SKIN

BRIEF ARTICLE

Pediatric Occurrence of Osteonevus of Nanta: A Case Report

Julian Melchor, MD[1], Genevieve Patrick, MD[2], Alexandra Mathis, MD[3], David Pascoe, MD[4], Lawrence Rosenberg, MD[5], Erin Carlquist, MD[6]

[1] Duke Family Medicine & Community Health Residency Program, Durham, NC, USA
[2] Florida State University College of Medicine, Tallahassee, Florida, USA
[3] HCA FL Orange Park Transitional Year Residency Program, Jacksonville, FL, USA
[4] Family Dermatology of North Florida, Tallahassee, Florida, USA
[5] Southeastern Plastic Surgery, Tallahassee, Florida, USA
[6] KWB Pathology Associates, Tallahassee, Florida, USA

ABSTRACT

Secondary ossification is a process that occurs in various cutaneous conditions. When this occurs within a melanocytic nevus, it is termed Osteonevus of Nanta (ON). This rare phenomenon typically presents in elderly female individuals usually located on the upper trunk. In this report, we present a unique presentation of ON in a pediatric patient.

A 2-year-old female with no significant past medical history presented to a plastic surgeon for excision of an asymptomatic soft tissue mass on the right scalp. The parents described the gradual postnatal development of the 1.2 x 0.4 x 0.3 cm red-brown nodule. Excisional biopsy was performed, and histopathological analysis found the lesion to be most consistent with ON.

To our knowledge, there are no other published reports of ON in the pediatric population. This exceedingly rare case serves to expand our current understanding of this lesion and may provide further insight into the pathogenesis of these nevi.

INTRODUCTION

Secondary ossification of the skin is a process that occurs in various cutaneous conditions, such as basal cell carcinoma, scars, and sites of previous trauma.[1] When secondary ossification forms within a melanocytic nevus, it is termed Osteonevus of Nanta (ON).[1] This rare phenomenon, with an estimated incidence of 0.6-1.45%, typically presents in elderly women on the superior portion of the body.[2,3] A majority of cases specifically occur on the face or scalp and may occur secondary to a traumatic process.[1] Due to limited clinical occurrences of ON, it is often misdiagnosed as melanoma or junctional nevi.[2] Based on limited findings from available literature, there are no characteristic clinical findings that would support a diagnosis of ON without histopathological examination. Histological features that support a diagnosis of ON are nests of melanocytic cells within osseous tissue.[2] Current recommendations suggest these lesions be completely excised as there have been reports of progression to melanoma.[2,4] In this case report, we present a unique presentation of ON in a pediatric patient as evidenced by histopathological examination.

CASE REPORT

A 2-year-old female with no significant past medical history presented to the outpatient plastic surgeon clinic with her caregiver for excision of an asymptomatic soft tissue mass located on the right scalp. The parents reported the gradual postnatal development of this lesion. On clinical examination, there was 1.2 x 0.4 x 0.3 cm red-brown nodule. The patient's parents denied any family history of melanoma or any other dermatological-related conditions. Our clinical diagnosis was soft tissue mass versus cephalohematoma. An excisional biopsy of the mass was performed and sent for histopathological preparation due to clinical uncertainty and cosmetic purposes. Photomicrographs of the lesion with hematoxylin and eosin (H&E) and immunostaining are demonstrated (**Figure 1-3**).

Microscopic Findings and Clinical Course

Biopsy fragments were composed of both non-pigmented nevus and pigmented epithelioid and focally spindle/dendritic melanocytic elements devoid of significant atypia but associated with foci of ossification. Histopathological analysis found the lesion to be most consistent with combined nevus (nevocellular nevus and blue nevus with epithelioid nevus elements). Ki-67 staining was performed to evaluate the level of atypia and demonstrated reassuring signs of predominant cytoplasmic staining. Due to the rarity of this presentation, the histopathology was sent to another dermatopathologist for verification. Both dermatopathologists confirmed the diagnosis of Osteonevus of Nanta.

