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Unusual melanoma of the scalp with blue nevus-like features and local metastasis: A case report

Sheenagh JK George[1], Peter Green[2] and Noreen MG Walsh[2,3]

Abstract

We present an unusual case of melanoma in a 76-year-old female covering approximately 80% of her scalp. Partial sampling of the lesion revealed focal blue nevus-like features at the microscopic level. We discuss issues related to blue nevus-like melanomas and highlight the unique clinical presentation of the current case.

Keywords

Blue nevus, blue nevus-like melanoma, scalp melanoma, local metastasis

Introduction

Blue nevus-like melanoma is one of the rarest subtypes of melanoma, accounting for a mere 0.3% of all melanomas.[1] Controversies exist regarding the nomenclature and diagnostic criteria for such lesions. We report a patient with scalp melanoma displaying partial blue nevus-like morphology and extensive local metastasis.

Case report

A 76-year-old female was seen in clinic for scalp changes. Her attention was drawn to localized asymptomatic vertex scalp changes by her hairdresser. She continued to develop new lesions over the ensuing months and presented to her family physician who referred her to a dermatologist. She was not aware of previous nevi on the scalp and denied a family history of melanoma. She denied associated pain or neurologic symptoms.

Her past medical history included a diagnosis of T3 N0 rectal carcinoma 10 years previous; this was treated with resection, radiation, and chemotherapy. A splenic lesion was found 2 years later and the patient subsequently underwent splenectomy; pathology demonstrated extramedullary hematopoiesis.

Physical examination revealed findings all localized to hair-bearing scalp. Multiple asymptomatic papules and plaques with variable pigmentation, from tan, pink, and blue-black were distributed throughout the hair-bearing scalp. Surface texture ranged from peau d'orange and mammilated to smooth and nodular (see Figures 1 and 2).

At the vertex zone between midscalp and crown, a collection of distinct grey and blue-black papules were noted (see Figure 3).

Two punch biopsies were obtained, one from a deeply pigmented component of the lesion on the vertex of the scalp (specimen A), and the other from a pink/tan plaque on the left parietal temporal region (specimen B). The first sample (specimen A) displayed a sheet-like proliferation of spindled and dendritic, pigmented, melanocytes, admixed with numerous melanophages in the upper third of the dermis. Occasional small irregular junctional nests of melanocytes were evident (see Figures 4 and 5). Mitotic figures in the dermal component of the lesion were sparse but, with the benefit of an immunohistochemical stain for Ki-67, many lesional cell nuclei were seen to be highlighted by this marker of cell proliferation. The histopathological findings in the second sample (specimen B) differed from those in the first biopsy (see Figures 6–8). In this instance, a sheet-like proliferation of plump atypical epithelioid melanocytes without melanin pigment occupied the upper third of the dermis and did not involve the dermo-epidermal junction. Scattered mitotic figures were apparent and a brisk proliferative index was confirmed by a Ki-67 stain. Both biopsies

[1]Dalhousie University Medical School, Halifax, NS, Canada
[2]Department of Medicine, Dalhousie University, Halifax, NS, Canada
[3]Department of Pathology, Dalhousie University, Halifax, NS, Canada

Corresponding Author:
Sheenagh George, Dalhousie University Medical School, Suite 4-193 Dickson Building, 5820 University Avenue, Halifax, NS B3H 2Y9, Canada.
Email: Sheenagh.george@dal.ca

