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WNT-activated deep penetrating melanocytoma arising within a congenital melanocytic nevus in a pediatric patient: a case report

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Abstract:

Deep penetrating melanocytoma is an uncommon melanocytic tumor characterized by activation of the WNT/β-catenin pathway and can present a diagnostic challenge because of histologic overlap with melanoma and other melanocytic neoplasms. We report a case of WNT-activated deep penetrating melanocytoma arising within a superficial congenital-pattern melanocytic nevus in a 16-year-old male. A slowly darkening, pigmented papule on the left parietal scalp was initially biopsied locally and demonstrated a biphasic compound melanocytic proliferation with a superficial congenital-pattern component and a deeper nested population of pigmented melanocytes. Histopathologic features included rare dermal mitoses, superficial dermal fibrosis, scattered melanophages, and abundant lymphohistiocytic inflammation. Immunohistochemical studies demonstrated MART-1 expression, focal HMB-45 staining, mosaic p16 expression, negative PRAME staining, and nuclear β-catenin staining within the larger central melanocytes. Expert dermatopathology consultation favored a low-grade WNT-activated deep penetrating melanocytoma arising within the congenital-pattern nevus. The lesion extended focally to the specimen base following conservative excision, and close clinical surveillance was selected. This case highlights the importance of recognizing the characteristic biphasic architecture and immunophenotypic features of WNT-activated melanocytoma, particularly when arising within a conventional melanocytic nevus in a pediatric patient. Accurate distinction from melanoma and other melanocytic mimics can help guide appropriate management and avoid unnecessarily aggressive treatment.

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Introduction

Deep penetrating melanocytoma, historically termed deep penetrating nevus, is a distinctive melanocytic neoplasm first described by Seab and colleagues and now recognized within the spectrum of WNT-activated melanocytic tumors of intermediate biologic potential in contemporary classification systems [1-5]. These lesions most commonly arise in children and young adults and frequently involve the head and neck, often

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presenting as small, pigmented, symmetric papules or nodules that may develop *de novo* or within a pre-existing melanocytic nevus [3, 6, 7]. While most lesions exhibit indolent behavior, recent enlargement or darkening may prompt clinical concern for melanoma, particularly in atypical variants [6, 7].

Histologically, these tumors demonstrate a characteristic wedge-shaped or plexiform dermal proliferation of heavily pigmented epithelioid and spindle melanocytes extending along adnexal and neurovascular structures [3, 4]. Additional features include cytologic uniformity, low mitotic activity, and the presence of melanophages [3, 4]. Immunohistochemically, lesional cells express melanocytic markers such as S100, SOX10, Melan-A, and HMB-45, with distinctive nuclear β-catenin and LEF1 expression, often accompanied by cyclin D1 positivity, reinforcing activation of the Wnt signaling pathway and helping distinguish these tumors from histologic mimics [4, 8, 9]. Molecular studies commonly reveal mutations in CTNNB1, frequently in conjunction with MAPK pathway alterations such as BRAF, HRAS, or MAP2K1, supporting a stepwise model of tumorigenesis [9-12].

The differential diagnosis is broad and includes blue nevus, cellular blue nevus, Spitz nevus, pigmented epithelioid melanocytoma, and melanoma, particularly deep penetrating nevus-like melanoma, making accurate recognition essential due to the vastly different therapeutic and prognostic implications [3, 4, 7, 12, 13]. Most WNT-activated melanocytomas are effectively treated with complete surgical excision and carry an excellent prognosis; however, atypical lesions may demonstrate regional lymph node involvement, necessitating careful clinicopathologic correlation and follow-up [7, 13, 14].

Herein, we report a case of a deep penetrating (WNT-activated) melanocytoma arising within a conventional melanocytic nevus on the scalp of a 16-year-old male, highlighting its diagnostic features and relationship to existing literature.

Case Presentation

A 16-year-old Caucasian male with a noncontributory dermatologic history presented to a dermatology provider with concerns about a recently evolving brown mole on the left parietal scalp. The lesion had been documented two years prior by his pediatrician (Figure 1) but was determined to not be of concern at that time due to stability and benign appearance. Observation was deemed appropriate. Darkening coloration prompted re-evaluation by dermatology. A 9 x 7 mm brown nevus with a contained fried egg-type papule of darker coloration was documented on examination (Figure 2) and shave biopsy was performed.

