

Atrophoderma Vermiculata – Case Report

Fekete Gyl¹, Fekete Júlia Edit²

¹ Department of Dermatology, University of Medicine and Pharmacy, Tirgu Mures, Romania

² Public Health Center, Tirgu Mures, Romania

Atrophoderma vermiculata is a rare genodermatosis probably inherited in an autosomal recessive pattern, characterized by follicular inflammation and atrophy. Together with keratosis pilaris atrophicans facies and keratosis follicularis spinulosa decalvans it belongs to a group of closely related conditions, characterized by keratosis pilaris and atrophy of the skin. Clinical manifestations with reticular atrophy of the skin in a honeycomb appearance are characteristic. The course is generally slow, with progressive worsening. We report a clinical case of a 59 year-old woman who presented a symmetric reticular atrophy of the face, which appeared when she was 18 years old, with progressive worsening in the last 3 years. The lesions were located on the forehead and cheeks, without any clinical symptoms. The disease is considered rare, the true incidence is unknown.

Keywords: atrophoderma vermiculata, genodermatosis, atrophy of the skin

Received: 12 April 2011

Introduction

Atrophoderma vermiculata is a rare genodermatosis, probably inherited in an autosomal recessive pattern, characterized by follicular inflammation and atrophy. Together with keratosis pilaris atrophicans facies and keratosis follicularis spinulosa decalvans, it belongs to a group of closely related conditions characterized by keratosis pilaris and atrophy of the skin. The basic morphological manifestation of this disease is the honeycomb-like reticular atrophy, situated symmetrically especially on the cheeks, forehead and in the preauricular area. Skin lesions are caused by abnormal keratinisation of the pilosebaceous follicle.

Case presentation

We report a clinical case of a 59 year-old woman who was consulted for a symmetric reticular atrophy of the face, appeared when she was 18 years old. The lesions slowly developed until the age of 38, when they became stationary. The disease had a progressive worsening in the last 3 years, when new atrophic depressions appeared. The multiple depressions of the skin were irregular in shape, with a diameter between 1–5 mm and a depth of about 1 mm, sharply defined and delimited by irregular bridges of epidermis, resulting in a honeycomb appearance of the skin. The lesions were located on the forehead and cheeks, without any clinical symptoms.

The personal and family history of the patient had no relevance for the disease. Other skin diseases were not present (Figure 1 and 2). The performed rutin laboratory tests were in normal limits. The histopathological findings showed mild perifollicular and perivascular inflammation and fibrosis, as well as a decrease in number or atrophy of the hair follicles. We proposed a dermato-cosmetic treatment, but it was refused by the patient.

Discussions

Atrophoderma vermiculata is a rare disease, probably inherited in an autosomal recessive pattern and also known under various other names, such as atrophodermia reticulata, folliculitis ulerythematososa reticulata or honeycomb atrophy. It usually makes its debut in childhood, at the age of 7 to 12, but may also appear in teenagers or adults, irrespective of sex or race [1].

Clinically the disease is characterized by a symmetrical reticular atrophy, located in most cases on the cheeks, forehead and preauricular area, rarely with extension on the sides of the arms. Eyebrows, eyelashes and hairy skin of the scalp are not affected. Unilateral locations have also been described [2]. The lesions consist of irregularly shaped depressions of 2–3 mm in diameter and 1 mm depth, sharply defined and delimited by narrow bridges of the epidermis resulting in a honeycomb or worm-eaten aspect [3]. The disease is associated with facial erythema, keratosis pilaris and rarely with comedone and milium cysts [4,5]. The histologic aspect is not specific: follicular knots, atrophy of hair follicle, dermal sclerosis and perifollicular and perivascular inflammation can be found [6,7]. The disease generally takes a slow course, with progressive worsening until puberty and subsequent recovery, even by spontaneous regression.

Differential diagnosis can be made with reference to conditions of atrophic scars of the face such as lupus erythematosus, varicella and acne. Also degenerative, idiopathic and congenital atrophic skin diseases must be differentiated [8]. Atrophoderma vermiculata is very rarely associated with oral leukokeratosis [9], Marfan syndrome [10], neurofibromatosis, atrial septal defect, Rombo syndrome and Nicolau-Balus syndrome [11]. The disease has no effective treatment. Procedures such as cryotherapy, phototherapy, keratolytic treatment have proven ineffective. The dermato-cosmetic treatments like dermabrasion and/or collagen injections, dioxide laser or YAG laser treatment could be the best therapeutical alternative.



Fig. 1. Clinical aspect – frontal



Fig. 2. Clinical aspect – lateral

Conclusions

Atrophoderma vermiculata is a rare disease, probably inherited in an autosomal recessive pattern. The disease has a distinctive clinical manifestations and an unspecific histological appearance. The disease is considered rare, the true incidence is unknown. We consider this case interesting to publish due to the fact that it is the second case reported in Romania in the last two decades.

References

1. Oanta A, Irimie M. *Atrophoderma vermiculata* – a case report. *Rev dermatovenerologie*. 2010;4:347-351
2. Arrieta E, Milgram-Sternberg Y. Honeycomb atrophy on the right cheek. *Arch Dermatol*. 1988;124:1101-4.
3. Lorusso B, Benci M, Lotti T. Monolateral *atrophoderma vermiculatum*. / *Eur Acad Dermatol Venereol*. 1993;2:239-41.
4. Dupre A, Carrere S, Bonafe JL et al. Syringomes éruptifs généralisés, grains de milium et atrophodermie vermiculée. Syndrome de Nicolau et Balus. *Dermatologica*. 1981;162:281-6.
5. Kooij R, Venter J. *Atrophoderma vermiculata* with unusual localisation and associated congenital anomalies. *Dermatologica*. 1959;118:161-7.
6. Bassioukas K, Fragidou M, Nakuci M et al. *Atrophoderma vermiculata*. *Cutis*. 1997;59:337-40.
7. Frosch PJ, Brumage MR, Schuster-Pavlovic C et al. *Atrophoderma vermiculatum*. Case reports and review. *J Am Acad Dermatol*. 1988;18: 538-42.
8. Bucur Gh: *Boli dermatovenerice*. Enciclopedie. Editura Medicală Națională, 2002; 90-91.
9. Seville RH, Mumford PF. Congenital ectodermal defect: *atrophoderma vermicularis* with leukokeratosis oris. *Br J Dermatol*. 1956;68:310.
10. Sidwell RU, Harper JI. Vermiculate *atrophoderma* in a boy with Marfan syndrome. *Br J Dermatol*. 1999;141:750-1.
11. Ashinoff R, Jacobson M, Belsito DV. Rombo syndrome: a second case report and review. *Am Acad Dermatol*. 1993;28:1011-4.