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Case report

Improvement of nailfold capillary microangiopathy after immunosuppressant therapy in a child with clinically amyopathic juvenile dermatomyositis

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ABSTRACT

Clinically amyopathic dermatomyositis is an extremely rare entity in childhood and only 75 cases have been described in the literature. These patients present mild or no muscle involvement, while skin involvement is similar to that seen in classic dermatomyositis. Systemic inflammatory vasculopathy is a hallmark of juvenile dermatomyositis, especially affecting the microcirculation. Nailfold capillaroscopy is a non-invasive method that allows direct visualization of nailfold capillary loops around the proximal edge of the finger nails. The characteristic microangiopathic features observed at capillaroscopy in juvenile dermatomyositis are represented by decrease in the number of capillary loops (devascularization), associated with dilation and branching of capillary loops. Due to the dynamic character of juvenile dermatomyositis microangiopathy, several studies have described the usefulness of capillaroscopy in the evaluation of therapy response. The authors originally describe extensive nailfold capillaroscopy abnormalities in a four-year old child with clinically amyopathic dermatomyositis and its significant improvement after immunosuppressive treatment. Although nailfold capillaroscopy has never been systematically studied in clinically amyopathic dermatomyositis, the fact that its cutaneous involvement is indistinguishable from that seen in classic dermatomyositis suggests that nailfold capillaroscopy is likely to be useful as a prognostic tool and bear significant correlation with treatment. These findings suggest that nailfold capillaroscopy should be regarded as an important tool for monitoring clinically amyopathic dermatomyositis patients.

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Melhoria na microangiopatia capilar periungueal após terapia imunossupressora em uma criança com dermatomiosite juvenil clinicamente amiopática

Palavras-chave:
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RESUMO

A dermatomiosite clinicamente amiopática é uma entidade extremamente rara na infância e foram descritos apenas 75 casos na literatura. Esses pacientes apresentam envolvimento muscular leve ou ausente, enquanto o envolvimento da pele é semelhante ao observado na dermatomiosite clássica. A vasculopatia inflamatória sistêmica é uma característica da dermatomiosite juvenil que afeta especialmente a microcirculação. A capilaroscopia periungueal é um método não invasivo que possibilita a visualização direta das alças capilares das pregas ungueais em torno da borda proximal das unhas. As propriedades microangiopáticas características observadas na capilaroscopia na dermatomiosite juvenil são representadas pela diminuição na quantidade de alças capilares (desvascularização), associada à dilatação e ramificação das alças capilares. Em decorrência do caráter dinâmico da microangiopatia da dermatomiosite juvenil, vários estudos têm descrito a utilidade da capilaroscopia na avaliação da resposta terapêutica. Os autores descrevem originalmente extensas anormalidades na capilaroscopia periungueal em uma criança de quatro anos com dermatomiosite clinicamente amiopática e sua melhora significativa após terapia imunossupressora. Embora a capilaroscopia periungueal nunca tenha sido sistematicamente estudada na dermatomiosite clinicamente amiopática, o fato de que seu envolvimento cutâneo é indistinguível do observado na dermatomiosite clássica sugere que a capilaroscopia periungueal possa ser útil como uma ferramenta prognóstica e apresentar correlação significativa com o tratamento. Esses achados sugerem que a capilaroscopia periungueal deve ser considerada uma importante ferramenta para o monitoramento dos pacientes com dermatomiosite clinicamente amiopática.

Introduction

Juvenile dermatomyositis (JDM) is a rare disease that belongs to the group of idiopathic inflammatory myopathies.[1] Clinically amyopathic dermatomyositis (CADM) is an even rarer entity in pediatrics, with only 75 cases described in the literature.[2] CADM patients present mild or no muscle involvement and the cutaneous manifestations are indistinguishable from those seen in classical dermatomyositis (DM).[3,4]

Systemic inflammatory vasculopathy is an important characteristic of JDM affecting especially the microcirculation.[5] Nailfold capillaroscopy (NFC) is a non-invasive method that allows the direct visualization of nailfold capillaries.[6] Decreased number of capillary loops (devascularization) associated with enlarged capillaries and branching capillary loops are the most characteristic findings observed in JDM.[7] In addition, several studies have described an association between NFC abnormalities and JDM severity and activity.[8] To the best of our knowledge, NFC abnormalities have not been systematically studied in CADM. We describe herein the case of a 4-year-old child diagnosed with juvenile CADM with important changes in NFC, whose response to treatment was followed by significant improvement in capillaroscopy abnormalities.

Case report

In June 2008, a 4-year-old girl was attended with a four months complaint of malar rash, photosensitivity, and erythematous lesions over the proximal interphalangeal joints, elbows and knees, with no complaints regarding muscle strength loss or pain. The Manual Muscle Testing (MMT)[9] score was 80/80, Childhood Myositis Assessment Scale (CMAS)[9] was 48/52, muscular Disease Activity Score (DAS) was 2/11 and cutaneous DAS was 6/9.[10] Laboratory tests showed hemoglobin 13.6 g/L, hematocrit 38.5%, leukocytes 21,000/mm^3 (76% neutrophils, 16% lymphocytes), platelets 289,000/mm^3, erythrocyte sedimentation rate (ESR) 23 mm/1st hour, C-reactive protein(CRP) undetectable, aldolase 20.8 IU/L (normal < 7.6), creatine kinase (CK) 130 IU/L (normal < 204), lactate dehydrogenase (LDH) 575 IU/L (normal < 480), aspartate aminotransferase (AST) 29 IU/L (normal < 34), alanine aminotransferase (ALT) 14 IU/L (normal < 44), and positive antinuclear antibody (1/640, homogeneous fine speckled pattern). Capillaroscopy was performed in all fingers of both hands using a stereomicroscope (Olympus SZ40) at 10× to 16× magnification under epi-illumination at 45°, analyzing number of capillary/mm, enlarged, giant and branched capillaries, and avascular score.[11] Capillaroscopy showed a scleroderma (SD) pattern,

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