Calcinosis in juvenile dermatomyositis mimicking cold abscess

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ABSTRACT

We report a case of dystrophic calcification presenting as soft cystic swelling in a patient with juvenile dermatomyositis. A 15-year-old boy with lumbosacral cystic swelling, which was considered a cold abscess clinically, was evaluated for non-response to antitubercular therapy. The cystic swelling had liquefied calcium with a well circumscribed calcified wall on imaging, which was subsequently excised.

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INTRODUCTION

Juvenile dermatomyositis (JDM) is a multisystem disease in which mortality has declined considerably in recent years. However, morbidity related to the myopathy, calcinosis and long-term use of corticosteroids remains a major challenge.[1] Calcification is usually a late complication of dermatomyositis and often becomes recalcitrant to therapy. Different presentations including superficial plaques or nodules in the skin or subcutaneous tissue, deep linear deposits along fascial planes of the muscles, and extensive superficial, lacy reticular calcium deposits, have been described.[2] Calcinosis occurs most often in patients who have been treated inadequately and the treatment options once it occurs are limited with variable results.

THE CASE

A 15-year-old boy was diagnosed to have JDM at the age of 13 years on the basis of proximal muscle weakness, heliotrope rashes, raised creatinine phosphokinase (CPK) and muscle biopsy suggestive of myositis. He was treated with steroids and methotrexate with intermittent flares requiring pulse steroid therapy. He now presented with complaints of swelling over the right lower back, which was increasing for the past 5 months. There was no history of pain, restriction of movement, trauma, loss of appetite or fever. Examination showed a bilateral erythematous malar rash reaching up to the forehead and Gottron papules over proximal interphalangeal joints bilaterally. Systemic examination was non-contributory. A well-defined soft, cystic, 9×9 cm swelling was present over the right lower back just adjacent to the spine. The overlying skin was free but the swelling was not freely mobile and reduced slightly on bending forward. There was no redness, warmth or tenderness. During the present admission the boy was on low-dose prednisolone and did not have muscle weakness though his skin lesions were still present.

A cold abscess was considered in view of the location and a history of prolonged immunosuppression. The boy was started on antitubercular therapy after sending the aspirated fluid (milky white in colour) from the swelling for appropriate investigations. Investigations were negative for acid-fast bacilli on staining, growth on liquid media (MGIT) and bacterial culture. An X-ray of the spine did not suggest vertebral involvement. As there was no response to therapy and the swelling continued to increase in size, possibilities of drug-resistant tuberculosis, dystrophic calcification and malignancy were considered.

Investigations showed normal haemoglobin (13.7 g/dl), total and differential leucocyte counts (5800/cmm; neutrophils 48%, lymphocytes 36% and monocytes 10%) and platelet count (250 000/cmm). The erythrocyte sedimentation rate (5 mm in first hour) and C-reactive protein (4.8 g/dl) were not raised. Serum CPK level was mildly raised (268 IU/L) while transaminases were within normal limits with serum calcium of 8.3 mg/dl and serum phosphate of 5.1 mg/dl. Renal and liver function tests were within normal limits. A repeat aspiration of the swelling again revealed a milky white coloured fluid which showed protein: 4.6 g/dl, sugar: 152 mg/dl and calcium: 10 mg/dl. Cytopathology was negative for acid-fast bacilli, and showed foamy histiocytes with necrotic area consistent with degenerated muscle fibres.

X-ray of the spine showed streaks of calcification at the site of the swelling. A CT scan was done and showed sheet-like calcification at multiple levels along with a fluid collection with a calcified wall over the back measuring 8×4.4×1.9 cm with hyperdense layering at the bottom (Fig. 1). The vertebral bodies were normal. A diagnosis of dystrophic calcification was made and surgical excision of the cyst was planned. A search for calcinosis at other sites did not reveal any. Intraoperative findings were a milky fluid collection within a calcified cyst wall, which was adherent to the underlying spinal muscles. Pathological examination of the tissue showed a mass composed of flattened soft tissue measuring 10×7 cm with chalky white deposits over the inner surface. The postoperative period was uneventful and the boy was started on bisphosphonate for calcinosis.

