

Agminated Heterogeneous Papules on the Neck

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A 19-year-old man presented to the dermatology clinic for evaluation of several new dark papules on the neck of 1 year's duration. He denied any personal or family history of skin cancer, cardiac abnormalities, or endocrine dysfunction. He also denied any recent changes in health or use of medication. A biopsy was performed at the site 2 years prior for a single blue nevus, but the patient denied history of other trauma or cutaneous eruptions localized to the area. Physical examination revealed numerous dark brown, blue, white, and flesh-colored papules and macules agminated into a well-circumscribed plaque on the left posterolateral neck without background hyperpigmentation. The total area of the plaque was roughly 3×4 cm. There was no associated edema or erythema. Cardiac murmur, thyromegaly, exophthalmos, neurologic deficits, regional lymphadenopathy, and similar skin findings on other areas of the body were not appreciated. Three scouting punch biopsies were taken of the various morphologies present.

WHAT'S THE DIAGNOSIS?

- Becker nevus
- cutaneous amyloidosis
- eruptive blue nevus
- metastatic melanoma
- NAME syndrome

PLEASE TURN TO **PAGE E25** FOR THE DIAGNOSIS

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The authors report no conflict of interest.

The views expressed in this article are those of the authors and do not reflect the official policy or position of the Department of the Navy, Department of Defense, or the US Government.

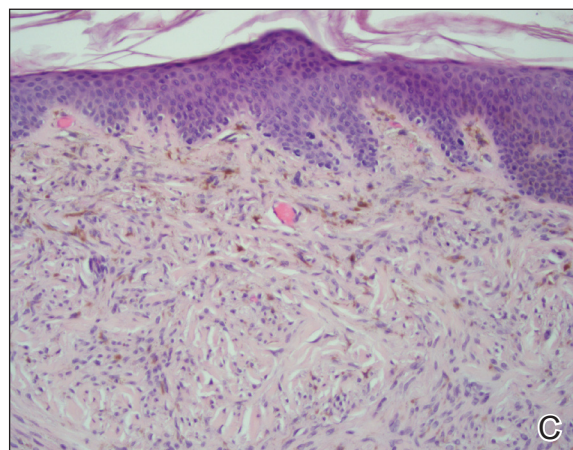
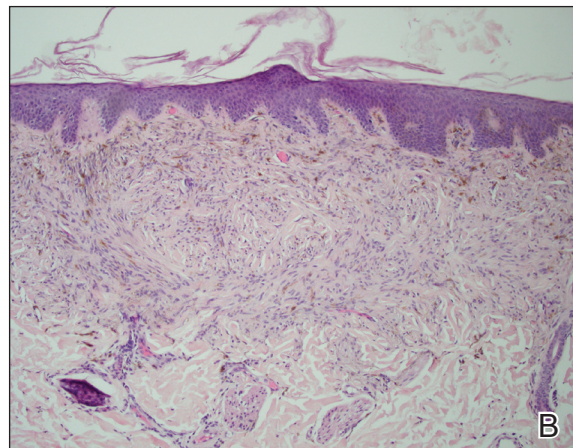
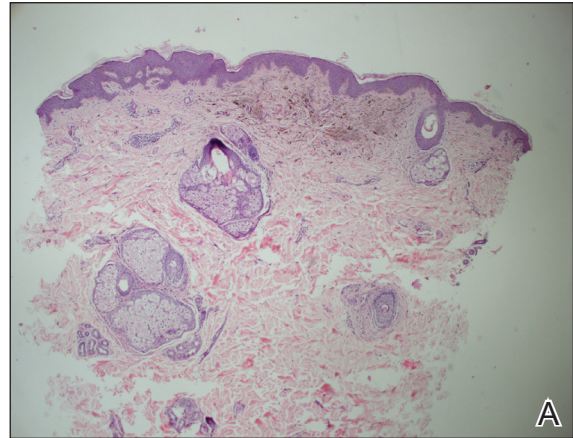
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THE DIAGNOSIS: Eruptive Blue Nevus

