Pediatric Rheumatology

Poster presentation

Efficacy of thalidomide for a girl with inflammatory calicinosis, a severe complication of juvenile dermatomyosistis

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Background

Juvenile dermatomyositis (JDM) is a systemic connective tissue disease characterized by typical skin rash and chronic muscle inflammation of unknown etiology. Calcinosis is one of the severe complications of JDM. It occurs in up to 30% of patients and results in major disability.

Case report

A girl, 14 years of age, was diagnosed as having JDM when she was 4 years old after a few months of increasing lethargy, muscle pain, muscle weakness, and rash. Calcinosis was recognized 18 months after disease onset. During 3 months, clinical manifestations and abnormal laboratory findings were effectively treated with prednisolone. However, generalized calcinosis rapidly progressed with high fever, multiple skin/subcutaneous inflammatory lesions, and increased level of CRP. Methylprednisolone pulses, cyclophosphamide, cyclosporine, azathioprine, and magnesium hydroxide/aluminum hydroxide were applied but failed. Examination of subcutaneous calcium milk revealed remarkably elevated levels of IL-6, TNF-α, and IL-1β. Being encouraged by partial effectiveness with etanercept, thalidomide was started when she was 12 years old. Clinical manifestations were subsided, and inflammatory markers showed remarkable improvement. However, recent examination by whole body PET-CT over 15 months thalidomide treatment still demonstrated hot spots around subcutaneous calcified lesion.

Conclusion

It suggested that thalidomide for calcium milk around the generalized calcinosis in a JDM patient could be effective to improve inflammatory manifestations and patient's QOL. We speculated that subcutaneous pooling of calcium milk may be the cause of inflammation and subsequent calcinosis in JDM, and TNF blockade by thalidomide will be beneficial for inhibiting systemic spreading of inflammation.