Histologic examination revealed compact orthokeratosis overlying a biphasic compound melanocytic proliferation (Figure 3). The superficial congenital-pattern component demonstrated single-cell and nested growth with bridging of adjacent rete ridges (Figure 4). The melanocytes had small, round nuclei with variably prominent nucleoli and variable pigmentation (Figure 5). Within the deeper central dermis, a second nested melanocytic population demonstrated intermediate to large amphophilic oval nuclei, abundant pale cytoplasm, and prominent melanin pigment (Figure 6). Rare dermal mitotic figures, superficial dermal fibrosis, scattered melanophages, and abundant lymphohistiocytic inflammation were also present.

Immunohistochemistry demonstrated MART-1 expression in the melanocytic proliferation without significant pagetoid scatter (Figure 7), focal HMB-45 staining of dermal melanocytic nests (Figure 8), mosaic p16 expression (Figure 9), and negative PRAME staining (Figure 10). Nuclear β-catenin staining highlighted the larger central melanocytes, supporting their classification within the deep penetrating nevus family. Overall, the histomorphologic and immunohistochemical findings supported a low-grade deep penetrating (WNT-activated) melanocytoma arising within a superficial congenital melanocytic nevus. The melanocytoma extended focally to the specimen base, and conservative re-excision or clinical follow-up was recommended.

Three months later, patient was seen in follow-up, at which time focal persistence of the pigmented papule was present at the biopsy site (Figure 11).

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With consideration of the histologic diagnosis, clinical persistence, and difficulty monitoring the anatomic region, the patient was referred to procedural dermatology for definitive excision. One month later, excision of the lesion was performed with conservative 2 mm margins circumferentially. Positive margins were identified on the excision specimen.

With no appreciable clinical recurrence and relative intolerance of the recovery from prior excision, the patient elected for surveillance and continues following closely with dermatology and primary care.

Discussion

Deep penetrating (WNT-activated) melanocytoma represents a distinctive melanocytic neoplasm characterized by activation of the Wnt/β-catenin signaling pathway and a clinicopathologic spectrum that ranges from benign lesions to tumors with atypical features and rare malignant potential [7, 9, 15, 16]. Our patient's presentation of a pigmented lesion on the scalp of an adolescent arising within a pre-existing melanocytic nevus aligns well with previously reported clinical patterns [3, 6, 9]. The head and neck predilection and occurrence in younger individuals have been consistently documented in the literature, reinforcing the characteristic demographic and anatomic distribution of these tumors [7, 11]. The association with a conventional nevus, as observed in this case, further supports the concept that WNT-activated melanocytomas may develop through stepwise tumor progression involving both MAPK and Wnt pathway activation [9, 10, 12]. The clinical evolution of darkening, however, prompted concern for melanoma and underscores the potential difficulty of distinguishing these lesions clinically.

In our case, the lesion demonstrated classic histopathologic features of a deep penetrating melanocytoma, characterized by a biphasic compound melanocytic proliferation with a superficial congenital-pattern nevus and a deeper nested dermal proliferation of pigmented melanocytes [3, 9, 12]. The lesion exhibited cytologic uniformity, superficial dermal fibrosis, scattered melanophages, a prominent lymphohistiocytic infiltrate, and rare dermal mitotic figures [3, 4, 13]. Despite the presence of rare mitoses, the overall cytologic and architectural features supported classification within the low-grade end of the biologic spectrum [2-4, 7, 15, 16]. Immunohistochemical analysis supported the diagnosis, with MART-1 highlighting the melanocytic proliferation and focal HMB-45 staining of dermal nests [4, 6]. Mosaic p16 expression and negative PRAME further supported a lesion with low malignant potential [2, 17]. Expert dermatopathology consultation favored a WNT-activated melanocytoma, with nuclear β-catenin positivity in the larger central melanocytes supporting their classification within the deep penetrating nevus family [9, 12, 13].

A critical aspect of this case is the distinction from melanoma and other melanocytic tumors with overlapping features, particularly in the setting of concerning clinical changes [5, 7, 16]. The deep infiltrative growth pattern and pigmentation can closely mimic blue nevus-like melanoma or pigmented epithelioid melanocytoma, while the epithelioid cytomorphology may raise concern for Spitz melanoma [3, 4, 6]. Recognition of the characteristic architectural symmetry, maturation with depth, and nuclear β-catenin expression is therefore essential to avoid misdiagnosis and unnecessary aggressive management [3, 8, 9]. Similar diagnostic challenges have been emphasized in the largest published series, underscoring the importance of integrating histologic, immunohistochemical, and molecular findings [3, 12, 13].

