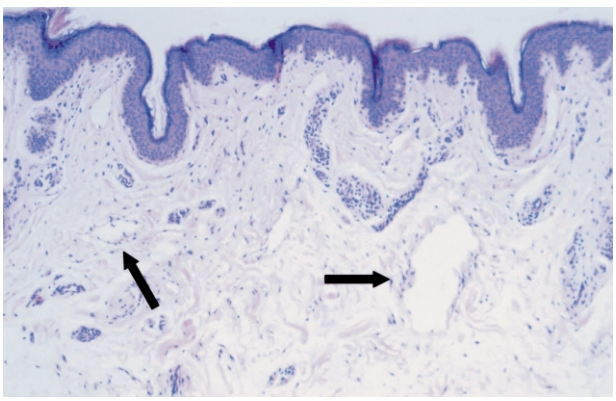

Figure 2—Skin of right upper eyelid demonstrating an increase of the number and size of lymphatic vessels (arrow) and a moderate perivascular infiltrate of lymphocytes. (hematoxylin and eosin stain, × 40).

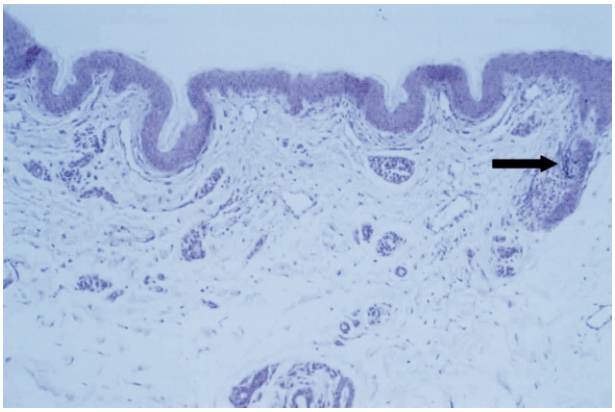

Figure 3—The skin of the right upper eyelid also demonstrates an almost complete absence of stainable elastic fibres. The arrow indicates a few remnants of perifollicular elastic fibre bundles. (elastica stain, × 40).

swelling got progressively worse with more frequent episodes and a beginning disturbness of eye function. The patient then consulted an ophthalmologist about two years ago. There was a noted effort to open the upper lid and a beginning restriction of the visual field in the two upper quadrants.

Surgery was performed on an outpatient basis to correct the blepharoptosis using a standard supratarsal crease incision. No resection of the aponeurosis was undertaken. The excess skin was excised and the lid fold recreated. The removed skin portion was submitted for histopathological examination.

The immediate postoperative result was satisfactory. After one month the patient developed a mild recurrence of the swelling in the lateral aspect of the left upper eyelid, requiring a second intervention. Eventually the patient was seen again after three months. The aesthetic result was then very pleasing and the symptoms of the visual field impairment were corrected (Fig. 1(B)).

Histopathology

A routine hematoxylin-eosin stain was done on the operative sections of the skin of the right upper eyelid (Fig. 2). There was mild oedema with increased capillary vascularity. The lymphatic vessels showed a moderate dilatation and additionally a spotty perivascular infiltrate of lymphocytes was present.

Additionally an elastica stain demonstrated a rarefaction of elastic fibres (Fig. 3).

Discussion

Blepharochalasis is a rather uncommon and insidiously developing disease usually appearing in young individuals. In most cases there is a bilateral presentation of the deformity, a restriction to one side is extremely rare and only a few cases have been reported.[2–5]

The course of the disease is characterized by a recurrent and painless swelling of the eyelids lasting only a couple of days. The results of these repeated attacks include thinning and wrinkling of the skin, a discoloration through increased pigmentation and a relaxation of the subcutaneous tissues. This condition was previously described as a cigarette paper appearance of the skin.[6] The sequelae are ptosis, pseudoepicanthic fold and proptosis, as formerly reported by various other authors.[2–4]

The condition can be divided into two different stages.[6] Initially, there is a hypertrophic or swelling stage in which the skin of the eyelid becomes oedematous. This also may be referred to as the active stage.

