

Juvenile Dermatomyositis Associated to Familial Mediterranean Fever

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Abstract Familial Mediterranean Fever (FMF) is a rare hereditary auto-inflammatory disease that can be exceptionally associated with many other dys-immune disorders; the most reported associations were with systemic vasculitis, spondyloarthropathies, inflammatory bowel diseases, systemic lupus erythematosus, multiple sclerosis, and juvenile chronic arthritis. The association of FMF with primary inflammatory myopathy remains exceptional and unusual; it has only been noted once before with adult polymyositis. We report an original observation of FMF associated with juvenile dermatomyositis in an eight-year-old boy, which, to our knowledge, has not been reported previously.

Keywords: *familial mediterranean fever, juvenile dermatomyositis, dermatomyositis*

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1. Introduction

Familial Mediterranean Fever (FMF) is a rare hereditary auto-inflammatory disease [1,2], which is particularly common among Sephardic Jews, Turkish, Armenian, and Arab, where the prevalence may be in the order of 1-2/1000 [1]. It is currently recognized as a ubiquitous disease, although in most cases the notion of an ancestor from the Middle East or the Mediterranean region is found [1,3]. This disease is genetically determined, caused by a mutation in the MEFV gene on chromosome 16 (16p13.3), and its transmission is predominantly autosomal recessive [1,2,3].

The association of FMF with other dys-immune disorders remains exceptional; the most reported associations were with systemic vasculitis, spondyloarthropathies, inflammatory bowel diseases, systemic lupus erythematosus, multiple sclerosis, and juvenile chronic arthritis [1,4,5].

The association of FMF with primary inflammatory myopathy remains exceptional and unusual; it has only been noted once before with polymyositis [6].

We report an original case of FMF associated with juvenile dermatomyositis (JDM), which, to the best of our knowledge, has not been reported previously.

2. Case Report

An eight-year-old boy with known FMF presented to our department with a feverish eruption of the face and hands.

The diagnosis of FMF was made at the age of 6 in front of recurrent abdominal pain with bilateral pleurisy, and confirmed by the presence of the M694V homozygous mutation of the MEFV gene. He was treated with colchicine with a good response.

The somatic examination found a febrile child at 38°C, a heliotrope erythroedema of the face (Figure 1), Gottron's papules of the extension faces of the proximal and distal inter-phalangeal and metacarpo-phalangeal joints of both hands (Figure 2), and provoked diffuse myalgia without muscle deficit.

Biology showed a marked inflammatory syndrome with erythrocyte sedimentation rate at 68 mm/H1 and a C-reactive protein at 42 mg/l, and rhabdomyolysis with creatinine kinase at 320 IU/l and lactic dehydrogenase at 480 IU/l. Other basic bioassays were within normal limits: total blood count, creatinine, serum calcium, fast glycaemia, transaminases, lipid parameters, and thyroid hormones. Standard X-rays of the chest and hands were without abnormalities.

Antinuclear antibodies were positive at 1/320 with positive anti-Mi2 antibodies. The electromyogram revealed a myogenic syndrome of the four limbs without associated neurogenic signs. Skin-muscle biopsy confirmed the diagnosis of acute dermatomyositis and did not show signs of colchicine-induced myopathy. No systemic visceral involvement was found, and investigations for underlying cancer were negative.

Under systemic corticosteroids at a dose of 1mg/kg/day and hydroxychloroquine at a dose of 200 mg/day, the evolution was rapidly favorable with complete remission.



Figure 1. Heliotrope erythrodema of the face



Figure 2. Gottron's papules on both hands

3. Discussion

Juvenile dermatomyositis (JDM) is a primary inflammatory myopathy of unknown etiology occurring, by definition, in children under 16 years of age [7,8,9]. Its incidence is estimated at 0.3-0.4/100,000 children, and is the most common inflammatory myopathy of the child with a maximum incidence around the age of seven [7,8,9]. It can rarely associate with other autoimmune diseases, particularly systemic lupus erythematosus, systemic sclerosis, and primary Sjögren's syndrome [10]. His association with the FMF has not been reported before. Indeed, in the Özçakar ZB et al series of 600 children with FMF, of whom 77 had concomitant disease, no association with JDM was noted [4].

This association represents a real diagnostic challenge for clinicians, especially since localized or diffuse myositis may be a clinical manifestation of FMF [11,12], and may even be the only manifestation revealing it [13]. Similarly,

colchicine, a basic drug for the treatment of FMF can induce an iatrogenic neuromuscular pathology that can simulate polymyositis (polumyositis-like syndrome) [14].

The association of FMF with an authentic primary inflammatory myopathy has been reported only once in the world literature by Eguchi M et al [6]. It was the case of a 37-year-old Japanese woman with FMF associated to a polymyositis. The exact mechanism of this association is not yet well understood, but this association comforts once again the immune dysfunction induced by FMF [6].

4. Conclusion

The association of FMF with primary inflammatory myositis remains exceptional and unusual. It deserves to be known because it can represent real diagnostic and therapeutic challenges.

Our observation is, to the best of our knowledge, the first reporting the association of FMF to dermatomyositis. It is also distinguished by its juvenile character.

Conflicts of Interest

None.

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