Isolated amygdala neurocysticercosis in a patient presenting with déjà vu and olfactory auras

Case report

DARRIN J. LEE, B.S.,1 CHRISTOPHER M. OWEN, M.D.,1 ELHAM KHANIFAR, M.D.,2 RONALD C. KIM, M.D.,2 AND DEVIN K. BINDER, M.D., PH.D.1

Departments of 1Neurological Surgery and 2Pathology, University of California, Irvine, California

Neurocysticercosis is the most common parasitic infection in the CNS and a leading cause of epilepsy. Since it is a circumscribed lesional cause of epilepsy, specific locations of neurocysticercal lesions may lead to specific clinical presentations. The authors describe a 17-year-old Hispanic boy who had a single enhancing bilobar mass in the right amygdala. Initially, the patient presented with secondarily generalized tonic-clonic seizures, which resolved with antiepilepsy drug therapy. On further investigation, he was found to have persistent olfactory and déjà vu auras. A right amygdalectomy without hippocampectomy was performed, and both the seizures and auras immediately resolved.

Pathological analysis revealed neurocysticercosis. To the authors’ knowledge, this case is the first reported instance of 2 distinct mesial temporal aura semiologies associated with localized neurocysticercosis in the amygdala and successfully treated with resection. Uniquely, the case demonstrates that both olfactory and déjà vu auras can emanate from the amygdala. (DOI: 10.3171/2009.2.PEDS08140)

Key Words • amygdala • aura • déjà vu aura • neurocysticercosis • olfactory aura

Neurocysticercosis, caused by CNS infection with the larval form of Taenia solium, is one of the most common causes of epilepsy in Latin America, China, India, and Africa.8 In the US, epilepsy due to neurocysticercosis has become increasingly prevalent.10,11 Patients commonly present with partial seizures with or without secondary generalization.12 In selected cases, surgical treatment is beneficial both for diagnosis and cure, but chronic lesions can lead to intractable epilepsy.20

Auras (Greek word for “breeze”), which have the same clinical and electrical definition as simple partial seizures, have been defined as “that portion of a seizure which occurs before consciousness is lost and for which memory is retained afterwards.”9 Despite the frequency of auras, they are not included in the International League Against Epilepsy (ILAE) standard classification of epileptic seizures.2 In a recent report from this commission, however, distinct epileptic auras were categorized as 1) epigastric, 2) fear and anxiety, 3) experiential (including déjà vu and jamais vu), 4) olfactory-gustatory (“uncinate fits”), 5) autonomous-vegetative, and 6) nonspecific auras.3 The localizing value of auras is based largely on studies in patients with mesial temporal sclerosis, which can involve multiple mesial temporal structures including the hippocampus and amygdala.10-12

In this report, we present a unique case of isolated amygdala neurocysticercosis in a patient presenting with olfactory and déjà vu auras and complex partial seizures with secondary generalization. Selective right amygdala lesionectomy led to the elimination of all auras and seizures. Thus, this case proves that these 2 types of auras can arise specifically from the amygdala.

Case Report

History and Examination. This 17-year-old Hispanic boy presented with a 1-year history of intermittent sei-
zures. He had traveled to Mexico approximately once per year. Within a span of 3 months, he experienced 3 tonic-clonic seizures with loss of consciousness. After the third seizure, he was placed on phenytoin. Since taking the anticonvulsant, he has experienced no tonic-clonic seizures; however, he did note multiple daily auras consisting of déjà vu experiences and/or a distinct recurrent unpleasant smell. The déjà vu experiences occurred so frequently that they were witnessed in real time on multiple clinic visits. The patient reported no concomitant headaches, language or memory difficulty, visual deficits, nausea, or vomiting. The neurological examination was entirely normal.

Brain MR imaging studies demonstrated a 2 × 1.5–cm multilobulated cystic abnormality in the right amygdala with peripheral rim enhancement (Fig. 1). Coronal and axial FLAIR images demonstrated perilesional hyperintensity. These images were consistent with either a neoplastic or infectious/inflammatory process. Only 1 lesion was observed.

Operation. After obtaining informed consent, a right pterional craniotomy and lesionectomy were performed using Stealth neuronavigation to make a tissue diagnosis and in the attempt to cure the auras and/or seizures. A transcortical temporal gyrus approach was used as in the transcortical selective amygdalohippocampectomy procedure. The lesion was encountered in the basolateral amygdala and an en bloc lesionectomy was performed (consisting of a right basolateral amygdalectomy without hippocampectomy). Marked perilesional white matter edema was observed. Entry into the temporal horn was verified with the appearance of ependyma and choroid plexus. The hippocampus was not involved. The uncus was removed and seemed edematous, but the lesion did not extend into the uncus. Other than the small corticectomy through the middle temporal gyrus, the lateral temporal gyri were left intact. The lesion itself was indurated and bilobed, and an intraoperatively cut cross-section demonstrated an amorphous yellowish material inside the cyst (Fig. 2).

