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# Milia-like idiopathic calcinosis cutis in an adult without Down's syndrome

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## Abstract

**Background:** Milia-like idiopathic calcinosis cutis is a rare entity. Only 19 cases have been reported so far, the majority of them developed in children with Down's syndrome. The mean age of the patients is 10.3 years, with a sex ratio of nine girls to ten boys. Hands are most commonly affected.

**Main Observation:** We report a case of a 69-year-old, otherwise healthy woman, who developed milia-like idiopathic calcinosis cutis on her forehead.

**Conclusion:** To our knowledge, we report the seventh case occurring in a patient without Down's syndrome, and the first case occurring in an elderly person.

## Background

Milia-like idiopathic calcinosis cutis (MICC) is a rare benign disorder first described by Sano *et al* in 1978[1], and termed MICC by Smith in 1989.[2] 19 cases have been reported so far, the majority of them develop in children with Down's syndrome. We report MICC occurring on the face in an elderly person without Down's syndrome.

## Case Report

A 69-year-old healthy woman was referred to our clinic because of numerous skin-colored, firm papules, 1-3 mm in diameter on her face, mainly distributed on the forehead (Fig. 1). The papules had been present for 9 years and were asymptomatic. She denied any previous trauma at the site of the lesions.

Histologic examination of a biopsy specimen from a lesion of her face and revision of histologic examination

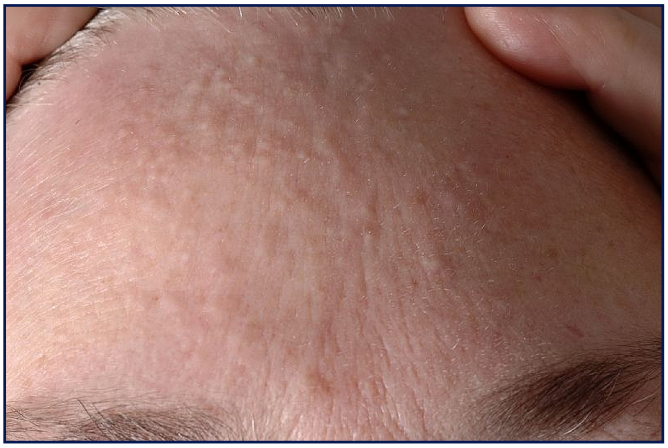

**Figure 1**

*Clinical presentation of our milia-like idiopathic calcinosis cutis.*

done 8 years ago in another clinic, showed focus of calcium in superficial dermis, without signs of syringoma or inflammatory infiltrate (Fig. 2). Laboratory findings, including serum calcium, phosphorus levels, and creatinine were within normal range.

On the basis of these pathologic findings and clinical features, the patient was diagnosed with milia-like idiopathic calcinosis cutis.

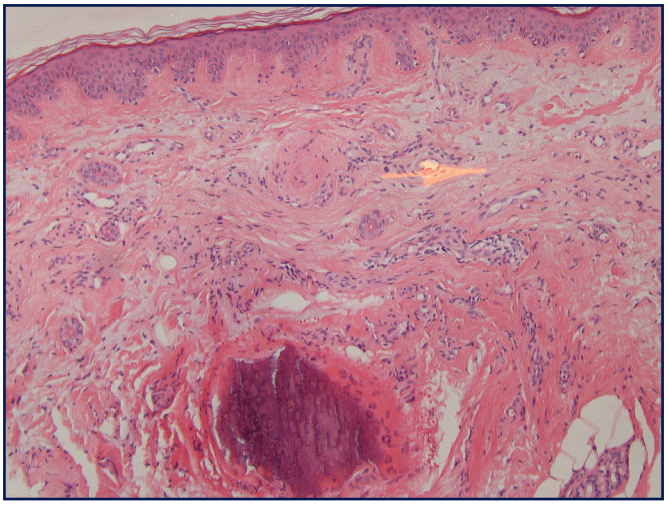

**Figure 2**

*Histology shows focus of calcium within the superficial dermis with HE staining.*

## Discussion

Milia-like idiopathic calcinosis cutis is a rare dermatologic disorder that was first described in 1978.[1] Clinically the lesions appear as smooth, firm, skin-colored papules resembling milia. They are sometimes surrounded by erythema.[6,10]

Some papules have a central crust, corresponding to a transepidermal elimination of calcinosis.[6,7,15] The disease affects primarily the hands and feet, but may occur on other parts of the body. Involvement of the face has rarely been reported.[1,6] Most cases of the disease have been reported in children with Down's syndrome and/or syringoma.[1-3, 6-12, 15,16]

In our case, adnexal cysts, adnexal tumors, verrucae planae were additional diagnostic considerations at the time of biopsy. To our surprise, histologic examination showed focus of calcium, without signs of milia, syringoma or warts. Laboratory findings, including serum calcium, phosphorus levels, and creatinine were within normal range, ruling out the diagnosis of secondary or metastatic calcinosis (see Table 1 for the general differential diagnosis of MICC[19]). The patient denied any previous trauma at the site of the lesions. On the basis of these findings and clinical features, the patient was diagnosed with milia-like idiopathic calcinosis cutis. To our knowledge this is the first report of an elderly person with MICC.

