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Linear atrophoderma of Moulin on the neck

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Abstract

Background: Linear atrophoderma is a rare disease, first described by Moulin in 1992 in 5 patients. It is an acquired unilateral hyperpigmented, depressed band-like areas following the lines of Blaschko. It affects children or adolescents of both genders involving the trunk or the limbs. It is considered to be a rare cutaneous form of mosaicism.

Main observation: A 39-year-old woman with a 22 years history of unilateral slightly depressed hyperpigmented lesion on her neck was admitted to us. The skin texture was normal and there were no signs of induration or sclerosis. The histopathological examination revealed a normal epidermis outlined by a hyperpigmented basal layer. In the papillary dermis proliferation of superficial vessels with mild lymphocytic infiltrate and melanin-laden macrophages were present. The collagen fibres and elastic fibres were normal. The clinical and histopathological features confirmed the diagnosis of linear atrophoderma of moulin. We discussed the case according to the other cases reported in the literature.

Conclusion: Approximately 28 cases of linear atrophoderma have been reported in literature. The present case has the charecteristic clinical and histopathological features of linear atrophoderma as defined by Moulin, but the localization of the lesion is unusual. (*J Dermatol Case Rep*. 2011; 5(3): 47-49)

Introduction

Linear atrophoderma of Moulin (LAM) is a rare entity first described by Moulin in 1992.[1] It is charecterized by asymptomatic hyperpigmented atrophic band-like lesions localized mostly on the trunk and following the lines of Blaschko. Baumann *et al*. suggested the term LAM for these band-like scleroderma-like skin lesions that do not show a preceding inflammation or induration or scleroderma.[2] They categorized this disease as belonging to the group of acquired linear dermatoses following Blaschko's lines (BL). They considered LAM as a variant of progressive idiopathic atrophoderma of Pasini and Pierini. In general, the condition begins in childhood or adolescense, and there is no evidence of any long-term progression.[3]

Case report

A 39-year-old female patient was admitted to the dermatology department with the complaint of eruption on left half of her neck. There were no clinical symptoms such as pain, pruritus or previous inflammation. The lesion occured 22 years ago. The patient reported that the lesion had developed as two small sized hypopigmented macule initially and then brown band-like hyperpigmentation had occured in a short period of time and remained unchanged. Her past medical history was unremarkable.

On physical examination, there was a slightly depressed band-like hyperpigmented lesion on the left half of the neck starting under the mandible to the midline of nuchae following Blaschko's lines (BL) (Fig. 1A,B). The superficial veins on the lesion

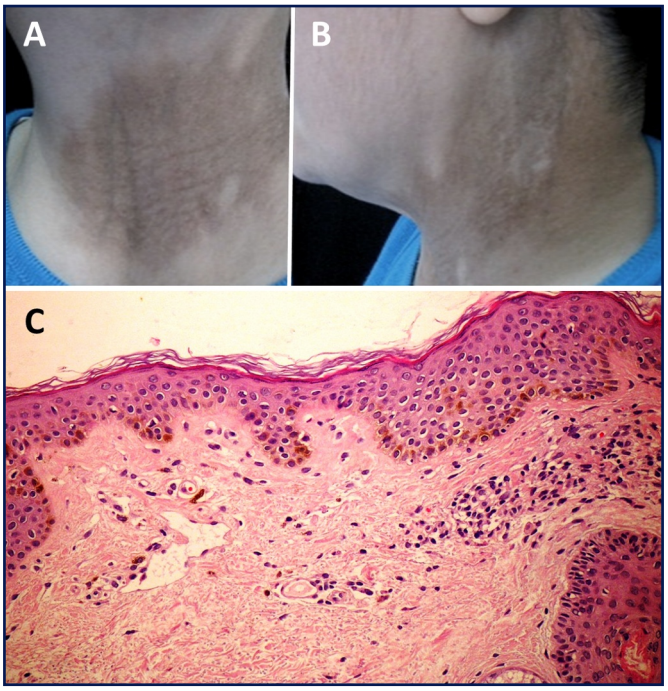

Figure 1

Slightly depressed band-like hyperpigmented lesion with visible superficial veins on the neck (A, B). Histopathology shows a normal epidermis outlined by a hyperpigmented basal layer. In the papillary dermis proliferation of superficial vessels with mild lymphocytic infiltrate and melanin-laden macrophages examined (H+E, 40x).

were clearly seen and two small hypopigmented plaque were also observed. It had a normal texture and there were no signs of induration or sclerosis.

