Kocher–Debré–Semelaigne syndrome with rhabdomyolysis and increased creatinine: a case report.

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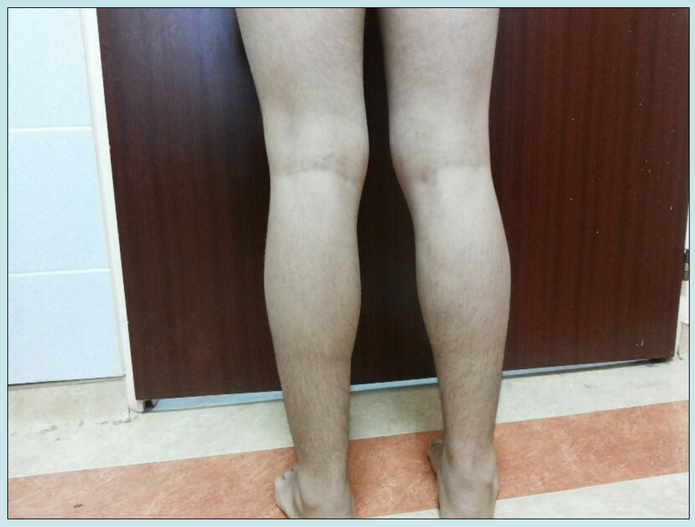
Objectives:

Kocher–Debré–Semelaigne syndrome (KDS) is a kind of myopathy in hypothyroidism characterized with hypertrophy of muscles. Muscle enzymes such as creatine kinase (CK) may be moderately elevated in hypothyroid patients. Though rhabdomyolysis due to hypothyroidism is rare, especially when other precipitating factors such as exercise, lipid-lowering drugs, alcohol and renal failure, are absent .
We describe a case of severe myopathy, massively elevated CK levels and high creatinine levels presenting with poly/dermatomyositis-like symptoms, diagnosed as KDS with rhabdomyolysis.

Methods:

A 15-year-old boy was admitted to our clinic complaining of generalized muscle pain, lethargy, dry coarse skin, swelling of hands and feet. His complaints had come on gradually within last 6 months. He denied any recent event of muscle injury. On examination he had generalized oedema of face and body, a dry pallor coarse skin, and diffuse goitre. His calf muscles were hypertrophied with minimal proximal muscle weakness in the lower limbs. He had muscle pain in the lower back, legs and arms with a firm feel. He had Tanner's stage 5. His height was 165cm (25th percentile) and weight was 61.7 kg (50th percentile). Deep tendon reflexes were diminished.

Creatinine	1.68 mg/dL
Blood urea	21 mg/dL
Free T4	0.18 ng/dL (0.89-1.76)
Thyroid stimulating hormone (TSH)	>150 µIU/mL (0.56-5.57)
Anti-microsomal antibody:	>1000 U/mL (normal<35)
Anti-thyroglobulin antibody	>3000 U/mL (normal <40)
Serum CK	4267 U/L (30-200)
Myoglobin	170.5 ng/ml (16-74)
Total cholesterol	300 mg/dl (130-200)
LDL cholesterol	199 mg/dl (50-160)
ALT	66 U/L (0-55)
AST	175 U/L (5-34)
Hemogram	Completely normal
Urine analysis	Completely normal



Results:

All these findings suggested an associated diagnosis of KDS and rhabdomyolysis, as a result of hypothyroidism due to autoimmune thyroiditis. The patient was treated with intravenous fluids and was commenced on thyroxine replacement therapy (75 µg of levothyroxine daily). Two weeks after admission, he had dramatic symptomatic improvement and muscular pseudohypertrophy decreased some. His CK, AST and ALT levels dropped to 193 U/L, 38 U/L and 25 U/L respectively, while his creatinine was 1.36 mg/dL. He was discharged. The therapy led to normal laboratory findings for serum creatinine after 4 weeks and for thyroid function test after 3 months, with levothyroxine dosage adjustment.

Conclusions:

•Good outcome of hypothyroid manifestations requires evaluation of thyroid function in cases of myopathy.
•Any patient presenting with myopathy of uncertain origin, accompanied by elevated serum muscle enzymes and serum creatinine should be evaluated for KDS and associated rhabdomyolysis.
•Hypothyroidism should be considered in cases with rhabdomyolysis of unknown etiology.

References:

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