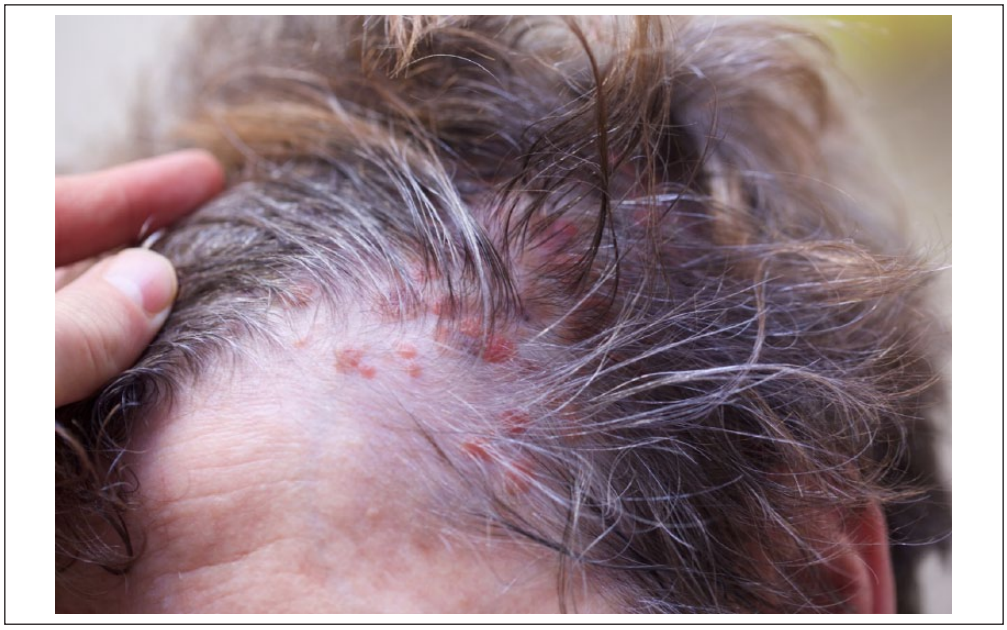

Figure 1. Partial view of frontal-temporal scalp demonstrating mixed morphology of lesions.

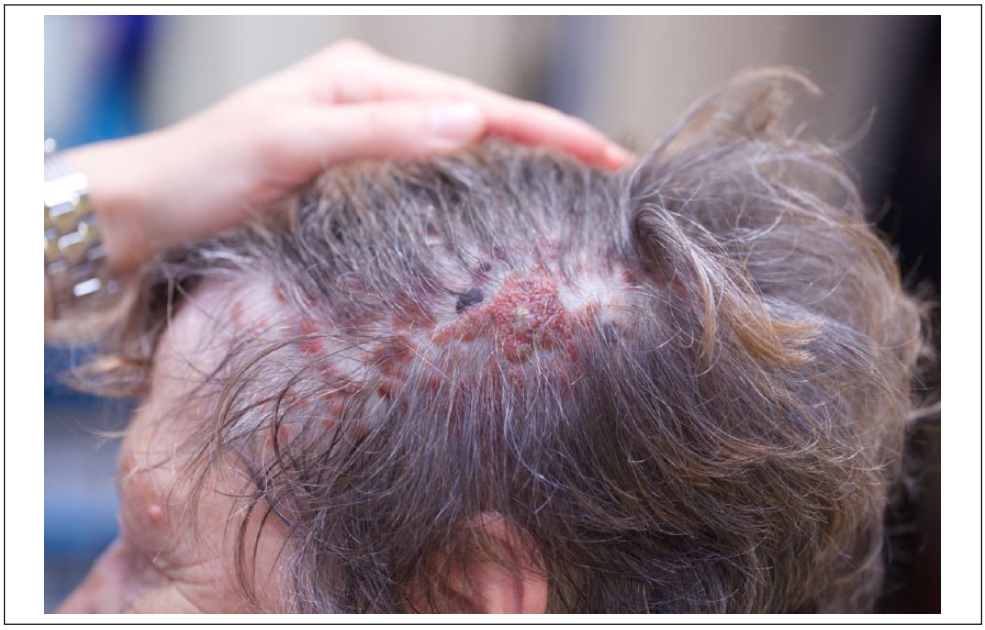

Figure 2. Lateral view of scalp demonstrating heterogeneity of texture among lesions.

were interpreted as showing evidence of malignant melanoma, the component in specimen A exhibiting blue nevus-like features and the component in specimen B being conventional epithelioid and amelanotic. In the setting of partial sampling, it was not possible to determine microscopically whether the melanoma was primary or metastatic. Genetic studies did not reveal mutations in *BRAF, GNAQ*, or *GNA11*.