Postoperative Course. Permanent pathology revealed a degenerated cysticercus with an extensive inflammatory reaction, including multinucleated giant cells (Fig. 3). Postoperative imaging demonstrated gross-total resection of the lesion (Fig. 4). The patient had transient incisional pain and headaches that subsequently resolved, and he was discharged home neurologically intact on postoperative Day 3 and instructed to continue the phenytoin. He has been completely aura and seizure free for 10 months postoperatively and has returned to work.

Discussion

In this patient with lesional epilepsy, frequent olfactory and déjà vu auras remained after initial medical management of complex partial and secondarily generalized seizures. Resection of an isolated neurocysterceral mass in the right amygdala led to complete freedom from both the seizures and auras. This case demonstrates that olfactory and déjà vu aura semiology can be related specifically to amygdala pathology—in this case a neurocysticercal cyst.

Many different sensory and experiential auras/simple partial seizures have been described, including visual, auditory, olfactory, and gustatory sensory auras; and déjà vu and jamais vu experiential auras.3–5 Using electrical stimulation and studies of mesial versus lateral temporal lobe epileptic discharges, Pierre Gloor11 and colleagues9,12 have localized experiential phenomena to the mesial temporal lobe as opposed to the temporal neocortex. Specifically, mesial temporal limbic structures, such as the amygdala and hippocampus, have been identified in perceptual hallucinations, including flashbacks and déjà vu experiences.11,12 Based on electrical stimulation studies, experiential phenomena are more common in the amygdala than in the hippocampus when associated with no afterdischarge,
whereas they seem more prevalent in the hippocampus in the presence of an afterdischarge. Similar findings have been observed in patients with spontaneous seizures. With or without afterdischarge, experiential phenomena are not as common in the temporal neocortex. The role of the neocortex is further elucidated by the fact that the early spread of an electrical discharge to the neocortex actually appears to prevent hallucinations or illusions, further emphasizing the role of the mesial temporal structures in experiential phenomena. By fortuitously finding a lesion confined to the amygdala and not other mesial temporal structures (hippocampus, parahippocampal gyrus), our case uniquely demonstrates that amygdala pathology alone is sufficient to generate olfactory and déjà vu auras.

There are 3 prevailing theories regarding the relationship between epilepsy and neurocysticercosis: 1) a noncausal relationship; 2) dual pathology, where 2 autonomous conditions exist coincidentally; and 3) a causal relationship between epilepsy and active or transitional intracerebral cysticerci. One study has suggested that neurocysticercosis is not necessarily epileptogenic, supporting the hypothesis that there is no relationship between the presence and localization of neurocysticercosis and the occurrence of seizures. If intraparenchymal neurocysticercosis is suspected based on imaging and serology, a trial of antihelmintic drugs, such as albendazole, along with steroids and a first-line antiepilepsy drug can be used to treat seizures. However, surgical treatment of neurocysticercosis can be successful with well-defined localization-related epilepsy even in the presence of disseminated calcified neurocysticercal lesions. Failed medical management may therefore require surgical removal for the relief of symptoms. Our case supports the third theory given that the cysticercal infection was present exclusively in the amygdala, seizures clearly arose from this area by semiology, and resection of the neurocysticercal cyst led to seizure and aura freedom.

While chronic epilepsy has been reported to develop in sites distant from cysticerci, there appears to be a relationship among seizure type, anatomical location of the cysticerci, and the presence of calcification. The epileptogenic effect of a cysticercal cyst may be due to inflammatory changes in the surrounding tissues, a proposal supported by the perilesional FLAIR imaging abnormalities in our case, although the detailed pathophysiology is unknown. While neurocysticercosis may be causally related to mesial temporal lobe epilepsy associated with hippocampal sclerosis, our case demonstrates that a neurocysticercal lesion can induce auras without associated hippocampal sclerosis. Finally, excellent relief from both seizures and auras can be achieved through gross-total resection of the neurocysticercal cyst.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**References**

Amygdala neurocysticercosis


Address correspondence to: Devin K. Binder, M.D., Ph.D., Department of Neurological Surgery, University of California, Irvine, 101 The City Drive South, Building 56, Suite 400, ZOT 5397, Orange, California 92868-3298. email: dbinder@uci.edu.