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Primary isolated osteoma cutis of the face

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Abstract

We report a healthy, 44-year-old woman presenting with an at least a 20-year history of hardened papules in the forehead region, extending to the scalp. The biopsy and histopathologic exam confirmed a diagnosis of osteoma cutis. We review the literature review and discuss the classification of the cutaneous ossification process presented, along with the results of the surgical treatment.

Keywords: osteoma cutis, cutaneous ossification, osteogenesis

Introduction

Cutaneous ossifications are unusual pathological processes that occur with the formation of bone tissue within the dermis and hypodermis; they are classified as either primary or secondary. The secondary forms are more frequent and can result from various stimuli, such as accidental or surgical trauma, acne scars, and skin tumors, whereas the primary forms develop in the absence of currently known triggering factors [1].

For the primary type, it is proposed that the mechanism of formation be divided into two subtypes: endochondral and intramembranous. The former is characterized by the involvement of the skin and subcutaneous tissue by direct extension of the bone structure. This form is typically more severe and progressive ossifying fibrodysplasia is the most representative type within the group. In the latter, intramembranous ossification begins in the dermis, comprising three entities: progressive

osseous heteroplasia (POH), Albright hereditary osteodystrophy, and primary osteoma cutis (POC) [1].

The plate-like osteoma cutis, although very rare, accounts for most cases of POC. Some authors point out that it could represent a localized, non-progressive form of POH [1].

We report a case of a patient with isolated lesions on the face, histologically compatible with primary dermal ossification, and we attempt to frame this POC subtype according to the literature.

Case Synopsis

A 44-year-old woman, complained of papular lesions on the forehead that emerged 20 years prior to presentation. She noted slight growth at the beginning, but the papules had been stable for at least 15 years. She denied any history of acne or previous trauma.



Figure 1. Indurated papular lesions grouped on the forehead.



Figure 2. Alopecia plaque, with indurated papular lesions.



Figure 5. After surgical excision, satisfactory cosmetic result.

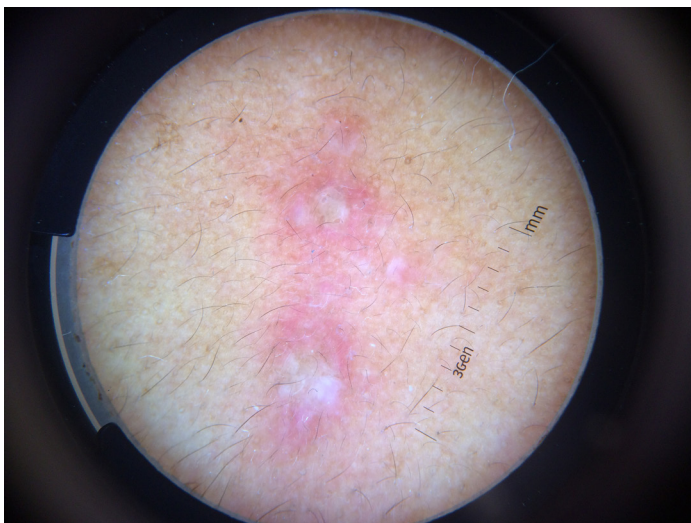


Figure 3. Amorphous yellowish-white central area, surrounded by discrete erythema. (Dermoscopy, 10x).

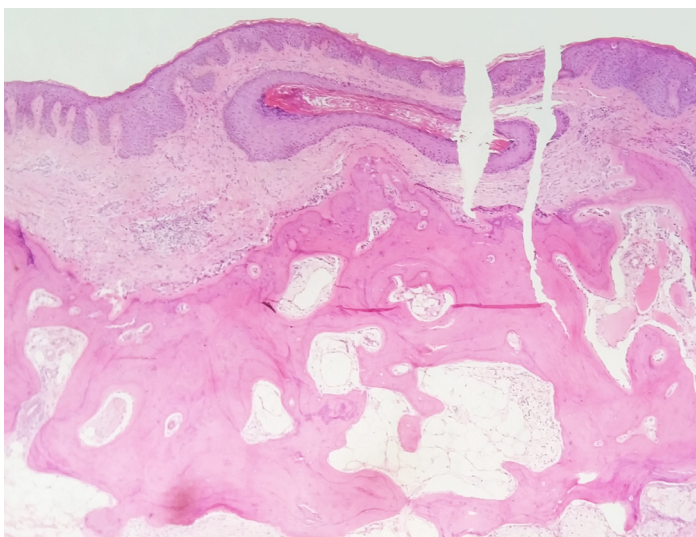


Figure 4. Bone trabeculae occupying the majority of the dermis, extending to the dermo-hypodermic junction (H&E, 10x).

