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The Clinical Picture

Multiple skin nodules in a 64-year-old woman

A 64-YEAR-OLD WOMAN has widespread hard subcutaneous nodules (FIGURE 1) which she says developed 18 years ago, about 10 years after the onset of proximal muscle weakness and rash. The nodules are mainly over bony prominences and tendons, but she also has them on her neck, trunk, arms, abdomen, hips, and buttocks.

Which of the following is not associated with this woman's clinical presentation?

- ❑ Rheumatoid arthritis
- ❑ Juvenile-onset dermatomyositis
- ❑ Adult-onset dermatomyositis
- ❑ Systemic sclerosis

Based on the clinical presentation, this woman has calcinosis cutis, which is associated with all the above conditions except rheumatoid arthritis.

Calcinosis cutis is most often a complication of juvenile-onset dermatomyositis and is seen in up to 40% of these young patients. It is much less common in adult-onset dermatomyositis. Calcinosis cutis is characterized by hard nodules under subcutaneous tissues, as well as in intracutaneous, fascial, and intramuscular tissue. They tend to occur over bony prominences and at sites of repeated microtrauma, such as the elbows, knees, buttocks, and flexor surfaces of the fingers.

Calcinosis cutis is also found in patients with systemic sclerosis, with a similar presentation. There are a few descriptive reports of calcinosis cutis in patients with systemic lupus erythematosus, but this is often with overlap syndromes and is not a common feature.

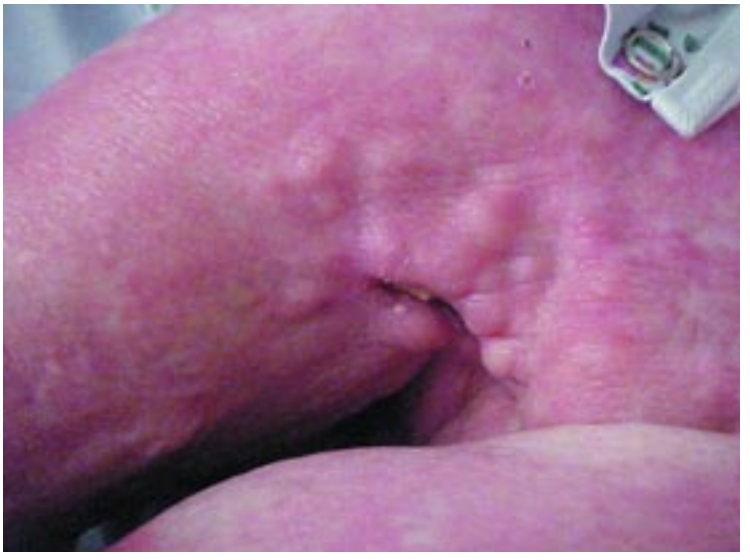

FIGURE 1.

Cause

The cause of calcinosis cutis is not known, but the underlying process is thought to involve the release of calcium from mitochondria in myocytes that have been damaged. The material in these nodules is most commonly calcium hydroxyapatite. When calcinosis appears, the muscle disease may be well controlled, and the calcinosis is not necessarily a marker of active myositis.

Complications

Complications associated with calcinosis cutis include ulceration of the skin overlying the nodules, drainage of material from open lesions, and dystrophy or liquefaction of material under the nodule, with marked inflammation or superinfection. Joint contractures secondary to intramuscular and fascial involvement can occur and interfere with mobility.

Diagnosis and treatment

This is a clinical diagnosis and does not require biopsy.

Once calcinosis develops it is difficult to treat, and spontaneous regression is uncommon. We have as yet no evidence from controlled studies to support the effectiveness of any therapy. However, we do know that early and aggressive treatment of myositis may prevent calcinosis in children. Studies have shown that early immunosuppression, including intermittent high-dose intravenous-pulse methylprednisolone, may decrease the occurrence of calcinosis.

We have anecdotal reports of treatment benefit with calcium channel blockers, bisphos-phonates, low-dose warfarin, aluminum hydroxide, and probenecid, but studies have not been done in large groups of patients.

Surgical excision may be helpful, but deep wounds may heal slowly or incompletely, and excised nodules tend to recur.

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SUGGESTED READING

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CME ANSWERS

Answers to the credit test on page 727 of this issue

1 **B** 2 **E** 3 **B** 4 **A** 5 **B** 6 **E** 7 **E** 8 **E** 9 **E**
10 **C** 11 **D** 12 **A** 13 **B** 14 **B** 15 **A**