Osteonevus of Nanta in a 3-year-old child with dermatoscopic characterization

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INTRODUCTION

Osteonevus of Nanta is a rare and peculiar benign skin lesion, histologically characterized by the presence of osseous metaplasia in a melanocytic nevus.[1] This condition was first described in an intradermal nevus by Heidingsfeld in 1908 and later named osteonevus of Nanta in 1911; since then, cases of osseous metaplasia occurring in blue and combined nevi have also been reported.[2,3] Osteonevus of Nanta is commonly diagnosed in adults, with most cases being reported in women between the fourth and the fifth decades of life.[4] Its precise incidence is not known, but it is considered a rare condition, with an estimated incidence of approximately 0.14% among pigmented skin lesions.[5]

Osteonevus of Nanta commonly presents as a firm, dome-shaped or pedunculated papule or nodule, usually asymptomatic. It is mostly found on the head and neck region but can occur on any part of the body. Histologic examination shows mature bone tissue adjacent to or interposed with melanocytic cells, typically at the base of the lesion.[6] Here, we report the case of an osteonevus of Nanta in a 3-year-old child, also illustrating its atypical dermatoscopic features.

Abbreviation used:

JXG: juvenile xanthogranuloma

CASE PRESENTATION

A 3-year-old White boy was referred to our Pediatric Dermatology Unit for a completely asymptomatic, papular, dome-shaped lesion on the left cheek. The lesion appeared one month after birth and had been reported as rapidly growing in the last 3 months.

Clinically, the lesion showed a maximum diameter of 4 mm, and it was elastic and firm to palpation (Fig 1, *A*). Dermatoscopy revealed hairpin and branched linear vessels in a periphery-to-center pattern; also, white-to-yellowish streaks coalescing into small patches were evident in the central area. The lesion was surrounded by a slightly erythematous border, configuring an overall setting in a sun-like pattern. There was no pigment network or other dermatoscopic signs suggestive of a melanocytic lesion (Fig 1, *B*).

The clinical suspicion was juvenile xanthogranuloma (JXG). However, given the nonspecificity of the clinical and dermatoscopic findings and the progressive enlargement in the last months, the lesion was surgically removed for histologic examination.

Histology revealed the presence of an intradermal nevus with features of maturation (cells becoming smaller and more dispersed with depth) associated with adjacent multiple foci of ossification (Fig 2, *A-C*); within the mature lamellar bone tissue, intraosseous

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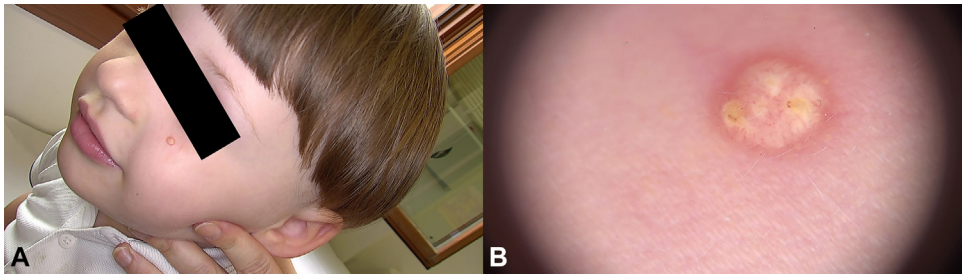

Fig 1. **A**, Dome-shaped papular lesion on the left cheek of a 3-year-old boy. **B,** Dermatoscopy of the lesion showing white-to-yellowish small patches in the central area, hairpin, and branched linear vessels at the periphery, all surrounded by a slightly erythematous border.

