

Dermal melanocytoses and variants

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Abstract

Dermal melanocytoses form a spectrum of benign intradermal melanocytic lesions composed of dendritic melanocytes, the most common of which is the blue naevus. A variety of clinical and histological appearances may be assumed, some of which can be worrisome both to the clinician and pathologist, with the most difficult differential diagnosis, in some instances, lying with malignant melanoma. This review presents and discusses the spectrum of histological appearances of this diverse group of lesions, and highlights the clinicopathological variants which are most likely to cause diagnostic difficulty. The lesions that form this spectrum and therefore will be discussed include the dermal melanocytoses such as the Mongolian spot, blue naevus and its variants, deep penetrating naevus, melanoma arising within a blue naevus and, finally, although not a member of this group, cutaneous metastatic melanoma mimicking blue naevus.

Keywords cellular blue naevus; common blue naevus; deep penetrating naevus; dermal melanocytoses; melanoma arising in a blue naevus

Introduction

Dermal melanocytoses form a broad spectrum of lesions characterized by the presence of banal pigmented dendritic and spindled melanocytes interspersed between dermal collagen fibres, and with a propensity to aggregate around adnexal structures and neurovascular bundles. Unlike other melanocytic lesions involving the dermis, they typically express HMB-45 and melan-A. The cells are thought to represent arrested neural crest melanocytes migrating to the skin during embryogenesis.¹ However, this hypothesis remains unproven.

The most commonly encountered of these lesions is the blue naevus which is divided into the common blue naevus (of Jadassohn-Tieche type) and the cellular blue naevus. However, there are many clinical and histological variants which arise from these two forms and much overlap blurs their boundaries. The histological features are not always immediately recognizable as those of a blue naevus and, without prior knowledge of these variants and careful examination, this can lead to diagnostic difficulties.

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This review describes the basic histological features of the dermal melanocytoses, paying particular attention to the common and cellular blue naevus and the many clinical and histological variants that exist, including the malignant counterpart, the malignant blue naevus. Deep penetrating naevus is also regarded, by some, to be part of the same spectrum and for this reason will be included in the discussion.

Metastatic melanoma mimicking blue naevus has been described in the literature and encountered in our own practice. An awareness of this potential pitfall is important and, therefore, this will form the last part of our discussion.

Naevus of Ito, Ota, Sun and Hori and the Mongolian spot

This group of dermal melanocytoses are seen far less frequently than their cousin, the blue naevus. Each one is composed of pigmented dendritic melanocytes distributed between normal collagen fibres and they only differ from one another in their clinical distribution.

Clinical features

All races are susceptible but they are found most commonly in the Asian population and there is also a female predominance. Most are congenital or develop in early childhood and are thought to represent developmental hamartomas or genodermatosis, although acquired cases do exist, in particular Sun's naevus and Hori's naevus. The *Mongolian spot* is familial and always presents at birth. It occurs on the lower back as a large macular area with blue-grey discolouration which disappears clinically with age, although it is still evident with Woods light and histologically.¹ *Naevus of Ito* and *naevus of Ota* present at birth in 50% of cases but they may also occur during childhood, adolescence and more rarely adulthood. There is also no resolution with time. Like the Mongolian spot they present as large blue-grey macules. However, naevus of Ito has a distribution along the lateral brachial and posterior supraclavicular nerves, thus presenting over the shoulder region. Naevus of Ota has a distribution along the ophthalmic and maxillary branches of the trigeminal nerve; thus presenting around the eye, sometimes involving the sclera, the iris, conjunctiva and the mucous membranes of the nose and mouth; they have also been associated with meningeal melanocytomas.² *Sun's naevus* is an acquired lesion affecting individuals between the ages of 8 and 40 years. They occur in the distribution of the zygomatic branch of the trigeminal nerve and are bilateral and symmetrical with a speckled appearance.² *Hori's naevus* is another acquired melanocytosis which presents as pigmented macules with a symmetrical distribution most frequently found on the face.³

Histology

All lesions are composed of variable numbers of individual pigmented dendritic cells aligned parallel to the epidermis and running between normal collagen fibres (Figure 1). Occasional clustering of melanocytes around adnexae and neurovascular bundles may be seen but there is no junctional component and no melanophages are present. The proliferation of melanocytes may be so scarce at times that they become practically invisible histologically. The only minor difference between these entities is the level of involvement; the Mongolian spot tends to involve the deep dermis whereas naevus of Ota, naevus of Ito and Sun's

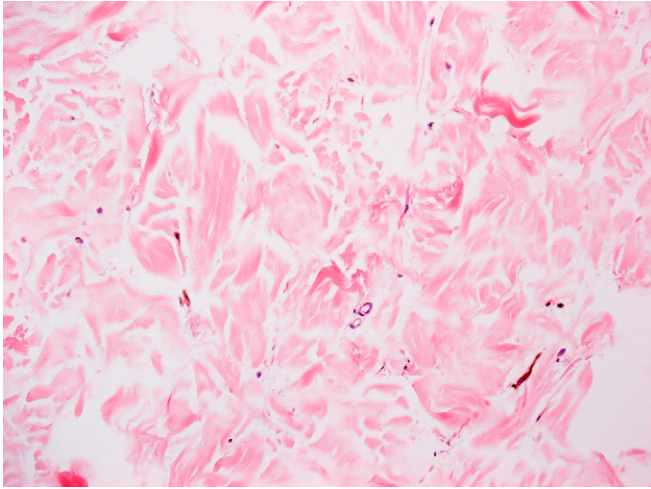


Figure 1 Mongolian spot, x40. Individual pigmented dendritic melanocytes are seen scattered between normal collagen fibres.

and Hori's naevus involve the papillary and superficial component of the reticular dermis.

