Response to reviewers' comments:

Thank you for the reviewers' comments concerning our manuscript entitled "Diagnosis and treatment of an inborn error of bile acid synthesis type 4: A case report" (ID: 67159). Those comments are all valuable and very helpful for revising and improving our paper, as well as the important guiding significance to our researches. We have studied comments carefully and have made correction which we hope meet with approval. Revised portion are marked in red in the paper.

Response to reviewer 1:

1. The English language needs minor polishing for some grammar and typos errors.

   Reply: We are very sorry for our negligence of some grammar and typos errors. And we have modified them. For the Running Title, we changed it to A case report of IEBas4. In DISCUSSION, we replace "Repeated" in line 5 of paragraph 3 with "repeated".

2. Response to comment: AMACR should be written in full name for the first time.

   Reply: We have made correction according to the Reviewer's comments. In the BACKGROUND, the full name of AMACR is written and the full name of AMACR in the INTRODUCTION is deleted.

Response to reviewer 2:

Reply: Tanks for your high praise.

Other changes:

1. In Discussion, we revise it in the second paragraph. The contents are as follows: In 2000, Ferdinandusse *et al* reported three adult patients with progressive sensory neuropathy, but presenting without fat-soluble vitamin malabsorption and liver disease. *AMACR* gene detection in these three patients revealed gene mutations, and fibroblast culture confirmed the damage to the synthetic pathway. In 2003, Setchell and co-workers reported a case of AMACR deficiency in a child with fat-soluble-vitamin deficiency, coagulopathy and mild cholestatic liver disease in the neonatal period. Analysis of the patient's blood and urine showed significant elevation of 25R-THCA. Genetic testing confirmed a mutation in the *AMACR* gene, and fibroblast studies also confirmed AMACR deficiency.