

### Answer to Dermacase *continued from page 1293*

#### 3. Nevus anemicus

Nevus anemicus, first described by Hans Vörner in 1906, is an uncommon, congenital, nonprogressive, localized skin anomaly.<sup>1</sup> This asymptomatic condition is characterized by a circumscribed pale-coloured macule or patch that can be of various size and shape (eg, rounded, oval, linear, or irregularly shaped) and that is sometimes surrounded by satellite macules. The lesions can appear on any part of the body, but are mostly found on or near the trunk, particularly the chest and back. The condition usually presents at birth or in early childhood, and occurs more commonly in women. Most of the time, nevus anemicus is an isolated finding, without associated abnormalities; however, a small number of cases have been linked to genodermatoses such as neurofibromatosis and phacomatosis pigmentovascularis.<sup>2</sup>

It was previously believed that there were autonomic alterations in nevus anemicus lesions, presumably increased stimulation of vasoconstrictor fibres or inhibition of vasodilator fibres of the blood vessels.<sup>3</sup> However, results of a sympathetic block study in nevus anemicus have since suggested that the primary defect could be due in part to increased local sensitivity to catecholamines.<sup>4</sup> This theory is further supported by the donor dominance demonstrated after transplantation of a nevus anemicus lesion to normal skin (ie, pale appearance is retained).<sup>5</sup>

Histopathologic findings for the nevus anemicus lesion are essentially the same as those for normal skin. An accurate diagnosis can usually be made based on the typical clinical presentation in conjunction with relevant physical examination. Diascopy is a simple and quick examination in which physicians apply pressure to the lesion and adjacent unaffected skin with a glass slide. Nevus anemicus becomes indistinguishable from the surrounding skin with diascopy. Another quick diagnostic examination is scratching or rubbing a line across both the lesion and the normal surrounding skin. Reactive erythema will appear in the normal skin but not within the nevus anemicus lesion. Ultraviolet light, such as that produced by a Wood lamp, does not accentuate nevus anemicus.

#### Differential diagnosis

Vitiligo is the most important differential diagnosis—nevus anemicus is often misdiagnosed as vitiligo, leading to inappropriate overtreatment. Vitiligo is estimated to affect 1% of the population in the United States, typically children or young adults, with an equal incidence in both sexes. The pathogenesis is not yet fully established, but many theories have been proposed, including autoimmune, neural, cytotoxic, biochemical, and viral mechanisms.<sup>6</sup> The condition is characterized by 1 to several

well-demarcated amelanotic macules or patches predominantly distributed on the periorificial areas, mucous membranes, face, extensor surfaces, hands, feet, and genitals. In contrast to nevus anemicus lesions, vitiligo lesions are well accentuated upon Wood light examination. Limited skin lesions can be treated with topical steroids; immunomodulators, such as tacrolimus and pimecrolimus; or targeted phototherapy with a 308-nm excimer laser.<sup>7</sup> Extended skin lesions require systemic phototherapy or even depigmentation of residual normal skin when there is extensive involvement.

With respect to clinical features, nevus depigmentosus is very similar to nevus anemicus. Nevus depigmentosus manifests as a congenital, nonprogressive, hypopigmented macule or patch that remains stable throughout the patient's life.<sup>8</sup> The skin lesion is thought to result from a developmental defect of the fetal melanocytes, particularly a defect in the transfer of melanosomes from melanocytes to keratinocytes.<sup>9</sup> Nevus depigmentosus can be differentiated from nevus anemicus during physical examination: erythema occurs after stroking the skin in nevus depigmentosus but not in nevus anemicus. In addition, nevus depigmentosus lesions appear off-white upon Wood lamp examination. No treatment is needed. Ablative laser with thin skin grafting could be considered in patients with considerable cosmetic concerns.

Tinea versicolor is a common superficial fungal infection characterized by scaly hypopigmented or hyperpigmented macules or patches that most commonly appear on the chest, back, and proximal extremities. The hypopigmented patches, although accompanied by minimal scaling, can simulate nevus anemicus, and need to be confirmed by a potassium hydroxide wet-mount preparation showing the presence of yeast (classic cases are described as having a "spaghetti and meatballs" appearance).<sup>10</sup> Several topical antifungal agents are effective treatment measures; selenium sulfide or ketoconazole shampoo might work as well.

Tuberous sclerosis (TS) is a rare, genetic, multisystem disease that can involve the eyes, teeth, heart, central nervous system, and, most commonly, the skin. Although two-thirds of cases are sporadic, the remaining one-third follow an autosomal dominant inheritance. Tuberous sclerosis is caused by mutation in either of 2 tumour suppressor genes, TSC1 (maps to chromosome band 9q34, encoding hamartin) or TSC2 (maps to chromosome band 16p13.3, encoding tuberin).<sup>11</sup> The common skin manifestations of TS include facial angiofibromas, periungual fibromas, shagreen patches (connective tissue nevi), gingival fibromas, confetti skin lesions, and hypomelanotic macules.<sup>12</sup> Hypomelanotic macules need to be differentiated from nevus anemicus lesions. They often first appear on the trunk or limbs at birth or within the first few years of life, but unlike nevus

anemicus lesions they might gradually fade or even disappear in adulthood. In addition, they are typically smaller in size, ranging from 0.5 to 3.0 cm, and often appear as so-called ash-leaf spots owing to their resemblance to European mountain ash leaves. Ultraviolet light accentuates hypomelanotic macules, especially in fair-skinned individuals. When in doubt, diascopic examination can further help to differentiate between the conditions. There is no definite treatment and the goals of management are aimed at reducing morbidity and preventing complications through a multidisciplinary team approach.

## Management

Treatment is generally not required for nevus anemicus. Patients might benefit from the application of camouflaging makeup for cosmetic purposes. Clinicians should be aware of associated abnormalities such as neurofibromatosis or vascular anomalies, as nevus anemicus might sometimes accompany these rare genodermatoses.



**Dr Chen** is a senior resident and **Dr Chiang** is an attending dermatologist in the Department of Dermatology at the Tri-Service General Hospital in Taipei, Taiwan.

### Competing interests

None declared

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