Recurrent Erysipelas in Myotonic Dystrophy Type 1 with IgG Deficiency

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Dear Sir,

Dystrophia myotonica type 1 (DM1), the most common muscular dystrophy in adults, is a hereditary, autosomal dominant, multi-system disease, caused by a CTG repeat expansion in a non-coding region of the dystrophia myotonica protein kinase (*DMPK*) gene on chromosome 19q13.3 [1, 2]. DM1 is frequently associated with IgG deficiency [3], which is independent of the CTG repeat size [4]. Here we report a DM1 patient with IgG deficiency and recurrent erysipelas.

The patient is a 61-year-old HIV-negative female with a history of recurrent erysipelas, occurring 4 times during the last 5 years. The erysipelas developed twice in the face (2001 and 2002) and twice in the gluteal region (2003 and September 2005). Her relatives reported increasing cognitive impairment and social regression during the last 6 years. In February 2005 she was admitted because of gait disturbance from weakness of the distal lower limbs. Clinical neurological examination revealed ptosis, dysarthria, weakness for foot extension (M5–), wasting of the distal upper and lower limb muscles, clinical myotonia and absent Achilles tendon reflexes. Nerve conduction studies were normal but electromyography revealed myotonic discharges and myogenic motor unit architecture. Ophthalmological examination showed incipient cataract bilaterally. Blood chemical investigations revealed slightly elevated glutamate oxalate transaminase but nor-

mal creatine kinase. Clinical cardiological examination and echocardiography were normal but electrocardiography showed arteriovenous block I and left anterior hemiblock. Molecular genetic analysis revealed a CTG expansion of 130 repeats in the *DMPK* gene. The diagnosis of DM1 was established. After the last episode of gluteal erysipelas in September 2005, she underwent a long-term oral penicillin therapy for 6 months until February 2006.

In May 2006 she was re-admitted because of fever up to 39.3°C and somnolence for 3 days. The skin in the gluteal region presented with an extensive bilateral erysipelas (fig. 1). This was altogether the fifth episode of an erysipelas, the third in the gluteal region. The C-reactive protein was 11.6 mg/dl (normal: <0.6 mg/dl), α_1 -globulin 5.0% (normal: 1.5–4.0%), but the leucocyte count and coagulation parameters were normal. The total serum IgG was

Fig. 1. Widespread erysipela in the gluteal region 7 days after onset in May 2006 (fifth episode) in the described DM1 patient.

reduced to 547 mg/dl (normal: 700–1,600 mg/dl). IgG subtypes were not determined. Chest X-ray showed no pulmonary infiltration. Clinical neurological examination was unchanged compared to previous findings. Cerebrospinal fluid examination was normal. Electrocardiography showed a normocardiac sinus rhythm and missing R progression. Under parenteral treatment with penicillin G (10 million IU/day) and flucloxacillin (6 g/day), fever and C-reactive protein decreased and the vigilance improved.

Decreased IgG is a well-known, previously described feature of DM1 but usually remains asymptomatic [5, 6]. Recurrent infections are usually no clinical characteristic of DM1. Only in single DM1 patients was hypogammaglobulinaemia associated with an increased rate of infections [6]. On the contrary, it has been shown that the prevalence of chronic lower respiratory tract infections, recurrent herpes simplex infections and recurrent erysipelas was increased among 313 patients with hereditary serum IgG deficiency,

particularly those with IgG3 deficiency [7]. Whether recurrent erysipelas in the presented patient are attributable to hypogammaglobulinaemia or a mere arbitrary coincidence and why hypogammaglobulinaemia was particularly associated with recurrent erysipelas and not more common infections remain speculative.

This case suggests DM1 with hypogammaglobulinaemia to be prone to recurrent erysipelas. This is the first report of a patient with DM1, low serum IgG and recurrent erysipelas.

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