

Photodermdiagnosis

What caused this patch?

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The healthy mother of a 6-month-old girl was concerned about an irregular, pale, relatively large patch on the abdomen of her child. (**Figure 1**) She reported that the lesion was present at birth, had increased in size with the child's growth, and may have become slightly lighter and more noticeable over the 3 months prior to her presentation. The fathers of the child, as well as 3 siblings, were healthy with no abnormal mucocutaneous findings. There was no history of autoimmune disorders in the family. What would be the main differential diagnosis in this case? How would you approach it?



Turn the page for the diagnosis and a brief discussion.

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Diagnosis

Nevus anemicus

Discussion

Nevus anemicus (NA), also known as pharmacologic nevus, is a nonmelanotic disorder in which pigmentation remains unaffected. NA usually presents as a unilateral pale area of variable size (3-10 cm) with an irregular or “broken up” outline on the trunk. It is usually present at birth but often noticed later in life. The trunk is the most common location. It occurs more frequently in females and is usually asymptomatic. Histologically, there are no abnormalities of the melanocytes or melanin content.

NA is caused by decreased blood flow through the capillaries in the dermal papillae, due to localized hypersensitivity of the blood vessels to catecholamines. Local blood vessels are very sensitive to endogenous catecholamines, remaining permanently vasoconstricted.¹ So, the paleness in NA is vascular, not pigmentary.

In addition to nevus depigmentosus (ND), the differential diagnosis includes other hypopigmented or depigmented disorders, such as hypochromic nevi, ash leaf spots of tuberous sclerosis complex, and vitiligo. An extensive review of the differential diagnosis is beyond the scope of this paper. In the following discussion we will focus on the main differences between NA and ND (**Table 1**).

Practical approach in the office

Diascopy On diascopy, the borders of the lesions will blanch with pressure due to constriction of the blood vessels in the surrounding normal skin; this will make the lesion indistinguishable from surrounding skin.

Friction With a tongue blade, you may make a few strokes across both the lesion and the normal-appearing surrounding skin. You should notice a flare on the skin surrounding the lesion, but not the central pale area of the NA.

Heat or ice cube Application of heat or an ice cube will often accentuate the lesion, as the border becomes hyperemic while the lesion stays pale. NA is also more noticeable when there is surrounding vasodilatation due to emotional stress.

Wood's lamp Wood's lamp examination does not accentuate NA, and may make the lesion inapparent, unlike true depigmenting disorders.

Getting to the roots of the term “pharmacologic nevus”

The following findings may justify the fact that nevus anemicus is best termed a pharmacologic nevus resulting from increased vascular sensitivity to catecholamines, and suggesting that several pharmacological anomalies may be involved:

- a. Intralesional injection of bradykinin, acetylcholine, pilocarpine, serotonin, nicotine,

Table 1 Comparison and contrast of nevus anemicus and nevus depigmentosus

<i>Nevus anemicus</i>	<i>Nevus depigmentosus</i>
1. Congenital patch manifested by skin pallor	1. Congenital dermatosis that manifests as unilateral, hypopigmented patches
2. Most commonly occurs as a single patch on the trunk	2. Usually occurs in a dermatomal pattern.
3. Caused by a localized vascular hypersensitivity to catecholamines which produces increased vasoconstriction and skin pallor	3. Caused by a block of transfer of melanosomes from melanocytes to keratinocytes
4. No loss of melanin occurs in the lesion	4. Loss of melanin is present causes the lesion
5. Is a pharmacologic nevus	5. Is an anatomic nevus
6. Wood’s lamp examination does not accentuate it, rather makes the lesion inapparent	6. Wood’s lamp examination makes the lesion apparent
7. Diascopy obliterates the border between lesion and normal skin, because the normal skin becomes blanched.	7. Diascopy would not obliterate the border between lesion and normal skin.
8. Scratching a line across both the lesion and normal surrounding skin will produce erythema in the normal skin but not within the lesion, similarly application of cold or heat will not produce changes within the lesion	8. Scratching a line across both the lesion and normal surrounding skin will produce erythema both in the normal skin and within the lesion
9. Histology normal, and melanocytes are preserved and normally distributed.	9. Histology is abnormal showing loss of melanosomes in the melanocytes.

- b. 5-hydroxytryptamine, and histamine fails to induce the anticipated vasodilatation or erythema in the affected area. However, following axillary sympathetic block or intradermal injection of pilocarpine (an alpha-blocking agent) vasodilatation and erythema are produced in the lesion.
- c. It also has been proposed that an abnormality in endothelial adhesion molecule induction (E selectin expression) may be involved.

The donor site dominance

In autograft exchange transplantation studies, the skin transplanted within the NA retains characteristics of the donor

site. This phenomenon is known as the donor site dominance.

Nevus depigmentosus

Nevus depigmentosus (ND) is also called achromic nevus or nevus achromicus. ND is a cutaneous abnormality consisting of a hypopigmented macular lesion which can present as circumscribed irregular, oval, or round or as a unilateral band or streak arranged along one or more Blaschko lines that is present at birth and remains stable over time.²⁻⁴ Most lesions measure a few centimeters in diameter and have irregular but well-defined borders. When it is very extensive, it is indistinguishable from hypomelanosis of Ito. The name is a bit of misnomer as the areas of leukoderma are actually hypomelanotic not amelanotic. Wood’s light makes the lesion more noticeable.

On histopathology, melanocytes can be normal or slightly reduced in number in ND. Transfer of the melanosomes from melanocytes to keratinocytes is believed to be abnormal in ND. A few patients with ND have been reported to have associated seizures, mental retardation, pes cavus ipsilateral to the hypopigmentation, and hemihypertrophy.^{5,6} However, in a recent review of 50 cases of ND from India,⁷ no abnormal neurologic features were found.

In a more recent article Diepell *et al.*⁸ reported the case of a patient with ND associated with nevi flammei affecting two contralateral quadrants, venous insufficiency of the right leg, and asymmetry of the arms.

We hope this short review helps our readers differentiate NA from ND.

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