

## Case Report

**Corpora amylacea mimicking low-grade glioma and manifesting as a seizure: Case report**Seung J. Lee<sup>1</sup>, Minsu Kim<sup>4</sup>, Carlito Lagman<sup>1</sup>, Timothy T. Bui<sup>1</sup>, William H. Yong<sup>2</sup>, Isaac Yang<sup>1,3</sup>Departments of <sup>1</sup>Neurosurgery, <sup>2</sup>Pathology and <sup>3</sup>Jonsson Comprehensive Cancer Center, University of California, Los Angeles, Los Angeles, California, USA, <sup>4</sup>Department of Neurosurgery, Yeungnam University College of Medicine, Daemyung-dong, Nam-gu, Daegu, KoreaE-mail: Seung J. Lee - [seungjlee@mednet.ucla.edu](mailto:seungjlee@mednet.ucla.edu); Minsu Kim - [MinsuKim@mednet.ucla.edu](mailto:MinsuKim@mednet.ucla.edu); Carlito Lagman - [carlitolagman@gmail.com](mailto:carlitolagman@gmail.com); Timothy T. Bui - [timothybui@mednet.ucla.edu](mailto:timothybui@mednet.ucla.edu); William H. Yong - [wyong@mednet.ucla.edu](mailto:wyong@mednet.ucla.edu); \*Isaac Yang - [iyang@mednet.ucla.edu](mailto:iyang@mednet.ucla.edu)  
\*Corresponding author

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**Abstract****Background:** Corpora amylacea (CA) are accumulations of polyglucosan bodies typically found in astrocytic foot processes, and rarely, can mimic neoplasm. CA accumulation has also been associated with seizure disorders. We report the first case of a histologically confirmed intracranial, intraparenchymal CA lesion mimicking a low-grade glioma and manifesting as a seizure.**Case Description:** A 43-year-old man presented after a general tonic-clonic (GTC) seizure. Brain magnetic resonance imaging (MRI) revealed a small lesion in the right mesial temporal lobe with radiologic features of a low-grade glioma. The patient underwent a right pterional craniotomy for resection of the lesion. Histology demonstrated abundant polyglucosan bodies without neoplastic features. The patient tolerated the procedure well, was free from seizures without antiepileptic drugs at 2-week follow-up, and is undergoing serial surveillance.**Conclusion:** The clinical manifestation of CA as a seizure in the context of an identified brain mass is extraordinarily rare. Nevertheless, CA should be considered in the differential diagnosis for patients with seizures and a radiologically identifiable low-grade lesion. Symptomatic CA lesions Mimicking a low-grade glioma should be surgically pursued with a goal of safe, maximal resection to confirm the diagnosis and to provide the patient with prognosis, which can significantly impact patient quality of life.**Key Words:** Case reports, corpora amylacea, glioma, pathology, seizures**Access this article online****Website:**[www.surgicalneurologyint.com](http://www.surgicalneurologyint.com)**DOI:**

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**Quick Response Code:****INTRODUCTION**Corpora amylacea (CA) are aggregates of polyglucosan bodies consisting of insoluble polysaccharides and protein.<sup>[1,2]</sup> CA accumulates in the human brain as a consequence of normal aging, stress, insults, and many neurological diseases, such as epilepsy, Alzheimer's disease, multiple sclerosis, and Lennox-Gastaut syndrome.<sup>[1-14]</sup> The exact role of CA accumulation in physiology and pathology are not fully elucidated.<sup>[1,3]</sup> We

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describe the case of a 43-year-old man who presented after a general tonic–clonic seizure (GTC), and was found to have a small lesion with radiologic features of a low-grade neoplasm, which was resected. The histological analysis of the lesion revealed prominent polyglucosan bodies without neoplastic features. To our knowledge, this is the first case of a CA lesion radiologically mimicking a low-grade glioma and causing a seizure.

## CASE DESCRIPTION

### History and examination

A 43-year-old, right-handed man without a prior history of epilepsy presented to his neurologist for evaluation of a GTC seizure. He had no other symptoms and was neurologically intact. He was prescribed an antiepileptic drug and underwent radiological evaluation.