The dermatological exam showed papules with discrete erythema without scale around a whitish center. The papules were firm to palpation and grouped on the forehead (**Figure 1**). In addition, a plaque of alopecia was noted, adjacent to the forehead lesions, which on closer examination showed similar papules (**Figure 2**). The examination also showed mild seborrhea of the facial skin, without comedones, papules, or pustules. The patient did not have acne scars.

A dermoscopy examination of the forehead lesions reinforced the clinical exam, identifying an amorphous yellowish-white central area, surrounded by discrete erythema (**Figure 3**).

An incisional biopsy was performed. A bone trabeculae occupying most of the dermis and extending to the dermo-hypodermic junction was observed in the histopathological exam, which is compatible with skin ossification (**Figure 4**). A simple skull radiograph and laboratory tests were normal.

After a discussion with the patient, performed a surgical excision with satisfactory cosmetic results (**Figure 5**). The patient is still in follow-up, with no recurrence after 6 months.

Case Discussion

Primary ossifications of the skin are rare. In a retrospective study of 425 cases of cutaneous ossification, 14% of cases were primary. Within this group, the cutaneous limited forms represented the

majority. These are encompassed generically within the POC group; they are more frequent in women [2].

The pathogenesis of the POC group is still uncertain, although some theories have been formulated. Among them, the following two are the most widely accepted: 1) primitive mesenchymal cells differentiate into osteoblasts and migrate to unusual places - in this specific case, to the skin; or 2) metaplasia of the skin's mesenchymal cells (fibroblasts) give rise to osteoblasts and culminate in bone formation [3].

Clinical aspects define the different subtypes of POC. Four forms are proposed: plate-like, miliary, isolated, and disseminated [4].

Plate-like POC is a subtype described in 1978 by Worret and Burgdorf, generally identified by a plaque that is hard on palpation. As described above, it represents the majority of cases of POC. It affects children and can present at birth or in the first few years of life. The same authors reviewed 12 cases and found the following diagnostic criteria: injury identified at birth or in the first year; no changes in calcium and phosphorus metabolism; no triggering factors (history of infection, trauma); presence of at least one bone plate, with or without other osteoma cutis [5]. However, cases of similar clinical lesions in adult patients have also been published [6].

Another well-known subtype is miliary POC of the face. This seems to predominate in adult women. Normochromic hardened papules are more commonly distributed over the face and scalp, but may also occur in the torso, buttocks, and extremities. There is a debate over whether this should be classified as a primary form, since a significant portion of cases are associated with a history of acne and it is possible that the ossification represents the end stages of a chronic inflammatory process. Thus, it may be best classified as a form of secondary ossification in a miliary pattern. Still, many patients are reported to develop the lesions without any identified triggering factor [7, 8]. With respect to the present case, the authors did not consider the findings to be consistent with miliary or plate-like forms.

A report published in 2006 by Ayaviri et al. described a female patient, 20 years old, with an approximately

4-year history of nodular coalescent lesions in the frontal hairline, extending from the forehead to the scalp, with no known trigger. The histopathology was compatible with osteoma cutis. The authors concluded that the case was an isolated form of POC. This subtype, along with HOP, and Albright hereditary osteodystrophy, would be related to changes in the GNAS (guanine nucleotide-binding protein) gene complex [9].

The pattern of the evolution of the injury and topography resembles that of the patient presented herein, with the small exception that the lesions were grouped, but did not coalesce. Another peculiar aspect of the case is the linear distribution. Orlow et al. published a case report of a 25-year-old woman with a 5-year history of a hardened plaque on the scalp, also linear. The authors diagnosed a primary plate-like osteoma [10]. Again, we would like to point out that our patient did not present with plaque, but rather with multiple papules that were distributed in a linear pattern, differing from other case reports.

There were insufficient lesions to consider this as a disseminated case of POC and there were no manifestations that might fit into the classification of Albright hereditary osteodystrophy (**Table 1**).

