

Asymptomatic bluish grey nodule on foot

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CLINICAL FINDINGS

A 35 year old male patient presented with asymptomatic bluish skin lesion for 5 years. The lesion was situated on dorsum of right foot. The lesion showed gradual onset and slowly progressive course. It started as a pigmented flat lesion then grew up to form a nodule. The condition was treated initially with topical keratolytic ointment but without significant improvement. There was no history of previous trauma at the same site. The patient had noticed the occurrence of erythematous changes on the lesion at some occasions. On examination, there was a solitary darkly pigmented nodule on the dorsum of the right foot. The lesion measured about 0.8 x 0.6 cm and it was slightly firm with smooth surface. The borders of the lesion were well defined, and it was not attached to underlying structures (Fig. 1). General examination revealed no significant abnormalities.



Fig. 1 Solitary, darkly pigmented nodule on dorsum of right foot.

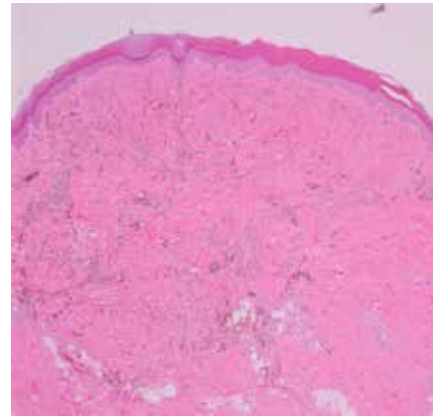


Fig. 2 Uncircumscribed dermal mass formed of pigmented dendritic spindle cells and melanophages situated in between collagen bundles.

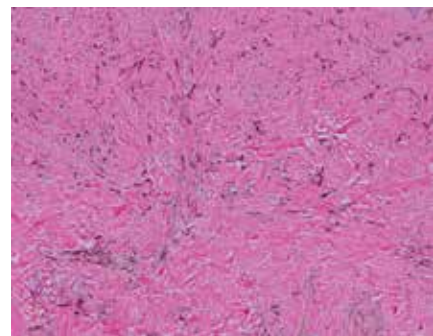


Fig. 3 Spindle shaped cells filled with numerous fine melanin and melanophages intersecting collagen bundles. Some of collagen bundles were thickened and sclerotic.

What is your clinical differential diagnosis?

Pigmented BCC

Dermatofibroma

Pigmented spindle cell nevus

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Pathological findings

Complete excisional biopsy of the lesion was done. Histological examination showed non-circumscribed dermal mass formed of dendritic spindle shaped cells and melanophages in upper and mid dermis. Spindle shaped cells were filled numerous fine granules of melanin and arranged in fascicles and as individual cells in-between collagen bundles. Some of collagen bundles were thickened and sclerotic. The overlying epidermis was hyperkeratotic with mild atrophy.

DIAGNOSIS

Common Blue Nevus

COMMENT

The common or classic blue nevus is a small slate-blue to blue-black macule or papule found most commonly on the extremities. Subungual lesions are rare. It is almost invariably acquired after infancy, but a giant congenital lesion has been reported.¹ The cellular variant is a much larger nodular lesion, often found on the buttocks but sometimes on the scalp or the extremities. The eyelid is a rare site.² Eruptive, plaque, target, amelanotic, linear, satellite, disseminated, and familial forms have been described. Eruptive multiple blue nevi have developed on the penis in a young adult.³

The term 'agminate blue nevus' has sometimes been used for the eruptive and plaque variants. The infiltrating giant cellular blue nevus may involve half the face and extend into striated muscle and the maxillary sinus. The epithelioid blue nevus is a variant which clinically resembles the common blue nevus, except for its distinct histological appearance, its tendency to be multiple, and its association with the Carney complex. A variant of epithelioid blue nevus presenting as a giant con-

genital nevus has been reported. However, epithelioid blue nevus is not always associated with the Carney complex. None of the four cases of epithelioid blue nevus of the genital mucosa was associated with the Carney complex. The various types of blue nevi can often be identified by dermoscopy, based on their unique color variations.⁴⁻⁶

The common blue nevus is composed of elongated, sometimes finely branching, melanocytes in the interstices of the dermal collagen of the mid and upper dermis. There are some melanophages.⁷ Histopathological features of our case matched with common blue nevus. Some lesions show dermal fibrosis (sclerosing blue nevus). A sclerosing 'mucinous' blue nevus with both stromal sclerosis and abundant mucin has been described. In about 3% of cases, there is minimal pigment present. Such cases have been called 'amelanotic' or 'hypopigmented' blue nevi. Occasionally, an overlying intradermal nevus is present: such lesions are called combined or 'true and blue' nevi. Combined nevi are characterized by the presence of two or more different types of melanocytic nevi in a single lesion. 'True and blue' nevi are the most common type of combined nevus. A rare finding is an overlying lentigo or junctional lentiginous nevus, a junctional Spitz nevus, or a dendritic component. This latter lesion which combines a proliferation of junctional dendritic melanocytes arranged individually along the dermoepidermal junction with a common blue nevus in the dermis has been called a 'compound blue nevus'.^{7,8}

A persistent (recurrent) blue nevus has also been described. It may extend significantly beyond the scar of the original excision, which may lead to a clinical misdiagnosis of melanoma. Melanoma *in situ* has also developed over a combined blue nevus.⁹

The cellular blue nevus is composed of dendritic melanocytes, as in the common type, together with islands of epithelioid and plump spindle cells with abundant pale cytoplasm and usually little pigment. Congenital pauci-melanotic cellular blue nevi have been described. Acquired amelanotic cellular blue nevi also occur. Heavily pigmented variants do occur. Melanophages are found between the cellular islands. The tumor often bulges into the subcutaneous fat as a nodular down growth which has a rather characteristic appearance. There are solitary reports of a lesion with subcutaneous cellular nodules, and one of bony infiltration by a scalp lesion. Nerve hypertrophy is often present with perineural aggregation of cells.¹⁰