From a pathogenetic standpoint, the coexistence of a conventional melanocytic nevus in this patient supports the model of dual-pathway activation, wherein an initial MAPK pathway mutation gives rise to a benign nevus, followed by acquisition of CTNNB1 mutations leading to Wnt pathway activation and development of a deep penetrating melanocytoma [9-12]. This stepwise progression provides a biologic framework for understanding these lesions within the continuum of melanocytic tumorigenesis [5, 7, 16].

Management of WNT-activated melanocytomas generally favors complete surgical excision with histologically negative margins [6, 7, 14]. While most lesions behave in a benign fashion, atypical variants have been associated with regional lymph node involvement, though this does not necessarily

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predict aggressive clinical behavior [7, 14, 15]. In the present case, conservative excision with clinical follow-up was deemed appropriate given the absence of high-risk histologic features, supporting the concept of management based on individual clinicopathologic assessment.

Conclusion

WNT-activated deep penetrating melanocytoma is a rare melanocytic neoplasm that can present diagnostic challenges because of its clinical and histopathologic overlap with melanoma and other melanocytic tumors. Recognition of its characteristic architecture and immunophenotypic features, supported by expert dermatopathology consultation, when needed, is essential for accurate diagnosis and appropriate individualized management. Continued documentation of such cases, especially with long-term follow-up, will further refine our understanding of their biologic behavior and contribute to the development of evidence-based management strategies.

Take Home Messages

Deep penetrating (WNT-activated) melanocytoma is an uncommon melanocytic tumor that may arise within a conventional melanocytic nevus, including in pediatric patients.

Its histopathologic features may overlap with melanoma and other melanocytic tumors, making careful assessment of architectural and cytologic features essential for accurate diagnosis.

Expert dermatopathology consultation and β-catenin immunohistochemistry can support accurate diagnosis and help guide individualized management.

Declarations

Author Contributions:

Linze M. Christensen: investigation (lead), writing – original draft (lead), writing – review & editing, resources, project administration. Stephanie R. Summers: investigation, visualization, writing – original draft, writing – review & editing. Amy M. Kerkvliet: investigation, visualization, writing – original draft, writing – review & editing. Marcus L. Frohm: conceptualization (lead), supervision (lead), project administration, writing – review & editing.

Funding: No external/specific funding was received for this work.

Ethics Approval: Ethical approval was not required for this case report according to Sanford IRB guidelines following the DHHS definition of research.

Patient Consent: Written consent has been obtained from the patient's guardian.

Conflict of Interest: The authors declare no competing interests.

Data Availability: All relevant information is included in the article. Additional patient-level data are not publicly available due to privacy and confidentiality considerations.

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Figure Legends

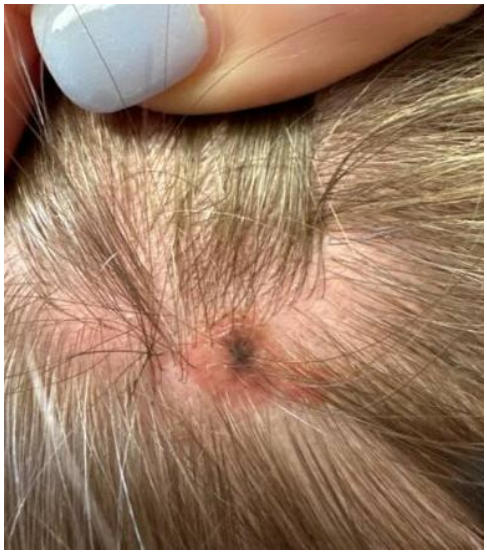

Figure 1: Clinical appearance of lesion at initial presentation, approximately 2 years prior to excision. Note the central focus of darker pigmentation.

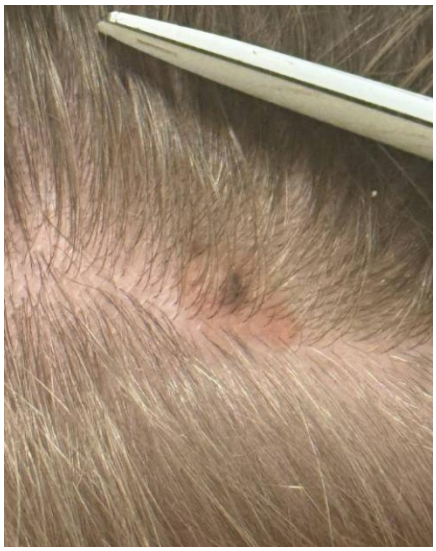

Figure 2: Clinical photograph at dermatologic evaluation prompted by darker pigmentation. Well-demarcated brown macule with a contained central darker papular component, producing a “fried egg” appearance on the left parietal scalp.

