Dear editor:  
I am very glad to receive your email and get the comments of the reviewers. We have seriously revised my manuscript and made some required changes according to the reviewer’s comments point by point.

Here are answers to the first reviewer’s comments:

1. We have added a figure of the GC-MS spectrum as figure2, showing the increased 2HG concentrations. The former ‘figure2’ was renamed as figure 3.

2. Answer to question 2: We did not separate the 2HG L-enantiomer from the 2HG D-enantiomer in the GC-MS analyses. The patient’s clinical presentation and imaging suggested L-2-HGA, and we confirmed the diagnosis according to the gene analysis.

3. We have added an arrow indicating the mutation on the Sanger plot.

4. In patients with autosomal recessive disease, some mutations are homozygous, and some are compound heterozygous. For L2HGA, there are a few compound heterozygous cases have been reported. For example, there was four L-2HGA patients with compound heterozygous mutations in the fourth reference. (Vilarinho L, Cardoso ML, Gaspar P, Barbot C, Azevedo L, Diogo L, et al. Novel L2HGDH mutations in 21 patients with L-2-hydroxyglutaric aciduria of Portuguese origin. Hum Mutat. 2005; 26: 395-6.)

5. We have reedited the manuscript for the expressing and grammatical error, including:
   a. who had characteristic developmental delay, ataxia and acrocephaly as the main symptom#main symptoms
   b. He also complained paroxysmal headache #He also complained of paroxysmal headache
   c. Urine test for organic acids showed a significantly increase level of 2-hydroxyglutaric acid#increased level of 2-hydroxyglutaric acid
   d. c.169G>A in exon 2 and c.542G>T in exon 5, not hitherto described. #not hitherto been described.
e. Novel gene mutation and associated clinical symptom can contribute for the understanding and identification of this rare disease. #associated clinical symptoms
f. Possible genotype-phenotype correlation waits further investigation. #waits for further study.
g. leading to the diagnosis of L-2-hydroxyglutaric aciduria. #leading to the probable diagnosis of
Answers to the second reviewer’s comments:
1. For the novel mutation, we used three software tools including SIFT, PlyPhen and Mutationtaster to predict the possible impact of the amino acid substitution on the structure and function of the protein L-2-hydroxyglutarate dehydrogenase. And we have added the result in the manuscript. But we did not do the real functional test.
2. It is the same question to the third one above.
Thank you for reviewing and giving valuable advices. If you find any other problems, please contact me and direct me to revise it.
With best wishes.
Tai Hongfei