中文題目：馬凡氏症候群與反覆性雙側蜂窩性組織炎

英文題目：Recurrent Cellulitis of Bilateral Lower Legs in Marfan Syndrome

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Introduction: Bilateral cellulitis is unusual. Recurrent cellulitis or erysipelas of lower extremities with symmetric distribution is even rare. Herein, we present a case of recurrent bilateral lower leg lesions in a middle-aged woman with Marfan syndrome who was repeatedly hospitalized for antibiotic treatment.

Case Presentation: Marfan syndrome is an autosomal dominant connective tissue disorder caused by gene mutation of fibrillin-1. In Marfan syndrome, overactivity of fibroblasts leads to increased production and accumulation of hyaluronic acid in extracellular matrix, increasing extracellular osmolarity and causing lymphedema of soft tissues. After a complete course of parenteral antibiotic treatment, in addition to subsequent compression therapy, chronic lymphedema was greatly ameliorated, and no recurrence of cellulitis was observed in the following ten months.

Discussion: Chronic edema, especially lymphedema, is a major risk factor for recurrent cellulitis. Chronic lymphedema provides a medium for bacterial growth, altered lymphatic function and decreased lymphatic drainage which deteriorates the skin immunity. In Marfan's syndrome, fibrillin-1 (FBN-1) gene mutation increased the expression of transforming growth factor-β (TGF-β) in affected tissues. TGF-β stimulates fibroblasts and causes the hyaluronic acid deposition in the third space through cell free synthesis system. Hyaluronic acid increases extracellular osmolality and impairs lymphatic drainage that causes lymphedema of soft tissues. The mechanism is found in aorta and breast which causes aortic aneurysm progression and breast cellulitis.

Conclusion: This report highlights that Marfan syndrome with chronic lymphedema, although immunocompetent, is vulnerable to skin and soft tissue infections. Compression therapy may be beneficial to the patient for prevention of recurrent cellulitis.