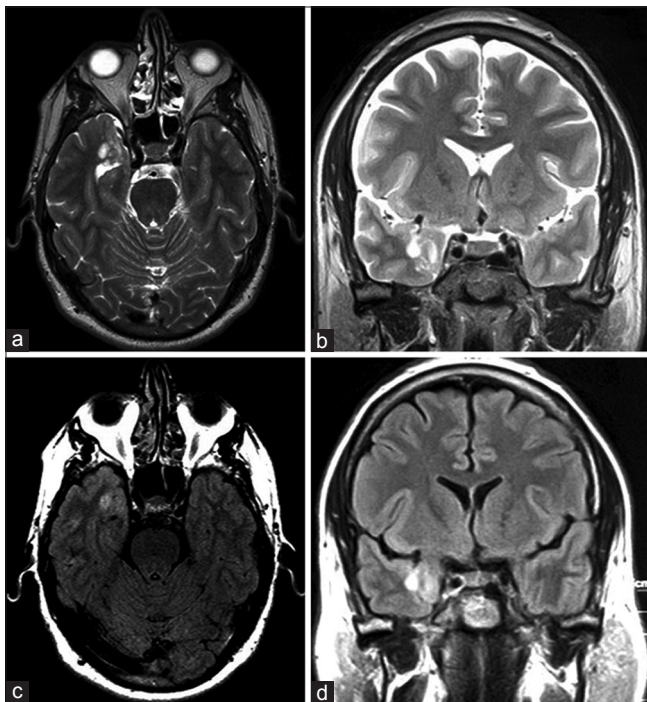
Magnetic resonance imaging (MRI) of the brain revealed a nonenhancing lesion measuring  $1.5 \times 1.8$  cm in the right mesial temporal lobe with minimal mass effect. The lesion appeared hyperintense on T2-weighted images [Figure 1a and b]. Fluid-attenuated inversion recovery (FLAIR) sequences delineated a nonspecific hyperintense focus in the right temporal lobe near the gray-white interface [Figure 1c and d]. Diffusion-weighted imaging (DWI, images unavailable) did not reveal any abnormal restricted diffusion. Positron emission tomography (PET, not shown) was

obtained, but did not reveal significant uptake of 6-[ $^{18}\text{F}$ ] Fluoro-L-DOPA ( $^{18}\text{F}$ -FDOPA) in the right mesial temporal lobe. The radiologic features of the lesion were suggestive of a low-grade neoplasm, and the patient was referred to neurosurgery.

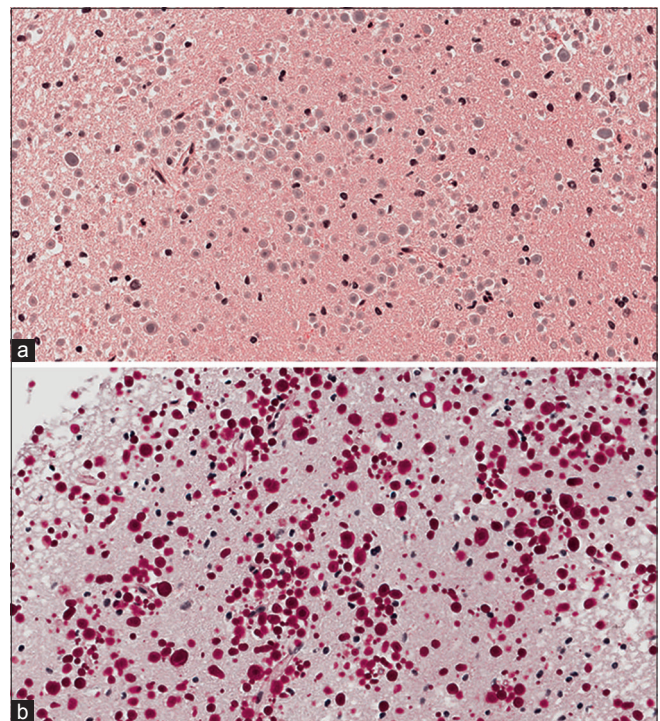
A right pterional craniotomy was performed for resection of the mass. The lesion was noted to have a firm consistency, ill-defined planes, and significant pial adhesions. The lesion was meticulously resected under the operative microscope, and the specimen was sent to pathologic examination. The patient tolerated the procedure well. Immediate postoperative imaging demonstrated confirmed gross-total resection without evidence of ischemia or abnormal enhancement. At 2-week follow-up, the patient was free from seizures and no longer required antiepileptic drugs. The patient has not returned in the 13 months since the surgery.