Clinical investigations included a magnetic resonance imaging (MRI) of the brain and computerized axial tomography (CAT) scan of the chest, abdomen, and pelvis, as well as a positron emission tomography–computed tomography (PET-CT) scan, which showed no evidence of metastatic melanoma. Review by an ophthalmologist excluded a choroidal or other ocular primary melanoma. Surgical resection was not feasible given the extent of disease on the scalp. The patient is currently receiving treatment with intralesional Interleukin-2 (IL-2) injections and has responded quite favourably. Three-month repeat whole body PET-CT scan again showed increased uptake on the scalp, consistent with known clinical disease, but no evidence of obvious metastatic disease. Given biopsy-proven

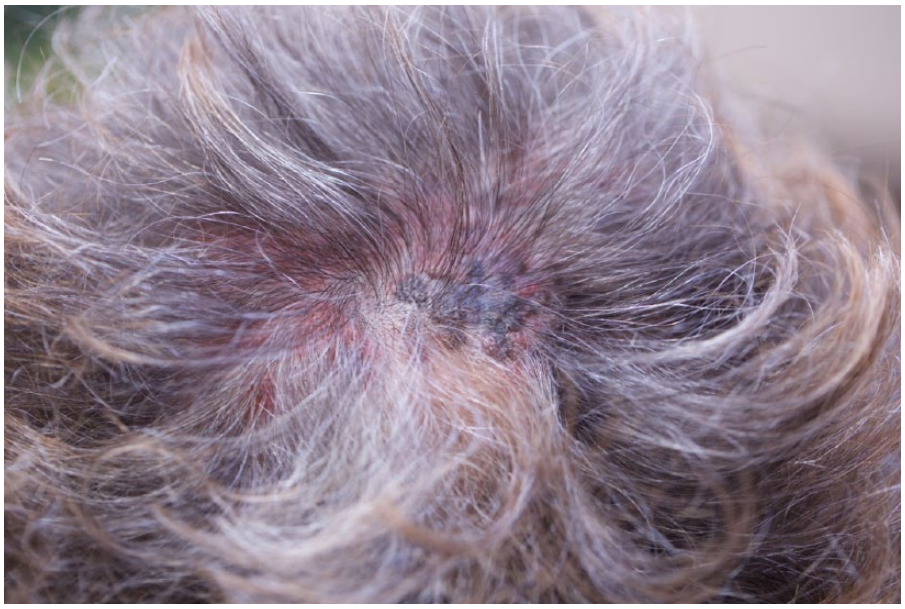

Figure 3. Vertex of scalp, frankly blue-black plaques clearly visible and surrounded by slight erythematous-pink plaques.

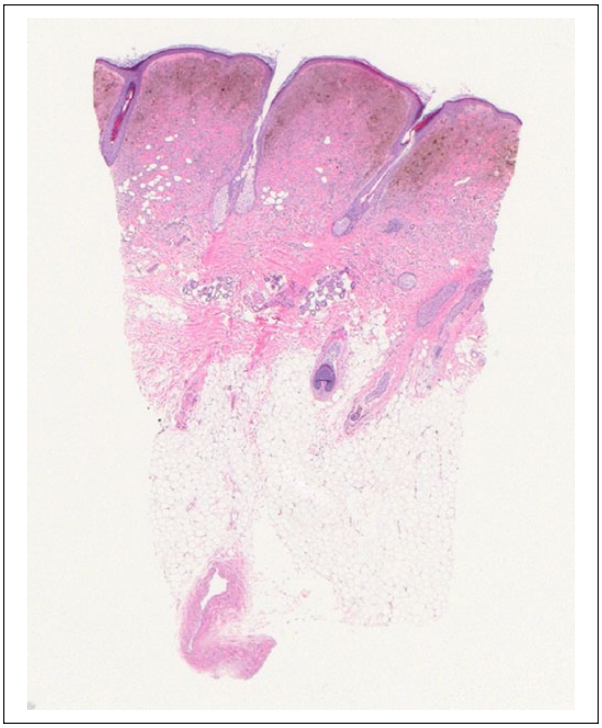

Figure 4. Taken from specimen A. At scanning magnification, the biopsy labelled A displayed a band-like proliferation of deeply pigmented cells in the upper third of the dermis, reminiscent a pattern seen in cellular blue nevi.

progression of scalp melanoma, she was offered Bacillus Calmette–Guérin (BCG) vaccine injection and developed significant systemic symptoms, and will thus continue to receive local therapy. Unfortunately, she remains at very high risk of developing visceral metastases.

