# Do you know this syndrome?*
## *Você conhece esta síndrome?**

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**CASE REPORT**

Eighteen-year-old, single, white female patient, student, born in and coming from Santo Amaro (SC), came to the Dermatology Outpatient Clinic with the complaint of recurrent bilateral eyelid edema, otherwise asymptomatic, for approximately five years. She had already sought medical assistance, but had had no treatment due to lack of diagnosis. Claimed no previous diseases or similar cases in the family.

Upon dermatological examination, presented lax skin in upper lids with a redundant local fold, reaching the level of eyelashes (Figures 1 and 2), with no other abnormalities.

A face computerized tomography scan had been requested in 2002, revealing a discrete thickening of the skin and subcutaneous tissue in the periorbitary region, with no other alterations. Laboratorial tests (blood count, blood glucose, renal and hepatic function, thyrostimulating hormone, free T4, anti-thyroid antibodies) in November/2004 were all normal, and thyroid ultrasonography indicated goiter with no nodules. Skin histopathological examination evidenced an epidermis with no particularities and a dermis with a slight mononuclear cell infiltration. Staining for elastic fibers showed, in the papillary dermis, fragmentation of elastic fibers (Figure 3).

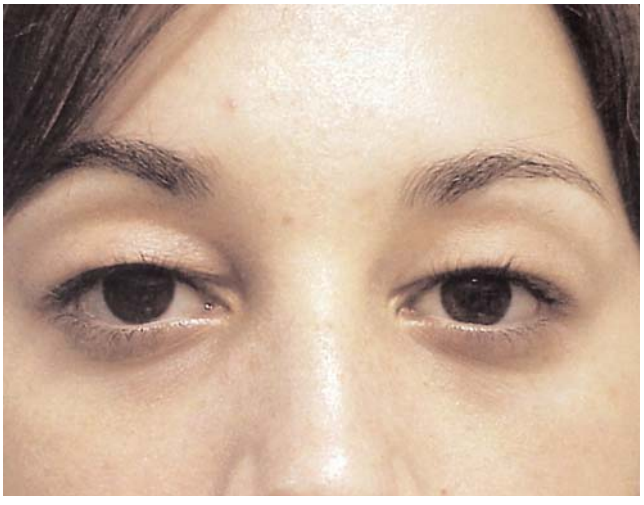

**Figure 1:** Skin laxity of upper eyelids (blepharochalasia)

**Figure 2:** Redundant local fold, in upper eyelids

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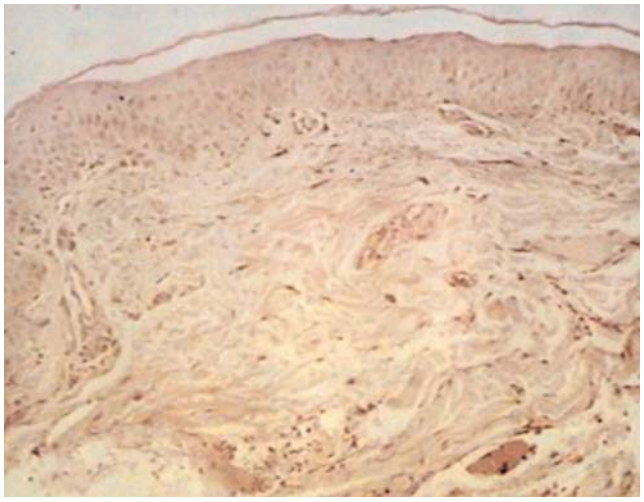

**FIGURE 3:** Histological examination showing elastic fiber fragmentation (10 X)

**WHAT IS THIS SYNDROME?**

**Ascher's syndrome**

With data obtained from history and physical examination, the main diagnostic hypothesis elaborated was that of Ascher's syndrome (AS), followed by bleopharochalasia. Such diagnosis was then confirmed by histopathological examination and laboratorial tests, leading to the onset of treatment (in nov/2004) with dapsone 50 mg/day, in an attempt to decrease the evolution of the process, until arrangement of a possible blepharoplasty. Since patient evolved with hemolytic anemia, medication was discontinued, and she is currently waiting for surgery

AS was described for the first time in 1920 and, since then, a little over 50 cases have been presented in literature. It may be that such slenderness of cases is due to little knowledge and small quantity of existing studies, not to mention subdiagnosis.[1]

Classified as a subtype of anetodermia, it is a rare syndrome, characterized by the variably expressive association of eyelid skin laxity (blepharochalasia) and upper lip edema (with a double lip appearance).[2] Thyroid increase, usually asymptomatic, is present in only 10% of cases, even though it has been originally described as a part of this syndrome.[3,4]

Etiology is unknown, but there is evidence for a defect in elastic fibers, which are either fragmented or decreased in number. Likely described etiological hypotheses are dominant autosomic inheritance, hormonal and allergic dysfunction, also being considered a traumatic origin.[5]

A certain degree of blepharochalasia, related to atrophic skin and eyelashes relaxation, is common in aging, but uncommon in young persons, being reported as recurrent episodes of bilateral eyelid edema, elastic tissue disease, lax cutis, or of idiopathic origin.[1] The greatest differentiation between AS and blepharocalasia is in histopathology, where a small presence or total absence of elastic fibers in the compromised tissue is seen associated to the atrophic skin in the second condition.

In up to 80% of the cases, symptoms appear before age of 20, developing as an almost simultaneous eyelid and upper lip edema, with abrupt onset. Lower eyelid edema is only noted in severe cases.[4]

There is no efficable treatment, the only one being repair of damages caused by skin laxity by means of plastic surgery. ❑

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**Abstract:** Ascher's Syndrome, a subtype of anetodermia, is a rare disease characterized by the association, in different degrees, of upper eyelid laxity, lip swelling and increase in thyroid size. Its etiology is unknown, but there is evidence for a defect in elastic fibers, which are either fragmented or decreased in number. A case is reported of a 16-year-old patient with a single complaint: recurrent episodes of bilateral eyelid edema.
Keywords: Blepharoplasty; Dapsone; Eyelids

***Resumo:*** *A síndrome de Ascher (SA), um subtipo de0 anetodermia, é rara e caracterizada por apresentar em associação, com maior ou menor expressividade, relaxamento da pele das pálpebras, edema de lábio superior e aumento da tireóide. A etiologia é desconhecida, mas há evidências de defeito nas fibras elásticas, que se apresentam fragmentadas ou diminuídas. Relata-se o caso de paciente de 16 anos de idade, com queixa única: episódios recorrentes de edema bipalpebral.*
*Palavras-chave: Blefaroplastia; Dapsona; Pálpebras*

**REFERENCES**

1. Gomes-Duaso AJ, Seoane J, Vozquez-Garcia J, Arjona C. Ascher syndrome: report of two cases. In J Oral Maxillofac Surg. 1997;55:88-90.
2. Kara IG, Kara CO. Ascher syndrome. Otolaryngol Head Neck Surg. 2001;124:236-7.
3. Krug-Jeromonim J, Pisera P. Ascher's syndrome: report of 2 cases. Pol Merkuriusz Lek. 1999;7:275-6.
4. Sanchez MR, Lee M, May JA, Ostreicher R. Ascher syndrome: a mimicker of acquired angiedema. J Am Acad Dermatol. 1993;29:650-1.
5. Alkan A, Metin M. Maxillary double lip: report of two cases. J Oral Sci. 2001;43:69-72.

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