MOTOR POLYNEUROPATHY MAY BE THE PRESENTING FEATURE OF ACUTE INTERMITTENT PORPHYRIA

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A 25-year-old female presented with progressive quadriparesis for the last two weeks. Appendectomy and cholecystectomy operations were recorded in her past medical history. Family history was unremarkable. In the physical examination, pulse was 132/dk and blood pressure was 166/110 mmHg. Motor examination revealed quadriparesis to be more prominent in the upper extremities and deep tendon reflexes were decreased bilaterally. The patient was admitted with a diagnosis of acute motor polyneuropathy that was also supported by electroneuromyographic findings. Routine blood tests showed anemia, hyponatremia and AST, ALT, ESR elevation. ANA IFA showed a granular pattern and anti-SSA was positive. Schirmer's test revealed dry eyes, but the salivary gland biopsy was reported to be normal. Methylprednisolone pulse therapy was started. Tachycardia and hypertension responded only to esmolol infusion. Acute intermittent porphyria (AIP) was diagnosed due to the clinical and laboratory findings and high urine porphobilinogen level (48.4 mg/d). High-calorie diet with general nutritional support was started. The patient was informed about AIP and a list of the drugs that should be avoided has been given. During the hospitalization muscle strength, physical and laboratory abnormalities improved markedly and she could be able to walk without any assistance.

We consider three important points in this case; 1- AIP should be kept in mind in the cases presenting with motor polyneuropathy even if the diagnosis was not done previously; 2- AIP has various systemic findings that may lead to the diagnosis; 3- SSA positivity may be seen in AIP that is not well-known among neurologists.