Clinical Image

Widespread Perifascicular Pathology in Juvenile Dermatomyositis

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CLINICAL IMAGE

Idiopathic inflammatory myopathies (IMs) are a heterogeneous group of autoimmune muscle disorders, which can be divided into four main categories including polymyositis (PM), dermatomyositis (DM), immune-mediated necrotizing myopathy (NM) and sporadic inclusion body myositis (sIBM) based on clinical, histological and serological characteristics. The diagnosis is established by muscle biopsy, in which certain pathologic findings are unique for each subset.

We report a case of a 4½ years old boy, who presented with progressive proximal muscle weakness of the upper and lower extremities associated with constantly elevated serum creatine kinase (CK) levels (2243 U/l, normal 26-170 U/l) and increase in liver transaminase levels AST 249 U/l (5-40 U/l) and ALT 166 U/l (5-35 U/l) in consecutive measurements. One month later a symmetrical violaceo-us rash was seen periorbitally. Autoimmune tests were normal. Electromyography revealed myopathic changes with spontaneous discharges in the examined muscles. Muscle biopsy of vastus lateralis showed perifascicular atrophy with increased HLA-I immunohistochemical expression without inflammatory infiltrates, multiple fibers deficient in cytochrome oxidase (COX) activity and prominent glycogen accumulation in perifascicular distribution Figure.

Dermatomyositis is typically characterized by perifascicular atrophy, even in the absence of inflammation, which involves both type 1 and type 2 muscle fibers. Muscle biopsy can also reveal characteristic mitochondrial pathology in perifascicular regions, as is seem by the multiple COX-negative fibers. However, there is an increasing body of evidence supporting a more widespread pathology in perifascicular areas, such as an occasionally significant glycogen accumulation, which has been previously described in juvenile DM and is not similar to the pattern of glycogen deposits observed in muscle glycogenoses. Interestingly these pathological findings in the present child of similar age with DM, may reflect a possible metabolic adaptation of the hypoperfused perifascicular areas to hypoxia. Whether glycogen perifascicular accumulation might be more frequently observed in juvenile dermatomyositis has to be elucidated.

REFERENCES


Figure 1 Histology of the vastus lateralis muscle biopsy.
Figure A: Hematoxylin and eosin staining shows perifascicular atrophy (4x)
Figure B: increased HLA-I immunohistochemical expression in the region of perifascicularatrophy (4x)
Figure C: Multiple fibers with deficient COX activity (COX-negative fibers) (4x)
Figure D: Periodic acid – Schiff staining shows increased PAS-positive staining in perifascicular distribution (4x).