

Multiple asymptomatic hard papules on cheeks in an elderly woman

A 55-year-old woman, recently diagnosed with dermatomyositis, presented with multiple, tiny, firm papular lesions on cheeks for many years. These lesions were asymptomatic with no history of ulceration or discharge. Past history was notable for the presence of severe facial acne vulgaris during adolescence and early adulthood. On examination, there were multiple small round to irregular-shaped skin-colored, hard, monomorphic papules on both cheeks [Figure 1]. There were no similar lesions elsewhere. Patient



Figure 1: Multiple, round to irregularly shaped skin colored, monomorphic papules on cheek

had characteristic cutaneous manifestations of dermatomyositis -- V sign, shawl sign, heliotrope rash, Gottron's sign and holster sign) with proximal muscle weakness.

Histopathology from the papule showed fibrocollagenous tissue with round to irregular, large homogeneous amorphous basophilic staining deposits in the deep dermis and subcutis. The dermis also showed spicules of crescentic eosinophilic material housing cells held within small lacunae, associated with mature adipose tissue. [Figures 2-4].

Question

What is your diagnosis?

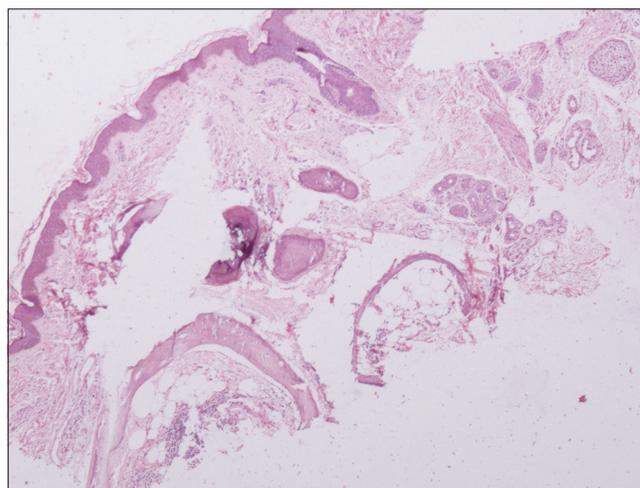


Figure 2: Round to irregular, large homogeneous amorphous basophilic staining deposits in the deep dermis and subcutis and spicules of crescentic eosinophilic material (H and E, 10x)

Access this article online	
Quick Response Code:	Website: www.ijdvl.com
	DOI: 10.4103/ijdvl.IJDVL_432_16

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Chander R, Garg T, Sanke S, Agarwal K, Chhikara A. Multiple asymptomatic hard papules on cheeks in an elderly woman. *Indian J Dermatol Venereol Leprol* 2017;83:513-5.

Received: June, 2016. **Accepted:** September, 2016.

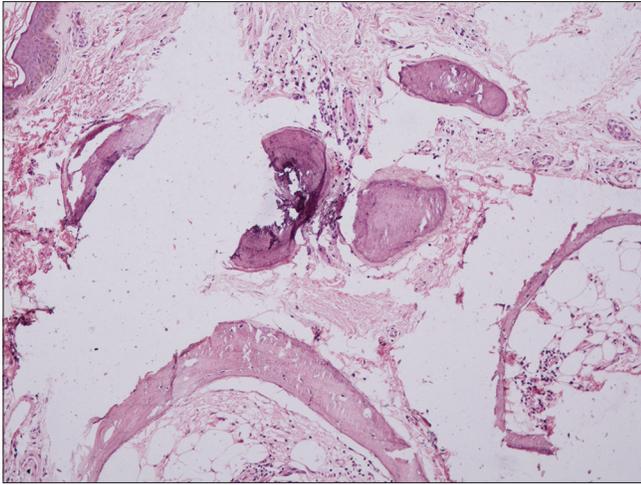


Figure 3: Round to irregular, large homogeneous amorphous basophilic staining deposits in the deep dermis and subcutis and spicules of crescentic eosinophilic material (H and E, $\times 100$)

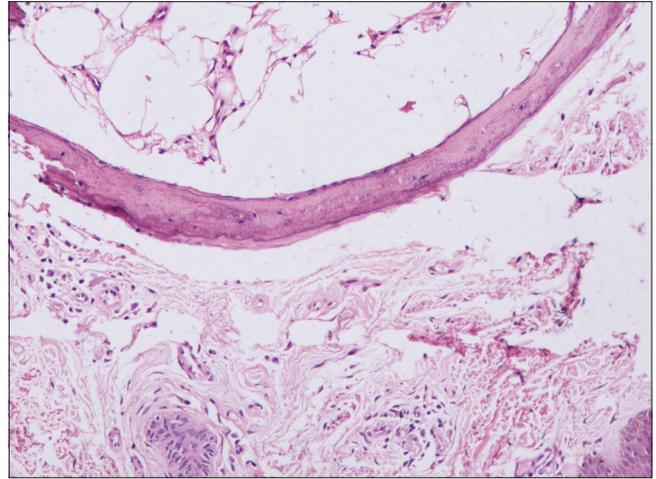


Figure 4: Crescentic eosinophilic material housing cells held within small lacunae, associated with mature adipose tissue (H and E, $\times 400$)

Answer

Diagnosis: Multiple miliary osteoma cutis of the face.