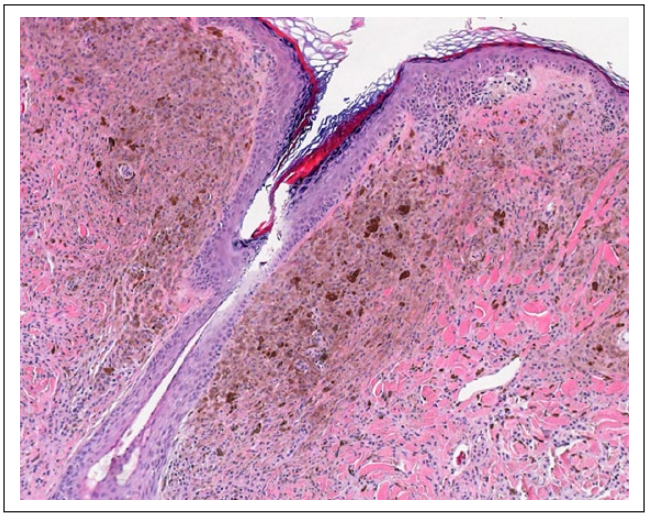

Figure 5. Taken from specimen A. At higher magnification, the cellular infiltrate is seen to be composed of lightly pigmented melanocytes in the background with scattered, admixed, deeply pigmented melanophages. Junctional nests of melanocytes are also evident, a feature not seen in blue nevi.

Discussion

First described by Tieche,[2] a student of Jadassohn, in 1906,[3] the term *blue nevus* refers to a group of benign melanocytic lesions which usually have a blue colour clinically (due to the Tyndall effect) and particular microscopy features. The latter are characterized by fascicular and sheet-like proliferations of slender spindled and dendritic melanocytes, often deeply pigmented and accompanied by melanophages. Different sub-types of blue nevi exist.

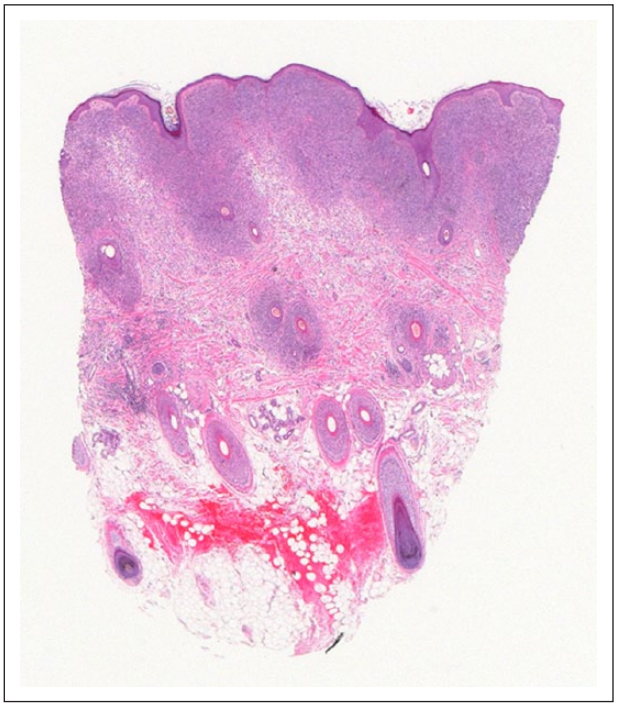

Figure 6. From specimen B. At scanning magnification, the biopsy labelled B displays a dense, sheet-like cellular infiltrate in the upper third of the dermis and in peri-follicular zones.

