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CASE REPORT

Ascher syndrome

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ABSTRACT

Ascher syndrome is a rare, benign skin disorder characterized by a double upper lip, blepharochalasis, and nontoxic enlargement of the thyroid gland. The exact cause is unknown, but it is considered to be a hereditary disease with an autosomal dominant trait. We report here a case of forme fruste Ascher syndrome in a 29-year-old man.

Introduction

Ascher syndrome is a rare, benign skin disorder first described in 1920. The syndrome is characterized by a triad of a double upper lip, blepharochalasis, and nontoxic enlargement of the thyroid gland. The nontoxic enlargement of the thyroid gland is not believed to be a necessary feature of the syndrome because it does not present in all patients.[1] The syndrome without nontoxic enlargement of the thyroid gland is considered as the forme fruste, or incomplete form.[2] We present here a report of a patient with incomplete Ascher syndrome coexisting with xanthoma of the eyelid.

Case Report

A 29-year-old man was admitted to our hospital as a result of ptosis of the upper eyelids and recurrent episodes of swelling of his upper lip for about 10 years. Recurrent swelling of the upper eyelid occurred without any causative factor and the swelling was abrupt in onset and gradually subsided after 2–3 days. Antihistamine treatment had no therapeutic effect. About 6 months later, his upper eyelids were seen to be gradually thinning with ptosis. There was no history of topical steroid use. Simultaneously, yellowish symmetrical progressively growing papules appeared on the medial surface of both upper eyelids and there was increasing swelling of his lips. There was no history of trauma or surgical procedure to his lips. There was no symptom suggestive of hypo- or hyperthyroidism, nor was there any relevant personal or familial medical history.

Physical examination showed blepharochalasis with obvious telangiectasia on his upper eyelids and bean-sized soft yellow plaques were bilaterally detected on the medial surface of both upper eyelids. The upper lip was obviously swollen and enlarged with a shallow transverse sulcus between the mucosa and the skin of the lip (Figure 1). When the patient smiled, a deformity with a fold of excess tissue on the mucosal aspect of the upper lip was remarkably prominent. An ophthalmological examination did not find any abnormality. There was no clinical evidence of thyroid enlargement. A complete blood count, routine urinalysis, stool microscopy, liver and renal function tests, serum levels of complement, and the erythrocyte sedimentation rate were normal. Blood lipids were within the normal limits. His thyroid gland was normal in size and consistency under ultrasonic examination and thyroid function tests were normal.

Biopsy samples were taken from the outside of his right upper eyelid. Microscopic examination showed a normal epidermis, significantly atrophic subcutaneous eyelid tissue with proliferative telangiectasia, and varying amounts of inflammatory cells infiltrating the surrounding vessels and adnexa in the dermis (Figure 2). Verhoff staining showed that the dermal elastic fibers were unremarkable on the normal side of the lesion, whereas the dermal elastic fibers were significantly reduced or missing on the thinning side of the lesion (Figure 3A and B). A diagnosis of forme fruste Ascher syndrome was diagnosed with the coexistence of eyelid xanthelasma.

Conflicts of interest: The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in this article.

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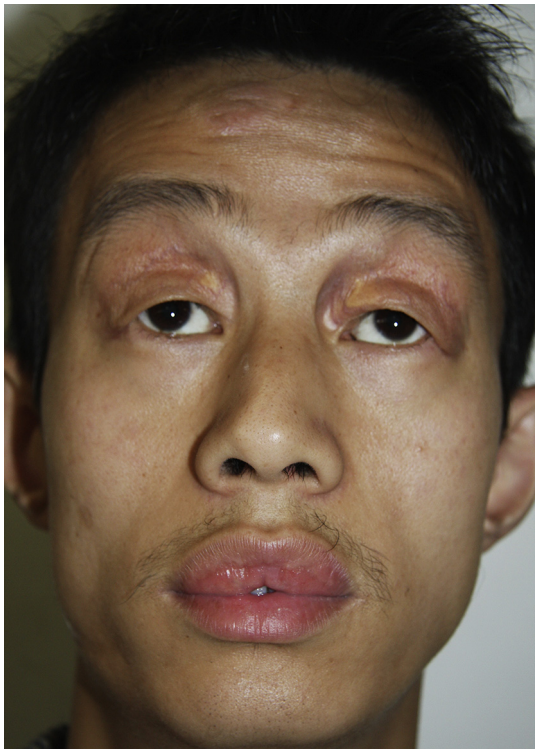

Figure 1 Patient with atrophic and swollen upper eyelids with telangiectasia, yellow plaques on the medial surface of both upper eyelids and remarkable deformity of his upper lip.