DISCUSSION

JDM is a chronic inflammatory myositis with predominantly cutaneous findings. Delay in the diagnosis and treatment can lead to development of pathological calcification.[3] About 20%–40% patients develop dystrophic calcification.[4] While the trigger for dystrophic calcification is postulated to be inflamed muscle or skin, the exact precipitants remain unknown. The usual time for development of dystrophic calcification from onset of disease is 2–3 years and mostly occurs over the skin and superficial tissues, occasionally causing a sterile necrotic abscess.[5,6] Although dystrophic calcification in the form of calcinosis cutis is common, a large cystic lesion has been reported[7–10] with only three cases in children. De Castro *et al*.[7] reported collection of milk of calcium in the calf muscle in a 15-year-old child with overlap syndrome. Samson *et al*.[8] described the magnetic resonance imaging findings of milk of calcium in a 16-year-old girl with JDM. Similar imaging findings of milk of calcium was described in patients with myositis.[9,10]

Our patient had JDM, was on prolonged immunosuppression and developed a paravertebral fluctuant swelling. The differential diagnosis for such a presentation includes tubercular abscess, other infective collections, lymphangiectasia and calcinosis. A

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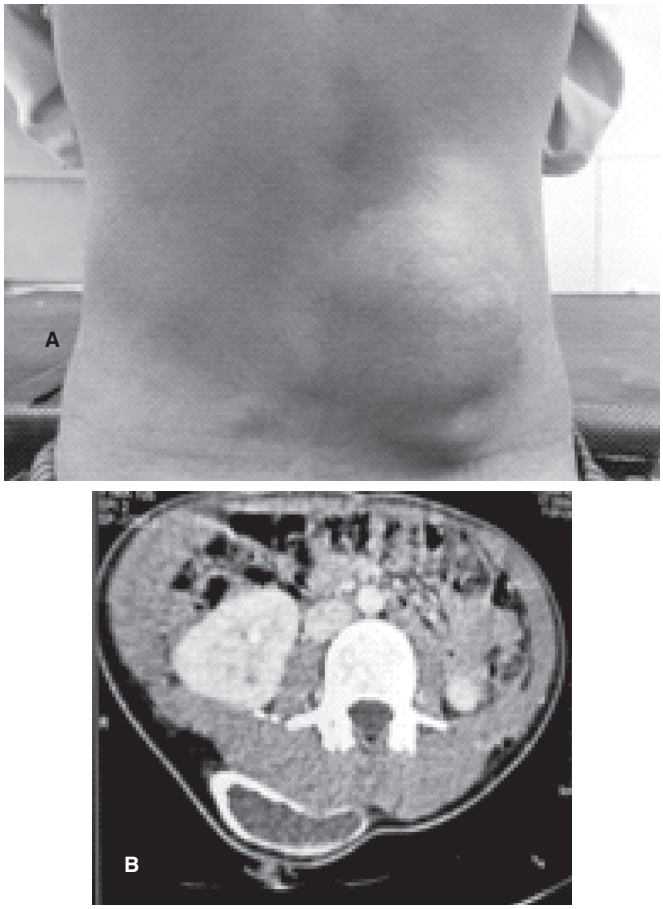

FIG 1A. Clinical photograph of back of the child showing a large swelling. B. CT scan of the trunk showing a cyst with calcification of the wall

paravertebral fluctuant swelling with milky white fluid in a child on prolonged immunosuppresants favoured diagnosis of tubercular abscess. However, there was no laboratory evidence for tuberculosis. A localized swelling with milky white fluid aspirated from a paravertebral swelling suggests lymphangiectasia. Laboratory findings including cytopathology did not suggest lymphangiectasia. Streaks of calcification at the site of swelling on the X-ray of the dorsolumbar spine and the CT scan showing sheet-like calcification at multiple levels along with a fluid collection with calcified wall favoured a diagnosis of dystrophic calcification.

Although most cases of subcutaneous calcinosis are treated with medical therapy, a localized swelling may require surgical removal. Different treatment options, including calcium channel blockers, warfarin and colchicine, have been tried but none is effective in the long term.[11] Currently, bisphosphonate is a promising drug for calcinosis of JDM with good response reported in some studies.[12,13] However, early treatment of the primary disease and avoiding disease flares is the mainstay of preventing calcinosis.

We have described a young boy with JDM and an unusual presentation of calcinosis. A cystic parietal swelling in patients with underlying myositis should be investigated for dystrophic calcification, which may need more aggressive treatment both for the primary disease and its complications.

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