CASE OF THE MONTH

A child with skin nodules and extensive soft tissue calcification

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Case history

A 9-year-old male presented to the dermatology clinic complaining of hard cutaneous nodules all over his body. The patient also had a chronic history of proximal muscle weakness of approximately 5 years' duration. On examination some of the nodules were whitish in colour and a white-coloured liquid discharge was seen exuding from the left calf skin surface. Multiple plaques were identified over the extensor surface of the hands, especially over the

knuckles. There was no family history of a similar disease. The patient had two older brothers who were both healthy. Radiographs of the upper and lower extremities were requested. A CT of the lower extremities was also performed after a few days.

What do you notice in Figures 1 and 2?

Can you see a pattern in Figure 3?

What additional findings can be observed in Figures 4–6?

What is your diagnosis?

Figure 1. Photograph of the patient showing the cutaneous nodules in the neck region.

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Figure 2. Photograph of both hands showing plaque-like lesions over the extensor aspects of hands and the knuckles. A few cutaneous nodules are also seen.

Figure 3. Radiograph of both thighs showing a typical sheet-like pattern of soft tissue calcification along the muscular planes and subcutaneous fascia.

Diagnosis

The distinctive pattern of diffuse soft tissue calcification in subcutaneous and muscular planes occurs in calcinosis universalis. The condition may be idiopathic or due to a connective tissue disorder, for example systemic lupus erythematosus, scleroderma and juvenile dermatomyositis (JDM). Biochemical tests can help to narrow the differential diagnoses.

In our patient, muscle enzyme levels were found to be markedly elevated with serum creatine phosphokinase and aldolase levels at 613 U l^{-1} and 29 U l^{-1} , respectively (the normal range for creatine phosphokinase is

Figure 4. Axial CT image of the bilateral thigh region confirming the extensive calcification within the muscle planes. Additionally, smaller nodular areas of calcification are also identified in the subcutaneous tissues.

Figures 5. Sagittal volume-rendered CT images of the left thigh showing the large intramuscular pockets of liquefied calcium with layering of calcium. One of the intramuscular pockets of liquid calcium is actively oozing calcium through the skin surface.

$60\text{--}170 \text{ U l}^{-1}$ and for aldolase is $0\text{--}6 \text{ U l}^{-1}$). The anti-nuclear antibody test was negative; serum calcium and phosphorus levels were within the normal range. The ultrasound of the abdomen was also normal. The patient history did not reveal any complaints characteristic of Raynaud's phenomenon or of scleroderma. On review of the dermatology, the plaque-like lesions on the extensor aspect of the hands were diagnosed as Gottron's papules. Correlation of the radiological findings, clinical evaluation and biochemistry in our patient suggested that JDM was the underlying disease and ruled out other diseases such as systemic lupus erythematosus and scleroderma. Inherited disorders, such as Ehlers-Danlos syndrome and pseudoxanthoma elasticum, in which the dystrophic soft tissue calcifications may resemble calcinosis universalis, were also not considered because features such as hypermobility of skin or laxity of joints, which are present in these disorders, were not seen in our patient.

Discussion

Calcinosis universalis refers to dystrophic calcification occurring in a diffuse sheet-like manner in the fascial

Figure 6. Axial volume-rendered CT images of the left thigh showing the large intramuscular pockets of liquefied calcium with layering of calcium. One of the intramuscular pockets of liquid calcium is actively oozing calcium through the skin surface.

planes, subcutaneous tissues and muscles. It is commonly seen in association with connective tissue disease, *e.g.* polymyositis or dermatomyositis. JDM is the childhood form of dermatomyositis and is a rare multisystem autoimmune disease of unknown aetiology, characterised by inflammatory myopathy. The incidence is two to three cases per million children per year [1]. The presence of skin rashes such as Gottron's papule and heliotrope rash is considered characteristic of JDM. Calcinosis, the hallmark of JDM, is seen in 30% of patients, with a prevalence range of 10–50% [2, 3]. Calcification is dystrophic and serum calcium and phosphorus levels are normal. Four different patterns of calcifications have been described in patients with JDM [3, 4]:

1. superficial plaques or nodules in skin and subcutaneous tissues
2. deep nodular deposits extending to muscles
3. deposits along fascial planes of muscles and tendons
4. hard deposits covering the entire body surface.

The incidence for patterns 1–4 is 33%, 20%, 16% and 10%, respectively [5]. In 22% of patients a mixed pattern is seen. The sites most frequently involved are the elbows, knees, digits and extremities. Commonly, the time period for the appearance of calcinosis after the onset of the illness is 1–3 years, but it can be as long as 20 years. Imaging with conventional radiography can identify the pattern of soft tissue calcification and aid the diagnosis. CT is more sensitive and specific than conventional radiography for deeper soft tissue calcifications [6]. CT also has the advantage of depicting ossifying lesions and the plane of calcification. MRI sequences, for example short tau inversion-recovery, may be useful in picking up oedema or

inflammation of the skin, subcutaneous tissues or fascia prior to the development of calcinosis.

Risk factors for the development of calcinosis are poorly understood, but it has been associated with a delay in diagnosis and the initiation of therapy. The natural course of calcinosis is variable and unpredictable, and a number of sequelae may be seen. Spontaneous extrusion or reabsorption may sometimes lead to regression. Improvement is more likely in those patients with inactive disease, those who engage in more physical activity, those with superficial plaques or nodules rather than deep deposits and those treated with an aggressive therapeutic regimen [3]. Radiologists can play an important role by aiding the diagnosis of this rare disease and paving the way for early initiation of therapy and, possibly, a favourable outcome. Other causes of soft tissue calcification include metastatic calcification (due to hyper- or hypoparathyroidism, paraneoplastic hypercalcaemia, excessive vitamin D or milk alkali syndrome), dystrophic calcifications in inherited disorders (Ehlers–Danlos or pseudoxanthoma elasticum), soft tissue tumour, gout, parasitic infections or idiopathic tumoural calcinosis [1]. Correlation of the typical radiological patterns of soft tissue calcification with biochemical tests (elevated muscle enzyme levels with normal serum calcium and phosphorus levels) and the presence of skin lesions help in achieving the correct diagnosis of JDM and in excluding other conditions.

Conclusion

Calcinosis universalis has unique and typical radiological findings, and the possibility of a rare disorder such as JDM should be considered as the underlying cause if the patient is a child and the biochemical tests and clinical picture are also suggestive.

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