



Use of Adalimumab in a Child with Juvenile Dermatomyositis and Calcinosis

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To the Editor: A 10-y-old girl diagnosed with juvenile dermatomyositis (JDM) was admitted to the rheumatology clinic again due to a new rash and muscle weakness. During the previous 3 y, she had continuous disease activity without any significant remission, and her calcinosis developed despite the rough treatment of glucocorticoids combined with multiple immunosuppressive agents (intravenous immunoglobulin, methotrexate, cyclosporine, thalidomide, colchicine, and mycophenolate-mofetil) and bisphosphonates. On physical examination, the girl had a faint heliotrope rash around the eyes, Gottron sign, proximal muscle weakness, and palpable subcutaneous nodules mainly at bilateral limbs and buttocks. Plain radiographs confirmed the existence of diffuse calcification. In addition, anti-nuclear matrix protein 2 (anti-NXP-2) was positive strongly, which is associated with calcinosis in JDM [1]. In view of the recalcitrant calcifications in refractory JDM and failure of these treatments, therapy with adalimumab, a fully humanized monoclonal anti-TNF- α drug, was initiated, apart from MTX, colchicine, and a low dose of glucocorticoid. Three months after initiation of adalimumab, a satisfactory clinical response was achieved with alleviation of her symptoms. After 1 y of treatment, the calcinosis had decreased as shown by the repeat radiograph.

Calcinosis, one of hallmark sequelae of JDM, can lead to pain, functional disability, joint contractions, muscle atrophy, skin ulcers, and secondary infections [2]. Although there is evidence in previous reports of the very poor efficacy of TNF inhibitors in myositis [3], including JDM and calcinosis, the clinical improvement in this case has demonstrated the potential for use of adalimumab in JDM

with calcinosis. Currently, multiple treatment approaches including anti-inflammatory medications, drugs that affect calcium or phosphate metabolism, and mechanical therapy have been attempted to target calcification in JDM. Nevertheless, the pathogenesis of calcinosis in JDM is still unknown and standardized therapy is still not found [4]. More attention is needed to get an improved understanding of the pathogenesis and develop a reproducibly efficacious therapy for JDM patients complicated with calcinosis.

Declarations

Conflict of Interest None.

References

1. Li J, Zhou Z. Calcinosis in juvenile dermatomyositis. *N Engl J Med.* 2019;381:e31.
2. Leung AKC, Lam JM, Alobaida S, Leong KF, Wong AHC. Juvenile dermatomyositis: advances in pathogenesis, assessment, and management. *Curr Pediatr Rev.* 2021;17:273–87.
3. Yang SH, Chang C, Lian ZX. Polymyositis and dermatomyositis - challenges in diagnosis and management. *J Transl Autoimmun.* 2019;2:100018.
4. Traineau H, Aggarwal R, Monfort JB, et al. Treatment of calcinosis cutis in systemic sclerosis and dermatomyositis: a review of the literature. *J Am Acad Dermatol.* 2020;82:317–25.

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