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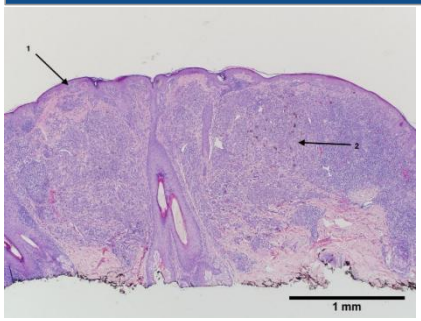

Figure 3: Hematoxylin and eosin-stained sections demonstrate a compound biphasic melanocytic proliferation. The superficial component is the congenital-pattern melanocytic nevus (1) and the deeper nested component is the deep penetrating melanocytoma (2).

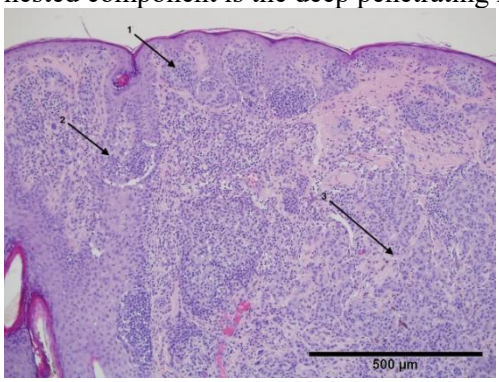

Figure 4: Hematoxylin and eosin-stained sections demonstrate the superficial melanocytic proliferation occurring in a nested and single cell distribution (1). The melanocytes are tracking down the hair follicle, evidence of congenital type pattern (2). There is mild papillary dermal fibrosis and associated lymphohistiocytic inflammation. The deeper dermal component shows intermediate to large amphophilic melanocytes with scattered melanin pigment (3). H&E, 100X

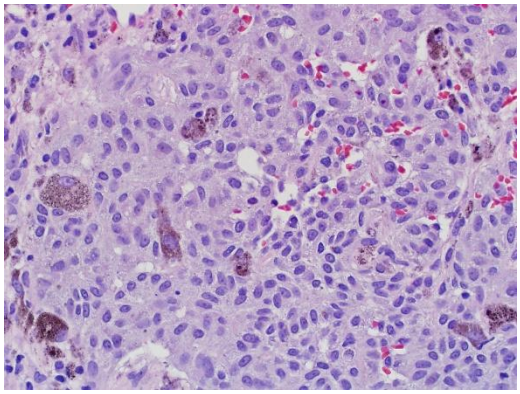

Figure 5: Hematoxylin and eosin-stained sections demonstrate the superficial melanocytic nests having small, round to oval nuclei, with variably prominent nucleoli. The melanocytes are both well nested and occurring singly. H&E, 400X

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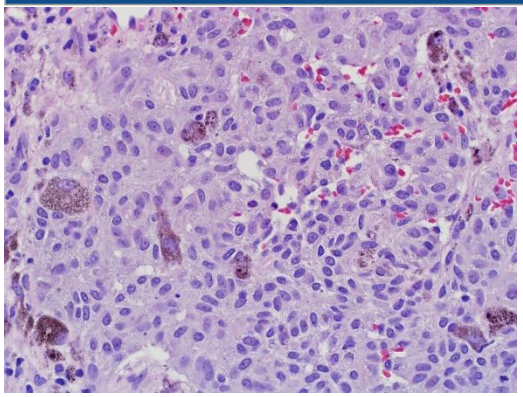

Figure 6: Hematoxylin and eosin-stained sections demonstrate the deeper dermal melanocytes with intermediate to large, amphophilic oval nuclei with abundant pale cytoplasm and prominent melanin pigment. H&E, 400X

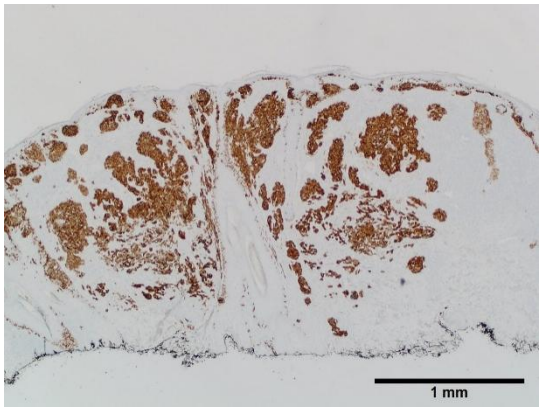

Figure 7: MART-1 immunohistochemical stain highlights both the superficial and deep melanocytic distribution. 40X