Review

Multiple miliary osteoma cutis of the face is a rare cutaneous disorder characterized by benign extra-skeletal bone formation in the dermis or subcutis, first described by Wilekens in 1858.¹ A review of literature yielded only 51 cases reported so far. The disease predominantly affects middle aged women and presents with skin colored hard papules and nodules resembling milia, predominantly on face whereas in men it may affect the scalp and chest.² Osteoma cutis can be of two types: Primary or secondary. Primary osteoma cutis is the idiopathic formation of bone. It may be an isolated finding or may be associated with Albright's hereditary osteodystrophy (pseudo-hypoparathyroidism or pseudo-pseudohypoparathyroidism). If there is no association, osteoma cutis can be classified into four types: Miliary osteoma cutis of face, isolated osteoma, widespread osteoma and congenital plate-like osteoma.³ Secondary osteoma cutis accounts for 80% of the cases, and arises as a consequence of varied previous inflammatory dermatoses (including nevi, acne vulgaris, scleroderma, pilomatricoma, dermatomyositis, trauma and neoplasms) or impaired calcium-phosphate metabolism.⁴ Though miliary osteoma cutis can be primary, it occurs more commonly secondary to precedent inflammatory processes, most commonly severe facial acne vulgaris.

Our patient also gave a past history of severe nodulocystic acne in her teenage years. Although osteoma cutis has been described in long-standing dermatomyositis, we believe that this particular presentation on the cheeks, in our patient, was unrelated to dermatomyositis as her connective tissue disease was of a very recent onset.

Various theories have been proposed for the pathogenesis of osteoma cutis. It is believed that severe inflammation can cause activation of mesenchymal stem cells into a matrix which calcifies itself into bone. Another hypothesis is that embryonic mesenchymal cells, erroneously migrate to the dermis, differentiating into the osteogenic lineage.⁵ Other authors have speculated about the skin fibroblasts being able to differentiate into osteoblast cells.

Histopathologically, osteoma cutis lesions may demonstrate calcification, lacunae, lamellae and/or bone marrow in the dermis or subcutis. Osteocytes and osteoblasts are embedded within the bone while osteoclasts and marrow elements may also be rarely seen. The lesions differ from calcinosis cutis in that they represent bone formation (dermal deposition of hydroxyapatite crystals)

compared to calcinosis cutis which demonstrates deposition of only calcium salts. A differential diagnosis of miliary osteoma is benign cartilaginous exostosis/osteochondroma, a benign tumor of mature hyaline cartilage and bone rarely seen in the maxillofacial region. It can be easily differentiated as clinically they lead to facial asymmetry and malocclusion while histologically they show chondrocytes of the cartilaginous cap arranged in clusters parallel to lacunar spaces.

Miliary osteoma cutis has no potential for malignant transformation. Treatment is directed to removal of existing lesions and prevention of the appearance of new lesions. Preventive measures including disodium etidronate have led to minimal improvement. The therapeutic treatment options include topical retinoid acid (0.025%-0.1%), curettage, surgical excision, needle microincision-extirpation, dermabrasion, erbium-yttrium aluminum garnet and carbon dioxide laser.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

**Ram Chander, Taru Garg, Sarita Sanke,
Kiran Agarwal¹, Aruna Chhikara¹**

Departments of Dermatology and STD and ¹Pathology, Lady Hardinge Medical College and Associated Hospitals, New Delhi, India

Correspondence: Dr. Sarita Sanke,
Department of Dermatology and STD, Lady Hardinge Medical College, Shaheed Bhagat Singh Marg, New Delhi - 110 001, India.
E-mail: sanke sarita@gmail.com

References

1. Essing M. Osteoma cutis of the forehead. HNO 1985;33:548-50.
2. Gfesser M, Worret WJ, Hein R, Ring J. Multiple primary osteoma cutis. Arch Dermatol 1998;134:641-3.
3. Mast AM, Hansen R. Multiple papules on the elbows. Congenital osteoma cutis. Arch Dermatol 1997;133:777-80.
4. Altman JF, Nehal KS, Busam KJ, Halpern AC. Treatment of primary miliary osteoma cutis with incision, curettage, and primary closure. J Am Acad Dermatol 2001;44:96-9.
5. Myllylä RM, Haapasaari KM, Palatsi R, Germain-Lee EL, Hägg PM, Ignatius J, *et al*. Multiple miliary osteoma cutis is a distinct disease entity: Four case reports and review of the literature. Br J Dermatol 2011;164:544-52.