Malignant transformation of these lesions is vanishingly rare with very few cases of cutaneous melanoma having been described in naevus of Ota and Ito.^{4,5}

Common blue naevus

The common blue naevus is the most frequently encountered of the dermal dendritic melanocytic proliferations.

Clinical features

Occurring slightly more frequently in females they are usually acquired, most often presenting in the second to fourth decades of life. However, some congenital cases have been described⁶ and occasional familial cases⁷ have also been reported.

The most common site involved is the distal upper limb, especially the dorsum of the hand, but they may also be seen on the head and neck, and cases have also been reported involving the oral cavity,⁸ cervix, prostate and lymph nodes.⁹ Lesions typically present as a single blue–black papule measuring no more than 10 mm in diameter.

Histology

The typical histological features consist of a symmetrical lesion situated within the superficial and mid dermis and composed of banal spindle shaped and dendritic cells within a slightly sclerotic stroma. These cells contain abundant melanin pigment (Figure 2a). Their nuclei are small and round to oval in shape with an inconspicuous nucleolus and mitoses are rare. There may be increased cellularity in the centre of some lesions but individual dendritic melanocytes are always found between the collagen fibres at the periphery (Figure 2b). Unlike the common acquired naevus, there is a lack of maturation gradient so little variation in size and shape of the cells is present with descent of the lesion into the deeper dermis. There is also a propensity for melanocytes to aggregate around adnexal structures and vessels, and perineural extension is also frequently seen.^{1,2,10} Melanophages are also evident throughout. A junctional component is not usually a feature, however there

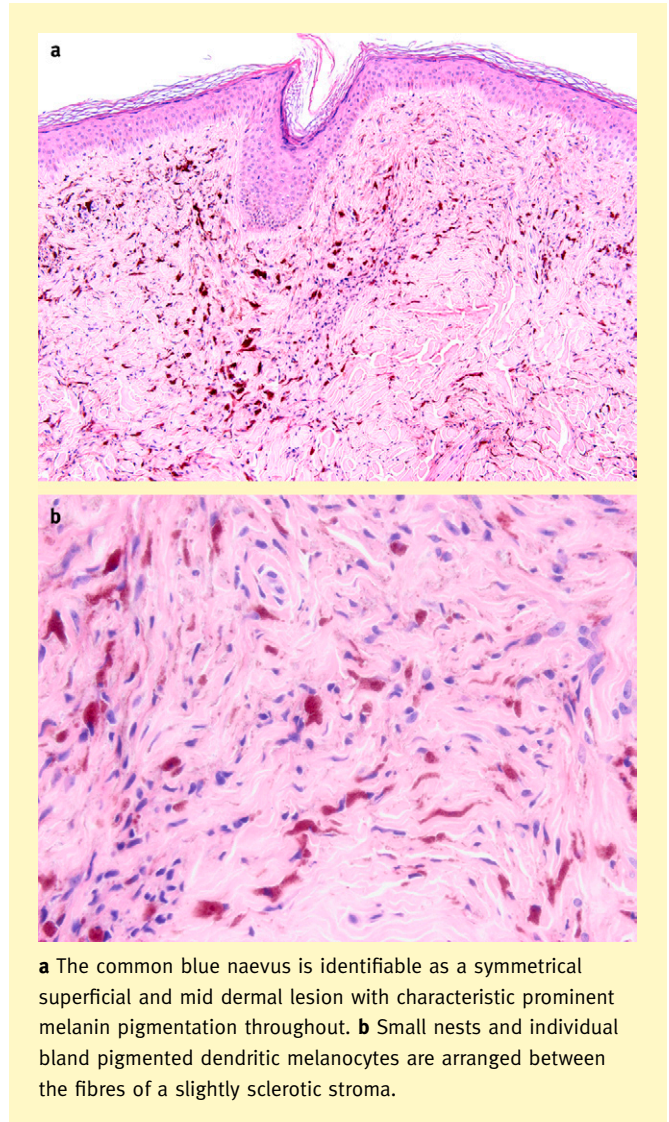


Figure 2

may be hyperpigmentation or even a mild melanocytic hyperplasia involving the basal layer of the epidermis.

Immunohistochemistry shows strong positive staining of melanocytes for S-100, HMB-45 and melan-A throughout the lesion.

Cellular blue naevus

Like the common type of blue naevus the cellular blue naevus tends to occur in the second to fourth decades of life and has a slight female predominance. They are usually acquired although congenital cases have been described.¹¹

Clinical features

The sacro-coccygeal area is the most common site of occurrence followed by the scalp, hands and feet.¹ More unusual sites have also been reported, including the cervix, vagina, spermatic cord and subungual.¹²

They present as a single blue – black papule, nodule or plaque measuring up to 2 cm in diameter; however, they can reach a giant size measuring > 10 cm.^{9,11} Ulceration is not a feature.

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