### Pathological examination

Histological examination of the lesion revealed normal gray matter with focally prominent polyglucosan body accumulations in the brain parenchyma. Numerous spherical polyglucosan bodies were identified on Hematoxylin and Eosin (H and E) frozen section slides without features of neoplasia. Permanent section H and E slides [Figure 2a] also demonstrated the presence of numerous polyglucosan bodies, far in excess of those found in the normal aging brain. Periodic-acid Schiff stain highlighted the polyglucosan bodies [Figure 2b], providing



**Figure 1:** Schematic diagram of T2-weighted axial (a) and coronal (b) images revealing a nonenhancing hyperintense lesion in the right mesial temporal lobe. Axial (c) and coronal FLAIR sequences (d) showing a hyperintense lesion in the white matter of the right mesial temporal lobe



**Figure 2:** Schematic diagram of a 20 $\times$  magnification of H and E stained specimen revealing polyglucosan bodies (a) and 20 $\times$  magnification of CA stained pink with periodic-acid Schiff staining (b)

a final diagnosis of a dense CA lesion. This retrospective review of a single patient's medical record did not require informed consent by the Institutional Review Board. Nevertheless, all images are completely de-identified.

## DISCUSSION

Glucose monomers within neuroglial cells are thought to polymerize in the setting of cellular degeneration to form aggregates of polyglucosan bodies (hence the term *corpora amalycea*, Latin for body and starchy, respectively); however, the physiologic and pathologic significance of these lesions are not fully elucidated.<sup>[1,4]</sup> Several studies report CA accumulation in normal aging, anoxic, and hypoxic brain, Alzheimer's disease, and seizure disorders, such as mesial temporal sclerosis, Lennox–Gastaut syndrome, and Lafora-type progressive myoclonic epilepsy.<sup>[1-15]</sup>

CA accumulation can also have a genetic etiology. Adult polyglucosan body disease (APBD) is a rare autosomal recessive genetic disorder caused by a glycogen branching enzyme deficiency, leading to CA accumulation in the central and peripheral nervous system.<sup>[2,3,11,14-17,20,23,26,27]</sup> Mochel *et al.*<sup>[3]</sup> demonstrated that bilateral hyperintense lesions on T2 and FLAIR sequences in the medulla, pons, internal and external capsule, and periventricular regions are characteristic in patients with APBD. Furthermore, these patients typically present with other neurological symptoms attributable to these lesions, such as seizures, cognitive deficits, neurogenic bladder, and neuropathy.<sup>[21]</sup> The unilateral lesion and lack of other neurological symptoms make APBD an unlikely diagnosis in our patient.

MRI is highly sensitive for brain tumors, although it lacks the specificity to distinguish low-grade from high-grade neoplasms.<sup>[12]</sup> One distinguishing feature of high-grade neoplasms is enhancement, which represents breakdown of the blood–brain barrier seen in infiltrative, high-grade lesions. Our patient's T2-weighted images showed a nonenhancing lesion, suggestive of a low-grade glioma. Furthermore, the lesion did not show abnormal diffusion on DWI sequences. Cerebritis and abscesses typically show abnormal restriction in DWI, which placed these diagnoses lower on our differential.<sup>[9,10,18,22,24,25]</sup>

PET imaging has also been proven to be useful in the diagnosis of low-grade gliomas. Chen *et al.* reported that the sensitivity in detecting brain tumors with 18F-DOPA is higher (sensitivity, 96%; 95% CI, 87–100%) than 18F-fluodeoxyglucose (18F-FDG) PET imaging (sensitivity, 61%; 95% CI, 41–81%).<sup>[5]</sup> Given the high sensitivity of 18F-FDOPA PET imaging and lack of uptake in our patient, a false negative was statistically unlikely. However, low-grade glioma remained high on our differential because of the patient's seizures and the MRI features of the lesion.

CA accumulation has been described in patients with temporal lobe epilepsy and other seizure disorders.<sup>[1,8,19]</sup> Kawamura *et al.* noted hyperintense lesions on FLAIR sequences that correlated with increased accumulations of CA with hippocampal atrophy.<sup>[13]</sup> In these patients, hippocampal atrophy was thought to be the primary cause of seizures. Our patient did not exhibit any signs of hippocampal atrophy and did not have a prior history of epilepsy. Lastly, medial temporal sclerosis has distinct radiologic features and rarely presents as a well-demarcated, spherical lesion.<sup>[6,7]</sup>

Abel *et al.*<sup>[1]</sup> reported the only other case of a CA lesion radiologically resembling low-grade glioma on MRI in a 49-year-old female with increasing migraine intensity. Her history was negative for seizures or other neurological problems. Because our patient remains seizure-free without the use of antiepileptic drugs postoperatively, we attribute the seizure directly to the CA lesion.

## CONCLUSION

This case illustrates an extraordinarily rare instance in which CA aggregates appeared as a well-demarcated, spherical mass lesion causing a seizure. Although uncommon, CA should be considered in the differential diagnosis for lesions with radiological characteristics of a low-grade glioma.

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## Conflicts of interest

There are no conflicts of interest.

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