

Massive calcinosis and severe osteoporosis in paediatric-onset overlap syndrome

Sirs,

Subcutaneous calcinosis may be a common long-term complication of juvenile dermatomyositis, systemic sclerosis and related conditions (1); subcutaneous calcifications are usually localised around knees and elbows. By contrast, widespread calcinosis forming a tumoral-mass (tumoral calcinosis) is much more rare in paediatric patients (2).

Herein we report the case of a 14-year-old boy affected with overlap syndrome who developed a rapidly progressive and widespread calcinosis of hypodermal and muscular fascia leading to severe movement limitation associated with severe osteoporosis.

The patient, born from unrelated healthy parents, was well up 10 years of age, when he developed polyarthritis of the hips, metacarpal and inter-phalangeal joints associated with an erythematous rash on knees and elbows. A diagnosis of psoriatic arthritis was made. Since oral steroids were only partially effective, methotrexate and etanercept were started with benefit.

One year later, the child developed severe Raynaud's phenomenon with ulcerations of the fingers of the hands and feet with ischaemia.

At the age of 11 he developed an important symmetrical weakness of the proximal upper and lower limb musculature, leading to difficulties in walking; malar rash and skin ulceration at fingers and elbows were present with normal CPK plasma concentration. The diagnosis of juvenile dermatomyositis was suggested and high dose prednisone was started with a good clinical response; during steroid tapering muscular straightness remained good; however, the skin lesions worsened and eschar formation occurred at the distal phalanx of the second finger of the right hand. One year later he developed skin induration at the lumbar spine and buttocks with the clinical feature of calcinosis.

On admission to our Institute, the patient showed a limited range of movement with an inability to walk for more than 10 m and muscular hypotrophy. Skin was normally elastic with lineal discromic areas along the limbs and the trunk. Severe acral vasculopathy with ulcerations of the fingers, acrocyanosis and onychodystrophy were also present. Nailfold capillaroscopy showed a scleroderma pattern with extensive avascular areas and neoangiogenic capillaries; no giant capillaries or haemorrhage were detected.

Fig. 1. Calcinosis of the lower limbs in a 14-year-old child with overlap syndrome.



Laboratory investigations displayed normal PCR and CPK plasma concentrations and increased ERS, LDH and ferritin (65 mm/h, 593 UI/L and 740 ng/dL respectively). Autoantibodies (ANA, anti-dsDNA, ENA and anti-Sc170) were absent. Serum and urinary calcium and phosphate were within normal ranges: moreover phosphate tubular reabsorption was normal. Upon closer inspection of the clinical and biohumoral data, the diagnosis of overlap-syndrome was pointed out.

The radiography of the lower limbs (Fig. 1) and hands showed a massive, widespread subcutaneous calcinosis, with involvement of the lower abdomen and paravertebral region. Gastro-oesophageal transit was normal. Chest CT showed a severe, widespread calcinosis of subcutaneous tissue associated with muscular hypotrophy. No signs of interstitial disease of lung were disclosed. MRI (STIR sequence) showed only a slight and diffuse heterogeneous alteration of signal within skeletal muscles associated with fascial involvement.

Since the radiological study was suggestive for a reduced bone density, DEXA as performed and showed an important osteoporosis (Z score -3.2).

This patient, affected with overlap syndrome, developed a massive, rapid onset calcinosis without detectable abnormalities of calcium/phosphate metabolism. It should be noted that calcinosis was present also in the paraspinal and abdominal areas, which are not usually affected in overlap syndrome. The massive calcinosis began

the pitoval symptom, severely limiting his physical activity: clearly, signs of inflammatory disease (*i.e.* myositis and arthritis) were less evident.

Although only anecdotal cases of massive calcinosis in patients with overlap syndrome have recently been reported, further investigations are needed to better define the underlying disease mechanisms of this unusual complication.

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