**Table 1**

*Disorders of cutaneous calcification.*

| |
|---|
| **Dystrophic** |
| Autoimmune connective tissue diseases, e.g. dermatomyositis, CREST |
| Cutaneous tumors, e.g. pilomatricomas |
| Infections, e.g. parasitic |
| Trauma |
| Panniculitis |
| Genetic disorders, e.g. pseudoxanthoma elasticum, Ehlers-Danlos syndrome |
| **Metastatic** |
| Chronic renal failure |
| Calciphylaxis |
| Benign nodular calcification |
| Hypervitaminosis D |
| Milk-alkali syndrome |
| Sarcoidosis |
| Hyperparathyroidism |
| Neoplasms (e.g. multiple myeloma, leukemia, lymphoma) |
| **Idiopathic** |
| Idiopathic calcified nodules of the scrotum |
| Subepidermal calcified nodule |
| Tumoral calcinosis |
| Milia-like calcinosis |
| **Iatrogenic** |
| Extravasation of intravenous solutions containing calcium or phosphate |
| Application of calcium-containing electrode paste for EMGs and EEGs |
| Application of calcium alginate dressings to denuded skin |
| Liver transplantation |

The cause of MICC is unknown. One hypothesis is that increased sweat calcium content causes calcium precipitation in the acrosyringium.[4] Alternatively, milialike calcinosis cutis may result from calcium deposition in persistently inflamed microepidermal cysts.[11] Some authors also hypothesize that lesions may result from localized persistent trauma.[13]

To our knowledge 19 cases of MICC have been reported thus far,[1-12,14-18] and only 6 of these cases were not associated with Down's syndrome.[4,11,14,17,18] The mean age of the patients is 10.3 years, with a sex ratio of nine girls to ten boys. In Table 2 characteristics of all reported patients with MICC are summarized.

Our case is unusual because the patient did not have Down's syndrome, the skin lesions were localized on the face and because MICC is unique in elderly persons. Clinicians should be aware of this rare benign disorder.

**Table 2**

*Characteristics of Patients with MICC.*

| | Reference | Sex/Age | DS | Ca2+/P levels | Perforation | Disease duration | Location | Syringoma | CA |
|---|---|---|---|---|---|---|---|---|---|
| (1) | Sano *et al.* | M/19 | + | ? | ? | ? | Hands | PS | - |
| (4) | Eng *et al.* | M/10 | - | N | + | 1 year | Thighs, scrotum foreskin | - | - |
| | | F/11 | - | Refusal | + | 5 years | Pubis | - | - |
| (5) | Shibuya *et al.* | M/12 | + | N | + | 1.5 years | Palms, soles | - | - |
| (2) | Smith *et al.* | M/6 | + | N | + | 2 years | Hands, elbows, knees, legs, feet, face, wrists | - | - |
| (6) | Maroon *et al.* | M/12 | + | ? | + | 2 years | Hands, forearms, thighs, neck, face | PS/PLS | + |
| (7) | Kanzaki *et al.* | F/6 | + | N | + | 1 year | Hands, wrists, feet | PS/PLS | - |
| (8) | Schepis *et al.* | F/11 | + | N | + | 3 years | Hands, feet | PS | - |
| (9) | Sais *et al.* | F/7 | + | N | - | 1 year | Hands, feet | - | - |
| (10) | Schepis *et al.* | M/10 | + | N | - | ? | Hands, feet | PLS | - |
| | | M/11 | + | N | + | ? | Hands, toe | PLS | - |
| (11) | Lee *et al.* | F/21 | - | N | - | 1 month | Hand | - | - |
| (16) | Patrizi *et al.* | F/6 | + | N | + | 1 month | Hands | - | - |
| (15) | Delaporte *et al.* | F/15 | + | N | + | 1.5 year | Hands, wrists | - | - |
| (12) | Schepis *et al.* | M/11 | + | N | + | 1 year | Hands | - | - |
| (17) | Kim *et al.* | M/2 | - | N | - | 1 year | Sole | - | - |
| (3) | Kotsuji *et al.* | F/7 | + | N | + | 2 years | Hands | - | - |
| (14) | Roth *et al.* | M/14 | - | N | + | 2 years | Hands, elbows, lips, knees | - | - |
| (18) | Mehta *et al.* | F/5 | - | N | - | 1.5 year | Chin, neck, trunk, intertriginous areas, extremities | - | - |
| | Present case | F/69 | - | N | - | 9 years | Face | - | - |

CA, calcified acrosyringium; PLS, perilesional syringoma; PS, palpebral syringoma; TE, transepidermal elimination; DS, Down's syndrome

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