INTRODUCTION

Calcinosis cutis refers to a group of disorders in which calcium deposits are being formed in the skin. Depending on the pathophysiologic mechanisms, calcinosis cutis has been classified as metastatic, dystrophic, idiopathic, or iatrogenic (1). In connective tissue diseases, calcinosis is mostly of the dystrophic type and it seems to be a localized process rather than an imbalance of calcium homeostasis (2).

Juvenile dermatomyositis (JDM) is a systemic, autoimmune inflammatory muscle disorder and a small vessel vasculopathy that affects children younger than 18 years, primarily the skin and the skeletal muscles (3). Calcinosis cutis occurs in 20-40% of patients with JDM (4). Some severe cases may be associated with advanced calcinosis but the exact mechanism that leads to their development and spreading remains unclear.

CASE REPORT

A female patient firstly admitted to the hospital at the age of 17 for skin rash, malaise and pain in peripheral joints and muscles. After evaluation a syndrome overlap with dominant atypical dermatomyositis (without muscle enzymes elevated) and systemic lupus erythematosus was suspected and treatment with prednisolone and choroquine initiated.

During the last 12 years, systemic corticosteroids and other immunosuppressive and immunomodulatory agents (methotrexate, cyclophosphamide, azathioprin, mycophenolat, colchicine), intravenous bisfosfonates, imunoglobulins and calcium antagonists were given in attempt to stop spreading and reduce a calcinosis.

After six months subcutaneous calcium deposits in form of multiple firm nodules appeared, firstly in gluteal region, than polytopically. The nodules were gradually enlarging, causing pain and exudation of chalky white material without a previous traumatic event. No metabolic disorders were observed; levels of PTH, phosphorus and calcium have always been in reference range; ANA and anti-dsDNA occasionally elevated.

During the last 12 years, systemic corticosteroids and other different immunosuppressive and immunomodulatory agents (methotrexate, cyclophosphamide, azathioprin, mycophenolat, colchicine), intravenous bisfosfonates, imunoglobulins and calcium antagonists were given in attempt to establish a better disease control, stop spreading and reduce a calcinosis along with tissue damage but without any significant improvement.

According to the clinical presentation and findings, early onset and a widespread calcinosis, a diagnosis of JDM is established.

Sedimentation rate remains elevated which, with a current health condition, despite the prolonged treatment, indicates that disease is in active phase. Also, the signs of incipient interstitial lung disease are notable.

Lastly, a treatment with abatacept, alternatively rituximab was recommended.

CONCLUSION

Although a number of drugs are often given for the treatment of calcinosis, the approach to management is still not established due to inconsistent responses. Revealing a patogenesis of calcium deposits would certainly bring a new insight to this and similares cases.

References