All biopsies demonstrated similar histologic features, including an intradermal proliferation of heavily pigmented, spindle-shaped dendritic melanocytes (Figure). The dermal pigment was most pronounced in the grossly darker papules, and there was not a substantial amount of background pigmentation at the stratum basale. Cytologic atypia, foci of necrosis, and mitotic activity were absent from all sections. There was no definitive junctional component identified, no multinucleated giant cells, and there was no overlying epidermal aberration. With some background pigmentation seen histologically, nevus spilus was considered, but because this acute eruption occurred in a young adult without appreciable gross background hyperpigmentation, the clinical context led to a diagnosis of eruptive blue nevus. After communicating the findings to the patient, he declined further treatment.

Eruptive blue nevus is an exceptionally rare subtype of blue nevus with few cases reported since the 1940s.¹⁻⁹ Generally, each case report found a triggering event that could possibly have precipitated the acute proliferation and evolution of nevi. Triggering events can include bullous processes such as erythema multiforme² and Stevens-Johnson syndrome,³ severe sunburn,⁴ trauma,⁵ immunosuppression,⁶ and a variety of endocrinopathies. No such history could be identified in our patient, except the biopsy.

Common blue nevi are benign, usually congenital, well-circumscribed, solitary, blue-gray macules or papules. Half of blue nevi cases are found on the dorsal aspects of the hands and feet but can present anywhere (eg, face, scalp, wrists, sacrum, buttocks). The blue-gray color appreciated clinically is attributed to the Tyndall effect, which occurs when long-wavelength light—red, orange, and yellow—is absorbed by the melanin deep in the dermis, while short-wavelength visible light—blue, violet, and indigo—is reflected with backscattering. On polarized dermoscopy, a homogeneous blue-gray hue is appreciated, but lighter segments may be present when collagen deposition is robust. Histopathologic findings confirm spindle-shaped dendritic melanocytes in the dermis without epidermal involvement. It generally is accepted that the etiology of these benign nevi is a failed migration of neural crest cells to the epidermis.^{10,11} Although the common blue nevus may be simple to diagnose, several subtypes have been described in the literature, including combined blue nevus, desmoplastic blue nevus, hypomelanotic/amelanotic blue nevus, and epithelioid blue nevus of Carney complex, and excluding a malignant process is of monumental importance.^{7,12}



Histopathology of an eruptive blue nevus revealed nests of melanocytes found in the superficial dermis (A), melanocytes in the upper dermis without basal layer pigmentation (B), and pigmented melanocytes in the dermis (C) (H&E; original magnifications $\times 4$, $\times 10$, and $\times 20$).

Biopsy is recommended for common blue nevi in the evaluation of newly acquired lesions, expansion of previously stable nevi, or for nevi larger than 10 mm in diameter. The nature of eruptive blue nevi warrants a biopsy to exclude melanoma or another malignant process. While the Becker nevus may manifest in adolescent males, it is clinically distinct from an eruptive blue nevus due to the size, relative homogeneity, and presence of hair within the lesion. Cutaneous amyloidosis may appear clinically similar to an eruptive blue nevus, but globular or amorphous material was not present in the papillary dermis of biopsied lesions in our patient. Since there was no cellular atypia or mitotic activity, melanoma and other malignancies were ruled out. Lastly, NAME syndrome by definition must include atrial myxomas, myxoid neurofibromas, and ephelides in addition to the nevi; however, our patient had only nevi and few ephelides. Once the diagnosis is established and benign nature confirmed, treatment is not necessarily required. If the patient elects to remove the lesion for aesthetic reasons, an excision into the subcutaneous fat is required to ensure complete removal of deep dermal melanocytes. Prior excisions of eruptive blue nevi have had no recurrence after more than 10 months.^{8,9}

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