INTRODUCTION: Automutilation is a common psychiatric behavioral disorder. However, it could be a revealing sign of a rare neurological disease such as neuroacanthocytosis.

CASE REPORT: We report a case of a 39-year-old patient, without previous neurological history. She was born to consanguineous parents. She has developed insidious oral automutilations. She was examined and followed by dermatologists and psychiatrists. No etiology was retained. Few months later, she developed movement disorders. Neurological examination has confirmed choreic movements in the head and upper limbs, tendon areflexia and cognitive impairment. Blood smear revealed the presence of acanthocytes (9%). Biological assessment showed a high level of muscular enzymes (CPK=1000 IU/L, LDH=600 IU/L). Cerebral MRI showed an atrophy of caudal nuclei. The EMG concluded sensitivo-motor axonal neuropathy, predominant in inferior limbs. Genetic assessment for Huntington disease was negative. We retained the diagnosis of neuroacantocytosis and the treatment was mainly symptomatic (neuroleptics and vitamins). Evolution and prognosis were poor.

DISCUSSION: Neuroacanthocytosis is an autosomal recessive affection, with a progressive evolution. It appears often in young male adults and rarely among women such in our case. Clinical symptoms include movement disorders (chorea, orofacial dyskinesia and oro-lingual automutilations), peripheral neuropathy and frontal dementia. Research of acanthocytes in blood smear is important for diagnosis, showing a percentage above 5%. Persistent elevation of CPK is usual. MRI and genetic research showed that an atrophy of caudal nuclei can cause mutation in chromosome 9q21-22 gene. Treatment is mainly based on vitamins, neuroleptics and psychotherapy.

CONCLUSION: Neuroacanthocytosis is a rare affection with a polymorphous clinical expression. We should consider psychiatric symptoms as an inaugural form of its revelation.

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