The authors report three cases of isolated sulphite oxidase deficiency with late onset, spontaneous recovery and a relatively mild clinical course.

Patients presented with encephalopathy (decreased consciousness) in one patient, a generalized seizure in another patient, choreoathetosis in one patient and «hypotonia and acute regression» in all.

Information about lens dislocation is not provided.

The clinical description is, in my view, insufficient: acute onset means hours? Days? Was there a decreased level of consciousness? Cranial nerve examination was normal? Were there clinical signs of ataxia? Hypotonia and hypertonia that these patients sequentially revealed was bilateral? Generalized? Reflexes were normal? Decreased? Hyperactive? Plantar responses were flexor? Ophthalmologic examination was normal?

MRI should be reported with details on which sequences were chosen to demonstrate the lesions and wether these lesions were hyper or hypointense in the text, not only in the legends

Pedunculus cerebri is not in common use in English Neurology literature. I would say from the pictures provided that the hyperintense lesions involve the substantia nigra (?)

I would not state that bilateral lesions in the globus pallidus suggest ISOD as the differential diagnosis for these lesions is extensive; dislocated lens and/or low plasma homocysteine would be important clues for this diagnosis

I also point out that the patient reported in Reference 11 recovered spontaneously from acute, recurrent encephalopathy without any dietary management
Finally I detect many orthographic and syntactic issues that I respectfully suggest should be reviewed

**Are the methods appropriate and well described?**
If not, please specify what is required in your comments to the authors.

No

**Does the work include the necessary controls?**
If not, please specify which controls are required in your comments to the authors.

Unable to assess

**Are the conclusions drawn adequately supported by the data shown?**
If not, please explain in your comments to the authors.

Yes

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Not suitable for publication unless extensively edited

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