DISCUSSION

Secondary cutaneous ossification characterizes the cutaneous bone formation associated with a pre-existing lesion. Osteonevus of Nanta, first described in 1908 and subsequently termed in 1911, characterizes the rare phenomenon of focal secondary ossification occurring within, or adjacent to, a melanocytic nevus.[6,7] These lesions are typically reported among older individuals, usually in the 3rd-5th decade of life, with an apparent predilection for women.[7,8]

Histopathologically, ON is defined as the formation of bone adjacent to dermal nests of melanocytes, yet there are various documented manifestations.[3] For example, ossification, composed of lamellar and amorphous compact bone, may present as a single or multiple foci within a nevus, as well as within the reticular dermis surrounding hair follicles.[1] Additional microscopic findings, such as mononuclear infiltrates, foreign body granulomas, or mixed granulomas, may also be observed within the lesion.[1,2] Bone tissue formation within pigmented cutaneous lesions, such as melanocytic, intradermal, combined, Blue, Spitz, and Becker nevi, is exceedingly rare but has been previously reported.[1,3,4]

There are several proposed mechanisms for ON's pathogenesis, including secondary ossification as a fibroblast metastatic response to dermal inflammation or trauma, hamartomatous proliferation of embryonic mesenchymal cells driven by osteogenic factors, and metaplasia of primitive mesenchymal cells.[4,7,8]

Although a less likely etiology in this pediatric patient, prior trauma or long-standing inflammation to a pre-existing melanocytic nevus can result in metaplastic ossification.[3] This typically occurs following repetitive traumatic injury of the follicles or disruption of

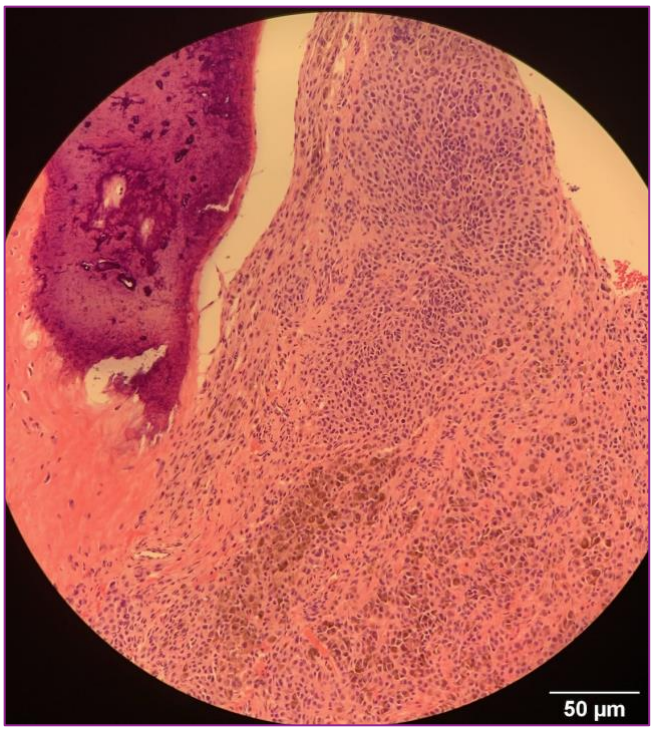

Figure 1. Macro H&E (50x) displaying both non-pigmented nevus and pigmented epithelial and focally spindle/dendritic melanocytic elements.

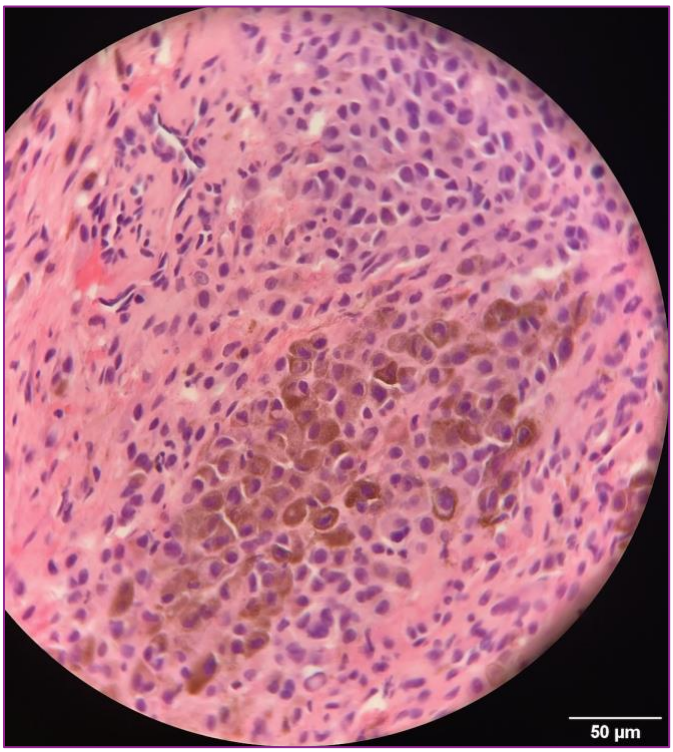

Figure 2. Micro H&E (630x) displaying the melanocytic components associated with foci of ossification.

