

# Anetoderma with positive Darier's sign

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## ABSTRACT

Anetoderma describes a localized laxity of the skin due to dermal elastolysis. It can be either primary or secondary to an inflammatory dermatosis. Rarely secondary anetoderma has been associated with mastocytosis. We report a 16-year-old man, with multiple pouch like lesions (anetoderma) mostly on the face, neck and upper trunk which had positive Darier's sign. Anetodermic lesions developed two years ago without preceding urticaria pigmentosa lesions. In skin biopsy perivascular mast cell and eosinophil infiltrations were observed.

**Key words:** Anetoderma; Mastocytosis; Darier's sign

## INTRODUCTION

Anetodermic lesions are localized areas of flaccid or herniated saclike skin with focal loss of dermal elastic tissue. This rare condition has been classified as primary or idiopathic anetoderma which flaccid lesions occur in normal skin and secondary anetoderma which is preceded by inflammatory, autoimmune, infectious or neoplastic processes. The most common sites of anetoderma are the trunk, thighs and upper arms [1]. Cutaneous mastocytosis is characterized by accumulation of mast cells in the skin with heterogeneous clinical presentations [2], which include maculopapular lesions (urticarial pigmentosa), erythrodermic and telangiectatic aspect. Rare and atypical presentations of mastocytosis has been reported in literatures consist of bullous, pseudoxanthomatous, congenital dermographism and anetodermic lesions [3,4]. Degranulating mast cells could be able to degraded dermal elastin [5] and induce anetoderma. Mild trauma results in erythema and urticaria around mastocytosis macule which named Darier's sign which helps us as a clinical guide for detecting anetodermic mastocytosis [2].

## CASE REPORT

We present herein a 16-year-old man, with multiple asymptomatic flaccid pouch like lesions mostly on the face, neck and upper trunk. On skin examination the lesions could be depressed deeply without resistance (Figs. 1a and b). Stroking of the lesions resulted in erythema and swelling (positive Darier's sign) but he had no dermographism on the normal skin. These lesions developed two years before the first visit without any inflammatory skin lesions such as acne vulgaris or insect bite. Past medical, drug and family history were negative. He had no atopy, allergic rhinitis or asthma. His parents did not remember any skin lesions during the infancy to suggest urticaria pigmentosa (UP). He mentioned that his lesions began as a soft skin color nodules which were gradually changed to flaccid pouch like lesions or spontaneously. Two skin biopsies were done. One from a soft nodule on the back and the other one from a pouch like lesion on the chin. Histopathologic sections of both specimens showed skin tissue covered by an atrophic epidermis under lied by edematous dermis with prominent separation of collagen bundles and perivascular infiltrations of lymphocytes, mast cells and eosinophils. Significant loss of elastic

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fibers in orcein staining was reported (Figs. 2a and b). No mucin identified in alcian blue stained sections. Immunohistochemistry (IHC) study revealed increased perivascular CD117 positive cells (Fig. 2c), which all of these findings were in favor of anetodermic mastocytosis.

Our patient had no systemic symptoms such as flushing, abdominal pain, diarrhea or bone pain. On clinical examination there was no organomegaly or lymphadenopathy. Laboratory evaluations including complete blood count, erythrocyte sedimentation rate, C- reactive protein, liver function test, urine analyses, were normal. Urinary 24-hour N-methyl histamine levels were slightly elevated. Due to lack of the systemic symptoms, serum tryptase level and bone marrow aspiration were not done. He complained of development of new lesions almost every six months, so phototherapy was considered for him.

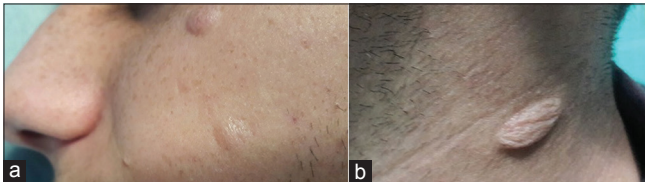
Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

## DISCUSSION

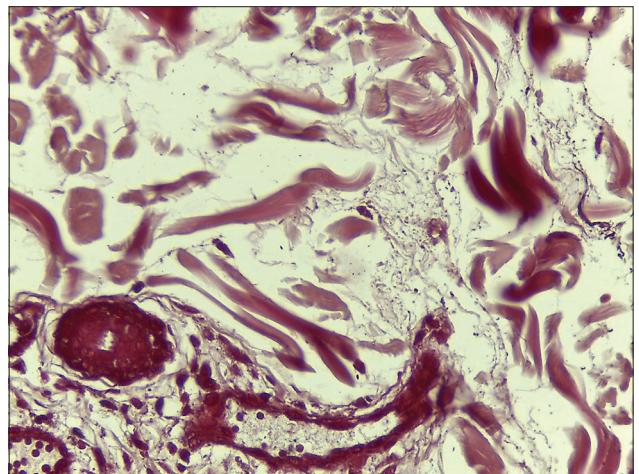
Anetoderma is a benign condition with focal loss of dermal elastic tissue resulting in localized areas of flaccid or herniated saclike skin, whose etiopathogenesis

