

EDUCACIÓN CONTINUA

Haga usted el diagnóstico

A 6-year old child with hyperpigmented patches in the right arm and chest

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A 6 year-old male patient was admitted to the dermatology department with an eruption on the right arm, chest and left leg. No clinical symptoms such as pain, pruritus or previous inflammation were present. His mother reported that the lesion had developed four years ago as small-sized hyperpigmented macules which enlarged in a short period of time as band-like hyperpigmentation, and then remained unchanged. His past medical history was unremarkable and there were no similar cases in the family.

On physical examination, band-like hyperpigmented patches on the right arm were observed. A similar lesion was located on the right side. When the right shoulder was on horizontal adduction, lesions of

chest and arm concurred in a large brown ring-like patch. Lesions on left leg showed the similar band-like distribution with a darker brown color (figure 1). There were no signs of induration or sclerosis.

Laboratory investigations included complete blood count, liver and kidney function tests, erythrocyte sedimentation rate, and antinuclear antibody test, which were negative or within the normal range.

A skin biopsy was performed. Histological examination showed a normal epidermis outlined by a hyperpigmented basal layer. In the papillary dermis, vessels were surrounded by mild lymphocytic infiltrate and melanin-laden macrophages were present (figure 2).



Figure 1. A. Ring-like pigmented patch involving the right arm and chest. B. Blaschko linear dark brown papulo-patches on the left leg.

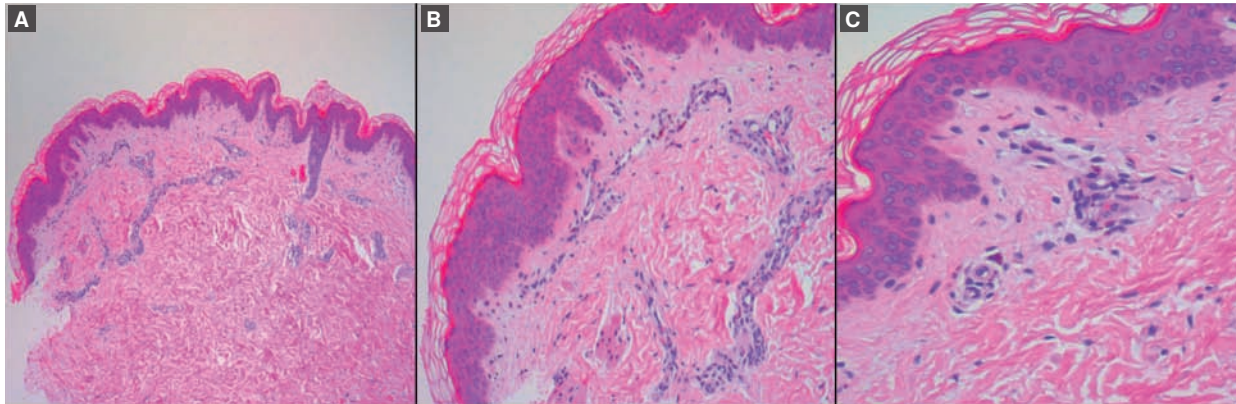


Figure 2. **A.** Panoramic view showing normal epidermis and collagen fibers. An infiltrate is observed in papillary dermis. Hematoxylin and eosin, 5X. **B.** Hyperpigmented basal layer of the epidermis with melanin laden macrophages. The infiltrate is mostly formed by perivascular lymphocytes. Hematoxylin and eosin, 10X. **C.** Closer view of the hyperpigmented basal layer, melanin laden macrophages and lymphocytic infiltrate. Hematoxylin and eosin, 20X.

What is the most likely diagnosis?

- a) Mongolian spot
- b) Linear atrophoderma of Moulin
- c) McCune-Albright syndrome
- d) Becker nevus
- e) Lichen striatus

Answers

Mongolian spot: Incorrect. It is a congenital condition featured by blue-color spots or patches usually on lumbosacral region or buttocks, which results from the entrapment of melanocytes in the lower half to two-third of dermis during their migration from the neural crest to the epidermis. This asymptomatic spot usually fades away by school age (1).

McCune-Albright syndrome: Incorrect. Caused by a mutation in the *GNAS1* gene, the hallmark symptoms of this syndrome are irregular, large patchy *café-au-lait* spots predominating on one side of the back without crossing the midline associated with early puberty and bone deformities. Blood test shows increased adrenal and growth hormones (2).

Becker nevus: Incorrect. Becker nevus is an uncommon pigmented disorder characterized by hyperpigmentation and sometimes hypertrichosis.

It is more prevalent in men (2-5:1) with a prevalence close to 0.52% (3). This condition usually develops in the peripubertal period, and it increases slowly, acquiring a geometric and asymmetric configuration. The most commonly involved areas are shoulders, anterior chest and the scapular area. This nevoid melanosis is referred to as Becker nevus syndrome when it is associated with other anomalies such as muscular hypoplasia, skeletal disorders and localized lipodystrophy (4).