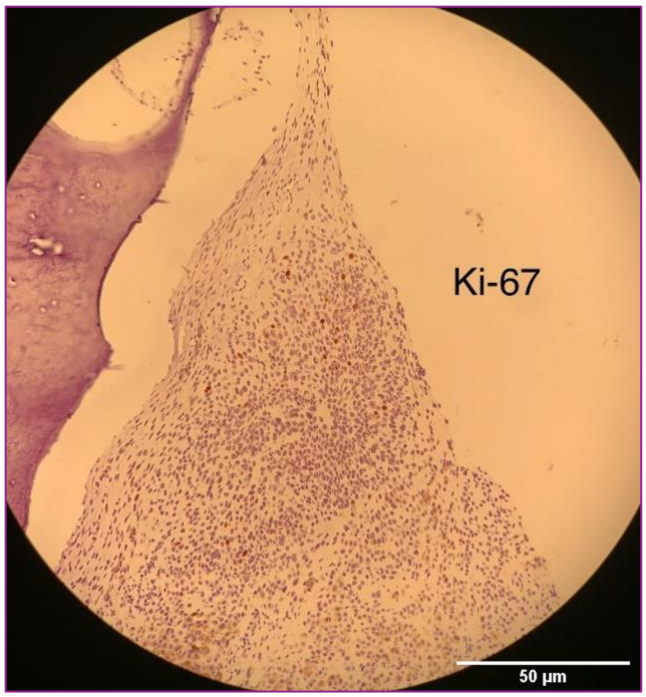

Figure 3. Ki-67 Stain (50x) appears to be devoid of any significant atypia due to the largely cytoplasmic cellular findings.

the melanocytes.[3] Conversely, hamartomatous proliferation of embryonic germ cells, from both ectodermal and mesodermal origin, may account for the concurrent presence of osseous components within a melanocytic nevus.[8] There may be a role of Cbfa1 transcription factor in the development of heterotopic secondary ossification of cutaneous lesions.[9] Cbfa1 is expressed in fibroblasts within the dermis during fetal development with a specific role in osteoblast-specific gene expression and differentiation.[10] It has also been implicated in autoregulation of bone matrix deposition of differentiated osteoblasts.[10] Therefore, mutations in this gene may result in dysregulated deposition of a bony matrix within the dermis.

Apart from well-documented associations of GNAS mutations with a multitude of heterotopic ossification disorders, there have been previous reports of GNAS-induced infantile osteoma cutis associated with dermal spindle cells.[11,12] This suggests another possible genetic etiology for development of secondary ossification centers within pre-existing lesions. Furthermore, the hypothesized role of estrogen in osteogenic bone formation may account for the disproportionate female predominance of ON, bolstering the evidence of osteogenic factor influence.[1,7] Finally, the theory of metastatic proliferation of primitive mesenchymal cells in ON development may be supported by reported cases detailing lesion progression to malignant melanoma.[2,4] Although the exact mechanism remains ambiguous for this pediatric patient, we hope to illuminate potential pathogenic possibilities of ON among this unique patient population.

Clinical ambiguity regarding the appearance and histopathological diagnosis of ON necessitates management airing on the side of safety.[2] The gold standard for diagnosis presently remains light microscopy.[2] Some clues that may be seen under dermoscopy may include pigmented globules, hypopigmented border, light brown to

translucent appearance, and a pigmented network.[2] The patient described in this case underwent a complete excisional biopsy of the lesion with independent histopathological confirmation by two pathologists. Close clinical follow-up was recommended following excision to monitor wound healing and any potential signs of recurrence. Furthermore, an ultrasound was performed 1-year status post excision to ensure rule out of malignancy. The ultrasound demonstrated a stable, small hypoechoic nodular focus in the superficial subcutaneous right posterior scalp soft tissue, indicating a good prognosis. The patient will continue to follow up with a dermatologist for regularly scheduled monitoring of the area.

CONCLUSION

To our knowledge, there are currently no other literature sources reporting the occurrence of Osteonevus of Nanta in pediatric populations. We present a case of ON in a pediatric patient treated with complete excision of the mass. This exceedingly rare presentation may provide further insight into the pathogenesis of these lesions and expand our current understanding of their associations.

Conflict of Interest Disclosures: None

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Corresponding Author:
Julian Melchor, MD
Duke University Medical Center
Department of Family Medicine & Community Health
Phone: (305)-878-0737
Email: jm960@duke.edu

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