has been unknown. This rare disorder occurs mainly in women aged 20–40 years. In the most usual form, crops of round or oval, pink macules 0.5–1 centimeter in diameter develop on trunk, thighs and upper arms, less commonly on the neck and face and rarely elsewhere. The lesions remain unchanged throughout life, and new lesions often continue to develop for many years [6]. Anetoderma has been classified as primary and secondary. In primary anetoderma, patients do not remember prior inflammation in the lesions and no abnormal inflammatory infiltrate in dermis or microthrombosis are seen in skin biopsy. Secondary anetoderma develops after many different dermatosis including acne, varicella, syphilis, leprosy, sarcoidosis, mastocytosis, granuloma annulare, xanthoma, pilomatrichoma, lymphoma and lupus profundus [7].

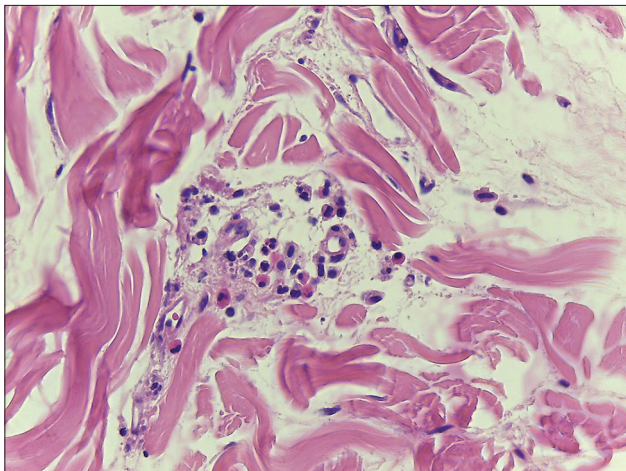
Our case was interesting to us because secondary anetoderma associated with mastocytosis has been rarely reported in the literature. It has been observed in only a



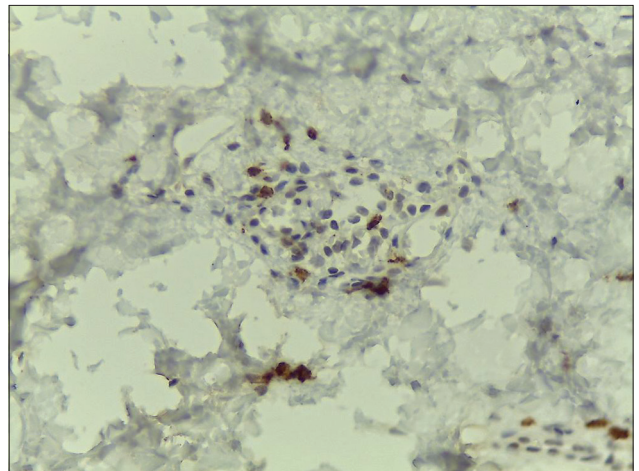
**Figure 1:** (a and b) Anetodermic lesions on the face and neck.



**Figure 2b:** In orcein staining mid dermal elastolysis was seen (x 40).



**Figure 2a:** Perivascular mast cell and eosinophilic infiltrations in dermis were seen (H & E, x 40).



**Figure 2c:** In Immunohistochemistry perivascular CD117 positive mast cells were seen (x 40).

few patients with UP which is the most common type of mastocytosis in children [3]. Kalogeromitros, et al reported anetodermic lesions developed on the sites of UP lesions [8] but our patient's family did not report any preceding UP lesions during the childhood and anetodermic lesions superimposed over the soft asymptomatic new skin color nodules or sometimes developed spontaneously. Positive Darier's sign on anetodermic lesions was the only point for diagnosis which is pathognomonic for mastocytosis. So Darier's sign should be investigated in anetodermic lesions with unknown origin in order to exclude cutaneous mastocytosis [2]. The exact mechanism of mechanical degranulation of tissue mast cells after rubbing the lesions is still obscure. Darier's sign might reflect abnormal mechanosensitivity of mast cells in certain skin lesions [9].

Pathogenesis of anetoderma is unclear but degeneration of the elastic fibers could be mediated by close contact of degranulating mast cells with elastic fibers [5]. The role of elastase and other serine proteinases such as tryptase and chymase in mast cells has been postulated in anetoderma [10]. On the other hand elastin fragments are chemoattractant for phagocytes such as macrophage [5] which aggravate dermal elastolysis and result in anetoderma.

Our patients had no pruritus or UP lesions but he complained of new onset lesions during last two years. Del Pozo, et al reported phototherapy with ultra violet A and psoralen (PUVA) was effective in control of anetodermic mastocytosis [2]. So we scheduled phototherapy for him to prevent new lesions and he was followed for detecting any systemic symptoms of the mastocytosis in futures.

## Abbreviations

(UP): Urticaria pigmentosa  
(IHC): Immunohistochemistry  
(PUVA): Psoralen- Ultraviolet A

## Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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