Later, it is followed by a second stage of either atrophy or hypertrophy, according to Tenzel and Stewart[7] and Custer et al.[8] In the opinion of Collin[4] it is doubtful that a late hypertrophic form exists.

The ultimately resulting redundant upper eyelid skin may interfere with upward fields of vision or pose a social embarrassment to the patient due to poor cosmetic appearance.

Castañares proposed a classification of eyelid deformities that is useful as a differential diagnosis.[9]

Dermochalasis is also a condition of an excess fold of skin of the upper eyelid. It is seen mainly in older patients and is characterized by hypertrophy of skin. *Hypertrophy of the orbicularis oculi muscle* is an entity different from others, although they may coexist. The pathogenesis involves constant smiling and squinting, as well as blepharospasm. *Herniated orbital fat* is by far the most common cause of eyelid deformity and seen in young as well as in older patients. Lid deformity may be secondarily caused by a *ptosis of the eyebrows* as a result of the general aging process of the face occurring in many individuals.

A last possibility would be a combination of the above-mentioned conditions.

The etiology and pathogenesis of blepharochalasis remains controversial. Basically, there are two possible pathogenetic causes. First, there is a rarefaction of elastic tissue, leading to a progressive laxity of the eyelid.

The second contributor to blepharochalasis could be a localized form of lymphedema. Lymphedema is an abnormal collection of interstitial fluid, caused by stasis of the lymph channels. It is confined to the dermis and subcutaneous tissue. The disease is classified as either primary or secondary. The most common form of primary lymphedema is lymphedema praecox, a condition typically diagnosed at puberty, probably associated with rising hormone levels.[10,11] The primary form of lymphedema can be further categorized into aplastic, hypoplastic or hyperplastic, based on lymphangiographic findings.[12] In our case a possible explanation could be a localized congenital malformation (i.e. hyperplastic form of primary lymphedema) of the lymphatic vessels causing an intermittent lymphedema.

We speculate that only the combination of an insufficient network of elastic fibres and a dysfunctional lymphatic system are able to produce symptoms and, thus, lead to blepharochalasis. The histopathological examination showed the coexistence of both pathogenetic mechanisms. The patient may possibly have a rarefaction of elastic fibres also in the left upper eyelid but due to intact lymphatic vessels there are none of the symptoms as seen on the other side.

Summarizing, blepharochalasis is a rare disease mainly in young adults and appearing typically on both

sides, a unilateral distribution being exceedingly rare. Awareness of this condition can prevent time-consuming and fruitless search for underlying systemic disorders. The excess of skin leading possibly to a visual obstruction in the later stages can be corrected by a simple blepharoplasty done on an outpatient basis.

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The tuberous male breast

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SUMMARY. Whilst tuberous female breasts are well described, the tuberous male breast is a very unusual variant of gynaecomastia. Two cases are presented, the development of the condition is considered and the surgical management is discussed. © 2003 The British Association of Plastic Surgeons. Published by Elsevier Science Ltd. All rights reserved.

Keywords: gynaecomastia, tuberous breast, surgery, tubular breast.

Gynaecomastia is a common condition causing significant embarrassment. Although gynaecomastia can be associated with hormonal imbalances, tumours, hepatic, thyroid and renal disease and drug interactions, the majority of cases arise in adolescent and middle-aged males in the absence of an underlying pathology.[1] Most pubertal males experience some degree of transient gynaecomastia, which usually subsides in less than two years,[2] whilst around 30% of middle-aged males manifest the condition.[3] We present two cases of a rare manifestation of the disease—the tuberous male breast.

The typical presentations of gynaecomastia have been classified by Simon et al.[4] (Table 1).

Lower-grade gynaecomastia can be managed with simple excision and/or liposuction of the abnormal tissue (Grade I–IIa), whilst higher-grade disease will usually require skin excision and, consequently, involves more

Table 1 Simon et al.'s classification of gynaecomastia[4]

Grade	Clinical appearance
I	small but visible breast development with little redundant skin
IIa	moderate breast development with no redundant skin
IIb	moderate breast development with redundant skin
III	marked breast development with much redundant skin