The giant cellular blue nevus of the scalp can be mistaken for a melanoma. Stromal desmoplasia (desmoplastic cellular blue nevus) and balloon cell change are rare occurrences. A brisk lymphocytic host response is a rare finding.¹¹

The concept of atypical cellular blue nevus was applied for a lesion that had clinicopathological features intermediate between typical cellular blue nevus and the rare malignant blue nevus. No metastases were recorded. The lesions were characterized by architectural and/or cytological atypia including necrosis. No atypical mitoses were present, indicating the importance of this finding in the distinction from malignant blue nevus. A study, involving experienced dermatopathologists, found a lack of consensus for the diagnosis of lesions thought to be cellular blue nevi, atypical cellular blue nevi, or malignant blue nevi. This paper also reviews the criteria proposed by various authors for the diagnosis of atypical cellular blue nevus.¹²⁻¹⁴

The epithelioid blue nevus is composed of in-

tensely pigmented globular and fusiform cells admixed with lightly pigmented polygonal and spindle cells. It shows symmetry on low power. It is a dermal lesion, which like the cellular blue nevus may show extension into the subcutis.¹⁵⁻¹⁸

The melanocytes are usually dispersed as single cells among the collagen bundles, although occasional fascicles exist. This pattern distinguishes this entity from the deep penetrating nevus. Although, some cases of epithelioid blue nevus resemble what has been called the superficial variant of deep penetrating nevus.¹⁹ There is usually no maturation in depth, a feature common to all blue nevi. The epithelioid blue nevus is often part of a combined nevus that may include Spitz nevus, desmoplastic nevus, or congenital nevus. The combination of epithelioid blue and Spitz features in single lesion has been called a blitz nevus.²⁰

Rare variants include the association of a blue nevus with osteoma cutis,²¹ and with a trichoepithelioma,²² and a bizarre blue nevus with striking cytological atypia, but without any other features of malignancy. Perifollicular pigment-laden spindle cells, similar to those seen in a pilar neurocristic hamartoma, are rarely present. Sebocyte like melanocytes were present in one desmoplastic blue nevus. Central myxoid change (myxoid blue nevus) is another rare histological finding. The angiomatoid cellular blue nevus has a conspicuous vascular component resembling hemangioma. Another variant of blue nevus is the 'ancient' blue nevus, a variant of cellular blue nevus with degenerative stromal changes.^{23,24} In addition to pleomorphic and multinucleate melanocytes, there were striking pseudoangiomatous features, hyaline angiopathy, and myxoid change. The melanocytes in blue nevi of all types express S100 protein, melan-A (MART-1), and HMB-45. They

The Clinicopathological challenges of Common Blue nevus.

Diagnosis	Clinical	Pathological
Dermatofibroma	<ul style="list-style-type: none"> • Presents as slowly growing, slightly pigmented, solitary nodule • Frequently develops on the extremities (mostly the lower legs) • Usually asymptomatic but may be pruritic, tender or painful • Can occur in patients of any age but more common in adults 	<ul style="list-style-type: none"> • A poorly defined proliferation of “fibrohistiocytic” cells within the dermis • The overlying epidermis may be acanthotic with increased basal layer pigmentation • The infiltrate is separated from the overlying epidermis by clear ‘Grenz’ zone • At the periphery of the lesion there is entrapment of collagen
Basal Cell Carcinoma	<ul style="list-style-type: none"> • Mainly sun exposed skin, in any hair bearing area (e.g. head and neck) • Also at sites with limited or no sun exposure • Clinical appearance often parallels the histologic subtype • Most common appearance is a papule or nodule with telangiectasias, which may be eroded or ulcerated (ulcus rodens / rodent ulcer) • Papules of BCC may clinically resemble a nevus or fibroma • Pigmented BCC may mimic a melanocytic neoplasm 	<ul style="list-style-type: none"> • Basaloid cells with scant cytoplasm and elongated hyperchromatic nuclei, peripheral palisading, peritumoral clefting and mucinous alteration of surrounding stroma • Also mitotic figures, apoptotic bodies • The presence of myxoid stroma and peripheral clefting has been suggested to be most helpful to distinguish BCC from other basaloid tumors
Pigmented spindle cell nevus	<ul style="list-style-type: none"> • Commonly in the lower extremities • < 1 cm, solitary, deeply pigmented and well circumscribed maculopapule or nodule 	<ul style="list-style-type: none"> • Some similarity with Spitz nevi • Symmetric with cytologic maturation • Nests and fascicles of spindled melanocytes along dermoepidermal junction and within dermal papillae • May be junctional or compound • Expansive, not infiltrative growth pattern • Extends no deeper than reticular dermis • Nevus cells typically contain abundant melanin pigment, may be associated with melanophages • Nuclei are monotonous, resemble normal keratinocytes and may have small nucleoli • Often has architectural or cytologic atypia • Variable lymphocytic infiltrate at base of lesion • Variable transepidermal elimination of junctional nests • No / rare mitotic figures

do not stain for carcino embryonic antigen (CEA). CD34 expression has been reported in a rare congenital form of cellular blue nevus with spindle-shaped cells, suggesting some overlap with neurocristic cutaneous hamartoma.²³ Melanosomes are present in both the dendritic melanocytes and the paler cells of the cellular blue nevus. Some au-

thors have highlighted schwannian features in this variant of blue nevus.⁵

Complete surgical excision is the best method for the treatment of blue nevus. Our case was treated by complete surgical excision and follow up for one year revealed no recurrence.

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