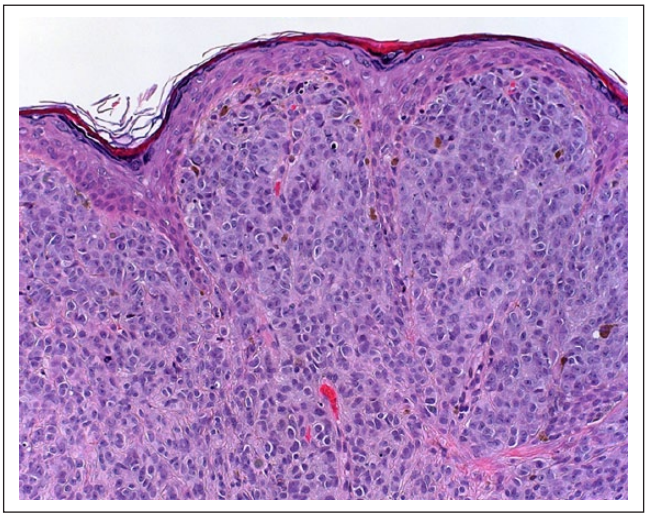

Figure 7. From specimen B. Under higher magnification, the dermal cellular infiltrate is seen to be composed of non-pigmented, atypical, epithelioid melanocytes with occasional mitotic figures.

The term *malignant blue nevus* was first used in 1953 to describe melanoma that demonstrated a readily identifiable benign component,[4] suggesting that the melanoma arose from or in connection with a blue nevus. Since then, the nomenclature has been controversial. The problem appears to be two-fold: using 'malignant' to describe a nevus is perceived by some as an oxymoron,[3] and *malignant blue nevus* as such fails to distinguish between melanoma derived from a blue nevus and melanoma that closely resembles a blue nevus but has no identifiable benign blue nevus component.[3] For these reasons, we use the term *blue nevi-like melanoma* in this communication.

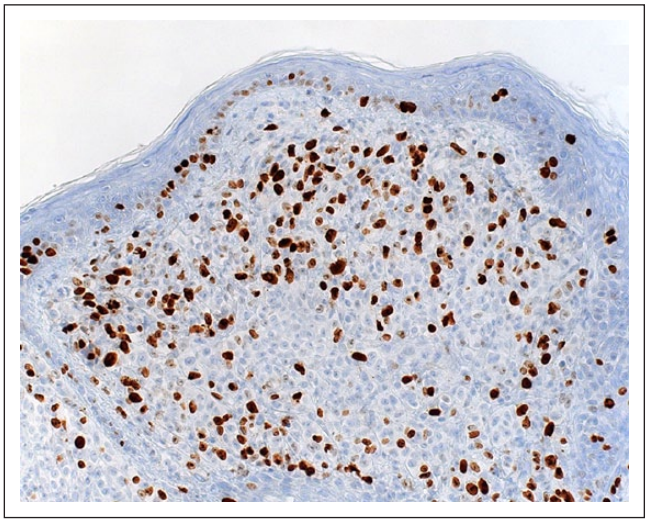

Figure 8. From specimen B. An immunohistochemical stain for Ki67 shows prominent nuclear positivity in lesional cells, indicative of a high proliferative index.

Two aspects of interest in our case include (1) the clinical pattern of local metastasis of a presumed primary melanoma of the scalp and (2) the partial blue nevus-like morphology of the lesion on microscopy. Whether the melanoma in our patient arose in a pre-existing blue nevus or whether it simply exhibits focal blue nevus-like features cannot be determined in the absence of complete excision and comprehensive microscopic evaluation.

As in the case of our patient, surgical excision is the gold standard of treatment for melanoma but is often not feasible with metastases. Traditionally there have been few other alternatives,[5] but in recent years, the field of cancer immunotherapy has grown prodigiously. IL-2, thought to stimulate the body's immune response,[6] is one such therapeutic used in the treatment of metastatic melanoma. Although IL-2 has great promise, it is not without serious risk. Predominantly involving a vasodilatory response, IL-2 therapy-toxicity can include such adverse events as capillary leak syndrome, pulmonary edema, and renal impairment.[7] This is similar to the serious autoimmune reactions observed with other oncological immune modulators, such as checkpoint inhibitor Ipilimumab, also used in metastatic melanoma.[8]

As the role of immunotherapy develops, the dermatologist's role in directing patient care expands as well. Coordinating care between surgeons, oncologists, and managing, the ever-growing list of adjuvant therapy

requires diligence and dedication. This case also highlights the importance of a multidisciplinary approach in complex melanoma management with multiple specialties contributing.

Declaration of conflicting interests

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Ethical approval

Ethical approval was obtained from the hospital's research ethics board.

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Informed consent

Written, documented, informed patient consent was obtained for publication of this case report for both patient information and images to be published.

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