Laboratory investigations, including complete blood cell count, liver or kidney function tests, erythrocyte sedimantation rate, antinuclear antibody tests, double stranded antinuclear DNA antibodies, anti-ssDNA antibodies, anti-SCL-70, anti-histon antibodies, anti-RNP antibodies, anti-Ro (SS-A), anti La (SS-B) antibodies were negative or within the normal range. Chest X-ray and ultrasound examination of the abdomen gave normal results.

A skin biopsy was taken from the lesion. Histopathological examination showed a normal epidermis outlined by a hyperpigmented basal layer. In the papillary dermis proliferation of superficial vessels with mild lymphocytic infiltrate and melanin-laden macrophages were present (Fig. 1C). The collagen fibres and elastic fibres were normal with Masson Trichrome and Elastic Van Gieson stains.

The diagnosis of LAM was made in consideration of the clinical examination and the histopathological results.

Discussion

There have been 28 cases of LAM described in the English literature since the first description of the disease by Moulin in 1992, to our knowledge. It is now believed that these criteria should be provided for the diagnosis of the disease as follows: onset during childhood or adolescence; development of hyperpigmented, slightly atrophic, unilateral lesions following Blaschko lines on the trunk or limbs; absence of prior inflammation or subsequent scleroderma; a stable, nonprogressive clinical course without a pattern of remission; and histologic findings showing hyperpigmentation of the basal epidermis and a normal dermis with unaltered connective tissue and elastic fibres.[4]

Some authors disagree with the diagnosis of this new entity and refuse some reported cases for histopathological differences. The main differences between the histopathology of cases reported to date are perivascular lymphocytic inflammatory infiltrate in the superficial dermis combined with abnormal collagen fibres.[4] Ang *et al*. proposed naming this condition Blaschko-linear atrophoderma of Pasini and Pierini because perivascular lymphocytic infiltrate and abnormal collagen fibres are more characteristics of atrophoderma of Pasini and Pierini than of LAM.[5] Histologic changes in the epidermis as atrophy, acanthosis, hypogranulosis, parakeratosis and hyperkeratosis have also been reported in some cases.[4] These cases probably seem to be describing epidermal nevi, linear inflamatory epidermal nevi, lichen striatus, or nevoid hypermelanosis.[6]

The lesion of our case developed when she was 17 years old. She did not describe any prior inflammation. The lesion was stable and did not progress in the past 22 years and no remission was seen. Histologic findings were: hyperpigmented basal layer, mild lymphocytic infiltrate in upper dermis with melanin laden macrophages and normal collagen and elastin fibres. All these findings enabled us to make the diagnosis of LAM, but the localization of the lesion was unusual. It was on the left half of the neck, but we found that it was following the Blaschko's lines of the neck area. Cecchi *et al* reported a 9-year-old Peruvian boy with exclusive involvement of the neck.[7] So this is the second report of LAM with this unusual localization.

The differential diagnosis of LAM include the skin disorders like linear nevoid lesions, genetic and acquired dermatoses, e.g., epidermal nevi, hypomelanosis of ito, lichen striatus, lichen nitidus, postinflammatory hyperpigmentation. In our case linear scleroderma and atrophoderma of Pasini and Pierini (APP) can be considered in differential diagnosis because the other dermatoses have different clinical features. Linear scleroderma has similar configuration, atrophy and hyperpigmentation but the absence of inflammation, induration or sclerosis in the present patient supported the diagnosis of LAM. APP may also resemble LAM but it does not follow Blaschko's lines and perivascular lymphocytic infiltration and abnormal collagen fibres are more charecteristic features in its histopathological examination. LAM was suggested a variant of APP, localized to BL by some authors.[8] However, the correlations between these two entities do not seem to have been clarified yet.[9]

The aetiopathogenesis of LAM has not been elucidated so far. It is thought to be caused by a somatic mutation that takes place early in embryogenesis, resulting in a geno and phenotypic mosaicism such as other dermatoses following BL.[10] But, no gene has been identified that is responsible for LAM to date. Our patient did not accept the genetic testing.

There is not a proven effective treatment of LAM. High-dose penicillin, topical corticosteroids, heparin, oral potassium aminobenzoate have been used but found to be ineffective.[11] As the disease occurs in a short period of time and does not progress, it seems to be hard to find an effective treatment model.

Conclusion

The present case has the charecteristic clinical and histopathological features of linear atrophoderma which was defined by Moulin but the localization of the lesion is different from the reported cases in the literature. We believe that genetic counselling of these cases would highlight the etiopathogenesis of this entity.

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