Specific Comments to Authors: In this paper the authors discuss a case of SCA3 with an unusual clinical presentation. I appreciate the effort of the authors to give the readers a different post of view in represent the discussed disease. I suggest moreover to better specify the clinical description of sca3 in order to subtitle the differences between normal presentation and abnormal one.

Reply: Thank you very much for the recommendation.

(1) We have revised the subtitle: Rare SCA3 with dopamine sensitivity.

(2) Spinocerebellar ataxia type 3 (SCA3) is an autosomal dominant genetic disease first described by Nakano et al. among Portuguese immigrants in the United States in 1972. It is characterized by progressive cerebellar ataxia accompanied by paralysis of extraocular muscles, dysphagia, lingual fibrillation, pyramidal tract signs, extrapyramidal system signs, and other clinical manifestations.

SCA3 is divided into several clinical subtypes: Type 1 (or Joseph type) is characterized by ataxia, ophthalmoplegia, pyramidal and extrapyramidal signs and is the most serious type of all. Type 2 (or Thomas type) is characterized by ataxia and ophthalmoplegia with or without pyramidal and extrapyramidal signs. Type 3 (or Machado type) is characterized by ataxia, ophthalmoplegia and peripheral neuropathy and exhibits a later onset than the other types. Subsequently, other clinical phenotypes have been described: type 4 with parkinsonian features, type 5 with spastic paraparesis without ataxia and type 6 with pure cerebellar ataxia. However, there is still no consensus among researchers. These subtypes will be inverted into one another during the disease's progression or exhibit 2 or more of them in one patient, which contribute to the clinical complexities and suggest that it could not have valuable role in assigning one patient to a definite subtype.

The early clinical manifestations of the disease are not typical, and the diagnosis is difficult. It needs to be distinguished from dopamine responsive dystonia and Parkinson's disease. The differential diagnosis is complex, and

there is no good drug in clinic.