Discussion

Ascher syndrome is a combination of blepharochalasis with progressive enlargement of the upper lip due to hypertrophy and inflammation of the labial salivary glands; it generally occurs within the first 20 years of life. Although some factors, including trauma and hormonal dysfunction, have been suggested, the exact cause is unknown and it may be inherited as an autosomal dominant trait.[3] Clinically it is characterized by a triad of double upper lip, blepharochalasis, and nontoxic enlargement of the thyroid gland. Goiter usually presents several years after the initial eyelid and lip edema, but is manifested in only 10–50% of patients and is not essential for the diagnosis of Ascher syndrome.[4] A double upper lip may be either congenital or acquired. Duplication between the inner and outer parts of the upper lip is usually caused by recurrent swelling. The lower lip may occasionally be affected.[1]

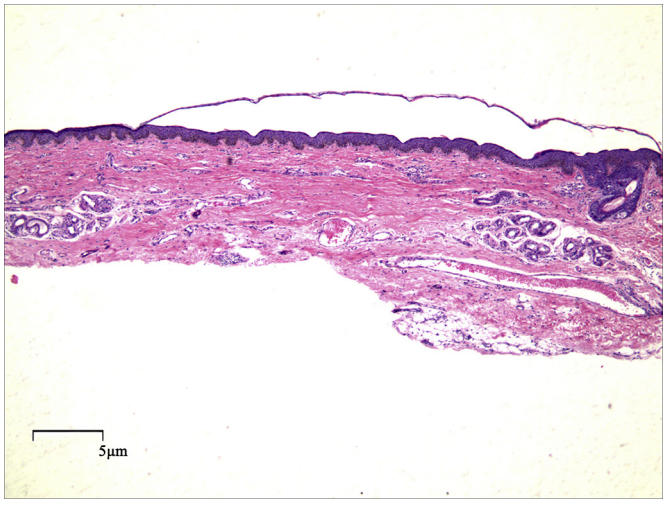

Figure 2 Histological examination shows a normal epidermis, significantly atrophic subcutaneous tissue with proliferative telangiectasia, and mild inflammatory infiltration in the dermis. Hematoxylin and eosin stain, original magnification ×40.

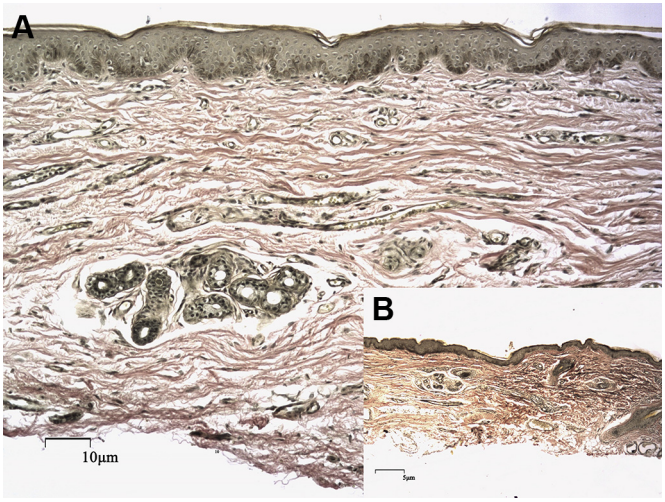

Figure 3 (A) Verhoff staining shows that elastic fibers are significantly reduced in the dermis. Verhoff staining, original magnification ×100. (B) Elastic fibers are normal in the dermis of the normal side (right) and significantly reduced on the thinning side (left). Verhoff staining, original magnification ×40.

Acquired-type Ascher syndrome usually results from trauma or oral habit, whereas the congenital type is a developmental anomaly.[4,5] In 80% of patients the swelling eyelids appear prior to the age of 20 years and lower eyelid edema is noted in severe cases of the disease.[6] Usually the patients present with transient swelling of the bilateral upper eyelids, which is followed by blepharochalasis with or without telangiectasia, sometimes with a reduction in the visual field as a result of narrowing of the palpebral fissure. Therefore angioneurotic edema-like episodes can be misdiagnosed as angioedema, early dermatochalasia, acquired cutis laxa, and variants of granulomatous cheilitis, especially in the early stages.[7] In addition, the swollen lips need to be differentiated from some vascular tumors, lymphangioma, mucoceles, salivary gland tumors, and inflammatory fibrous hyperplasia.[5]

Our patient presented with a forme fruste of Ascher syndrome coexisting with eyelid xanthoma. The pathogenesis of Ascher syndrome and eyelid xanthoma is different. Whether such a coexistence is a coincidence, or whether it indicates a possible correlation in the underlying pathogenesis, needs further investigation.

There is no specific pharmaceutical treatment for this syndrome. Cosmetic surgery is generally the treatment of choice when the condition interferes with vision, speech, and chewing. Surgical treatment to excise the excess tissue of eyelids or lips is usually recommended. Good functional and cosmetic results are usually obtained, although recurrence is occasionally reported.[8] Our patient did not request further treatment due to the expense and risk of surgery.

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