Lichen striatus: Incorrect. It is a rare, benign, self-limited linear dermatosis of unknown origin that predominantly affects children aged 5-15 years (5). It appears as a sudden eruption of small papules on an extremity, which may resolve with post-inflammatory hyper or hypopigmentation. Histopathological features include hyperkeratosis, acanthosis, focal spongiosis, lymphocytic exocytosis, basal layer vacuolation and lichenoid lymphocytic infiltrate (6).

Linear atrophoderma of Moulin: Correct. Linear atrophoderma of Moulin is a rare entity described by Moulin in 1992 (7). Since then, to our best knowledge, 32 cases have been reported in English literature. It is featured by asymptomatic hyperpigmented band-like lesions localized mostly on the trunk following the Blaschko's lines. Some authors consider linear atrophoderma of Moulin as a Blaschko-linear variant of progressive idiopathic atrophoderma of Pasini and Pierini (8). Linear atrophoderma of Moulin begins in childhood or adolescence developing into pigmented unilateral lesions in a short period of time in absence of prior inflammation or subsequent scleroderma.

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The clinical course is stable, non-progressive and shows no pattern of remission. The lesions in the case of our patient developed when he was two years old, they progressed in a short period of time and remained stable for the following four years. One interesting feature of our case is the ring-like pattern of the lesion.

The clinical features of linear atrophoderma of Moulin are very characteristic to suspect the diagnosis, which is confirmed by histological examination. In our skin sample, findings were: hyperpigmented basal layer and mild lymphocytic infiltrate in upper dermis with melanin laden macrophages and normal collagen and elastin fibers. These outcomes enabled us to make the diagnosis of linear atrophoderma of Moulin. The differential diagnosis includes different genetic and acquired dermatoses such as linear nevoid lesions, epidermal nevi, hypomelanosis of Ito, lichen striatus, lichen nitidus and postinflammatory hyperpigmentation. The etiology of this condition remains unclear. It is thought to be caused by a somatic mutation resulting in a geno- and phenotypic mosaicism such as other dermatosis following Blaschko's lines (9). Several therapeutic approaches such as penicilins, topical steroids, heparin and oral potassium aminobenzoate have been used for linear atrophoderma of Moulin with no effective results (10).

In our case, no therapy was initiated and the lesions have remained stable during the follow-up.

Conflicts of interest

Authors declared no conflicts of interest.

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References

1. **Reza AM, Farahnaz GZ, Hamideh S, Alinaghi SA, Saeed Z, Mostafa H.** Incidence of Mongolian spots and its common sites at two university hospitals in Tehran, Iran. *Pediatr Dermatol.* 2010;27:397-8. <http://dx.doi.org/10.1111/j.1525-1470.2010.01168.x>
2. **Kumar KH, Dhull P, Bisht Y.** Hansen's disease with McCune-Albright syndrome. *J Glob Infect Dis.* 2012;4:215-7. <http://dx.doi.org/10.4103/0974-777X.103901>
3. **Danarti R, Konig A, Salhi A, Bittar M, Happle R.** Becker's nevus syndrome revisited. *J Am Acad Dermatol.* 2004;51:965-9. <http://dx.doi.org/10.1016/j.jaad.2004.06.036>
4. **Cosendey FE, Martínez NS, Bernhard GA, Dias MF, Azulay DR.** Becker nevus syndrome. *An Bras Dermatol.* 2010;85:379-84. <http://dx.doi.org/10.1590/S0365-05962010000300015>
5. **Taniguchi-Abagge K, Parolin-Marinoni L, Giraldo S, Carvalho VO, de Oliveira Santini C, Favre H.** Lichen striatus: Description of 89 cases in children. *Pediatr Dermatol.* 2004;21:440-3. <http://dx.doi.org/10.1111/j.0736-8046.2004.21403.x>
6. **Sato H, Shirai A, Asahina A.** Case of lichen striatus presenting with multiple lesions along Blaschko's lines. *J Dermatol.* 2012;39:802-4. <http://dx.doi.org/10.1111/j.1346-8138.2011.01427.x>
7. **Moulin G, Hill MP, Guiland V, Barrut D, Chevallier J, Thomas L.** Bandes pigmentées atrophiques acquises suivant les lignes de Blaschko. *Ann Dermatol Venerol.* 1992;119:729-36.
8. **Bauman L, Happle R, Plewig G, Schirren CG.** Atrophoderma linearis Moulin. A new disease picture, following the Blaschko lines. *Hautarzt.* 1994;45:231-6.
9. **Utikal J, Keil D, Klemke CD, Bayerl C, Goerdts S.** Predominant telangiectatic erythema in linear atrophoderma of Moulin: Novel variant or separate entity? *Dermatology.* 2003;207:310-5. <http://dx.doi.org/10.1159/000073096>
10. **Atasoy M, Aliagaoglu C, Sahin O, Ikbali M, Gursan N.** Linear atrophoderma of Moulin together with leuconychia: A case report. *J Eur Acad Dermatol Venerol.* 2006;20:337-40. <http://dx.doi.org/10.1111/j.1468-3083.2006.01434.x>