Histopathology is the gold standard for diagnosis, and the findings are unequivocal: well circumscribed formations and aggregate nodular or irregular shapes of lamellar bone formation in dermis [6, 7, 11]. In our literature review, we were unable to find any articles that addressed the dermatoscopic aspects of osteoma cutis. However, we believe that the most important thing is to rule out other diagnoses.

POCs do not pose any risk of neoplastic transformation and their limited forms generally do not undergo significant growth. Therefore, the recommended treatment should take into account the patient's wishes regarding the cosmetic outcome. Among the options, surgery (simple excision), combined or uncombined topical retinoids, and ablative lasers are the most commonly recommended treatments [3, 7].

Conclusion

POCs are rare; however, they should be understood by dermatologists. The classification of POC is not

Table 1. Evaluation of the patient with cutaneous ossification.

Mechanism of bone formation	Endochondral			Intramembranous			
Clinical evolution	Progressive	Progressive	Limited				
Associated manifestations	Halux malformation	Without associated manifestations	Pseudohypoparathyroidism / Pseudo-pseudohypoparathyroidism / Obesity / Brachydactyly / Short stature	Without associated manifestations			
	Progressive Ossifying Fibrodysplasia	Progressive Osseous Heteroplasia	Albright Hereditary Osteodystrophy	Plate-like Osteoma Cutis	Miliary Osteoma Cutis	Isolated Osteoma Cutis	Disseminated Osteoma Cutis

Adapted from Fairley JA in: Bolognia JL, Jorizzo JL, Schaffer JV. *Dermatology*. 3rd ed. Philadelphia, PA: Elsevier Saunders; 2012. 729-36.

straightforward, but some fundamental observations can serve as guides: 1) primary or secondary; 2) progressive or restricted process; 3) clinical expression and associated manifestations. With this report, we seek to draw attention to this entity, in addition to presenting a satisfactory therapeutic result.

References

1. Fairley JA in: Bolognia JL, Jorizzo JL, Schaffer JV. *Dermatology*. 3rd Ed. Philadelphia, PA: Elsevier Saunders; 2012. 729-36.
2. Roth SI, Stowell RE, Helwig EB. Cutaneous ossification: report of 120 cases and review of the literature. *Arch Pathol* 1963;76:44-54. [PMID: 13975332]
3. Talsania N, Jolliffe V, O'Toole EA, Cerio R. Plate-like osteoma cutis. *J Am Acad Dermatol*. 2011; 64(3): 613-5. [PMID:21315967]
4. Sanmartín O, Alegre V, Martínez-Aparicio A, Botella-Estrada R, Aliaga A. Congenital plate-like osteoma cutis: case report and review of the literature. *Pediatr Dermatol*. 1993; 10(2): 182-6. [PMID:8346117]
5. Montor AB, Burgdorf WH, Sheward S. Plate-like cutaneous osteoma. *J Am Acad Dermatol*. 1987; 16(2 Pt 2): 481-4. [PMID:3102570]
6. Orme CM, Hale CS, Meehan SA, Long W. Plate-like osteoma cutis. *Dermatol Online J*. 2014; 20(12). [PMID:25526332]
7. Bouraoui S, Mlika M, Kort R, Cherif F, Lahmar A, Sabeh M. Miliary osteoma cutis of the face. *J Dermatol Case Rep*. 2011; 5(4): 77-81. [PMID:22408708]
8. Ayaviri NA, Nahas FX, Barbosa MV, Farah AB, de Arimatéia Mendes J, Ferreira LM. Isolated primary osteoma cutis of the head: Case report. *Can J Plas Surg*. 2006; 14(1): 33-6. [PMID:19554229]
9. Martin J, Tucker M, Browning JC. Infantile osteoma cutis as a presentation of GNAS mutation. *Pediatr Dermatol*. 2012; 29(4): 483-4. [PMID:22612068]
10. Orlow T, Shawaf AZ. Plate-like cutaneous osteoma on the scalp. *Dermatol Online J*. 2006; 12(4): 17.
11. Orlow SJ, Watsky KL, Bolognia JL. Skin and bones. II. *J Am Acad Dermatol*. 1991; 25(3):447-62 [PMID:1918486]