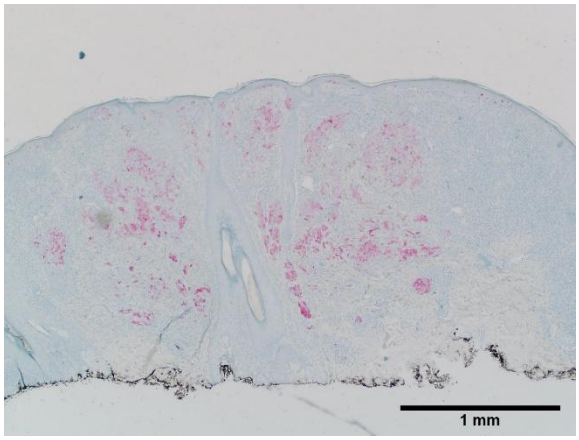

Figure 8: HMB-45 immunohistochemical stain focally highlights the dermal melanocytes. 40X

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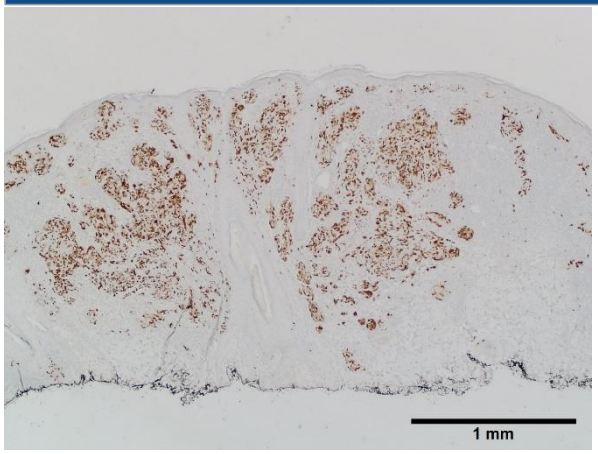

Figure 9: P16 immunohistochemical stain demonstrates a mosaic staining pattern. 40X

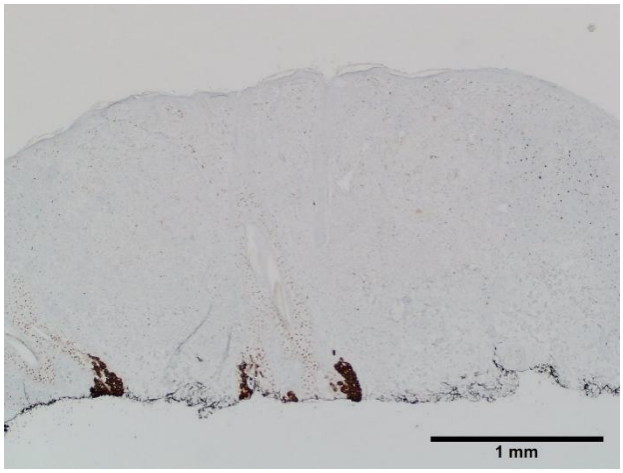

Figure 10: PRAME immunohistochemical stain is negative. The positive staining is the internal control demonstrated by adnexal structures. 40X

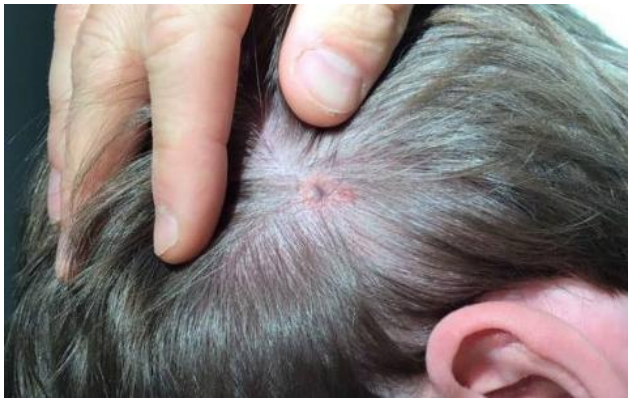

Figure 11: Clinical appearance of lesion 4 months post-shave biopsy. Focal persistence of the central pigmented papular component.