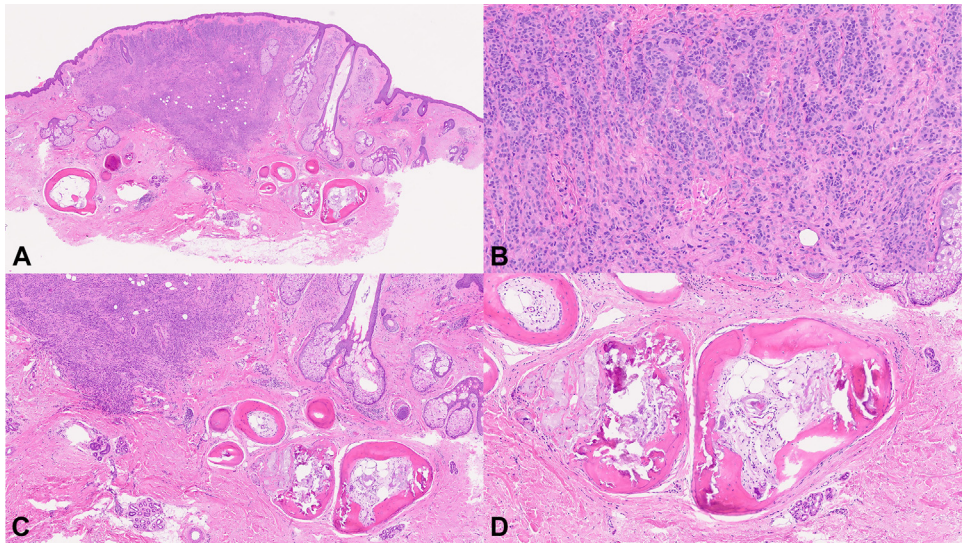

Fig 2. Histology of the lesion (hematoxylin and eosin stain) showing an intradermal nevus with melanocytes arranged in nests and multiple foci of ossification (**A-C**) at higher magnification connective tissue and mature fat cells accompanied by blood vessels were enclosed in the bone areas (**D**).

vascularization, and marrow elements were also detected (Fig 2, *D*).

DISCUSSION

Our case is one of the few reports of osteonevus of Nanta in the pediatric population. In their case series, including 33 histopathologically-confirmed osteonevi, Bezić et al[6] reported that the age of patients with osteonevi was significantly higher than the age of patients in the control group of nonossifying nevi. Furthermore, in the case series by Moulin et al[4] and Sasaki et al,[7] the patients' mean age was 46 and 45 years, respectively. The results of these studies provide evidence that ossification within nevi typically occurs in adults and tends not to occur in the pediatric age. The diagnosis of a growing papular lesion on the face of a child can be very challenging, and requires a comprehensive evaluation, including clinical, dermatoscopic, and sometimes histologic examination. As for our case, JXG was included in the differential diagnosis. JXG is a benign form of non-Langerhans cell histiocytosis, which presents as solitary or multiple yellow-orange firm papules or nodules. It typically occurs in early childhood but can develop at any age, and it is usually asymptomatic. In their retrospective study, Peruilh-Bagolini et al[8] recently reviewed the clinical and dermatoscopic characteristics of JXG, concluding that it usually appears as a symmetric lesion, whose main dermatoscopic characteristics include yellow/orange-pink/red colors, yellow globules, shiny white streaks, together with irregularly distributed vascular structures of different types. The association of a yellow-orange structureless central area and a slightly erythematous border (also referred to as "setting sun pattern") has been previously suggested as a dermatoscopic pattern characteristic of JXG; however, it should be noted that the specificity of this

pattern appears to be limited.[8] In the presented case, the presence of a setting sun-like pattern, with a central yellowish area circumscribed by a mildly erythematous ring, in association with abundant vascular structures, could be suggestive of JXG. However, in most cases JXG is a symmetric lesion, a characteristic that was absent in our case.

In our case, the lesion was also lacking any sign suggestive of a melanocytic lesion. On the contrary, in all the reported cases of osteonevi where dermatoscopy was available, some evidence of a melanocytic lesion was present. López-Robles et al[2] reported the case of an osteonevus arising in a blue nevus where, dermatoscopically, a brown pigmented network was evident in the center of the lesion, surrounded by whitish to light pink areas at the periphery.[2] In their case series of 2 ostonevi, Sławińska et al[9] found brown structureless areas in both cases. Finally, Breunig et al[5] reported a case of osteonevus of Nanta where a light brown pigmentation, blue-gray areas, and multiple black dots were present.[5]

Regarding pathophysiology, the exact mechanism underlying ossification in osteonevus of Nanta is not known. Two primary hypotheses have been proposed: the presence of either bone metaplasia or a hamartomatous lesion.[3] With regards to the former hypothesis, repeated trauma, chronic inflammation, or even melanocytic proliferation are capable of inducing the differentiation of dermal fibroblasts into osteoblasts.[7] According to the second hypothesis, there are mesenchymal stem cells (capable of differentiating into osteocytes) present at aberrant sites that ultimately lead to the formation of a hamartoma with coexistence of cells from both mesodermal and ectodermal origins. Finally, the potential role of estrogens in bone formation has been postulated to explain the higher incidence of osteonevus of Nanta in adult women.[2] Osteocytes express estrogen receptors and their activation can promote bone formation.[6] Our case of osteonevus occurring in a male prepubertal child suggests that estrogen receptors activation in osteocytes might not be strictly necessary for bone formation in the context of a melanocytic nevus.

In conclusion, osteonevus of Nanta may have heterogeneous clinical characteristics and the gold standard for diagnosis remains histopathology; further studies are certainly needed in order to elucidate clinical and dermatoscopic characteristics of this peculiar lesion. In clinical practice, the diagnosis of osteonevus of Nanta should also be considered, although the diagnosis of papulo-nodular lesions without specific clinical features could be very challenging, especially in the pediatric age.

Conflicts of interest

None disclosed.

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