

Calcified Pseudoneoplasm of the Neuraxis (CAPNON)–A Rare Cause for Temporal Lobe Epilepsy: Not all Warrant a Surgical Intervention

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Abstract

Epilepsy is a common neurological condition with varied etiological causes, with temporal lobe epilepsy being the most common. Among the varied etiologies of temporal lobe epilepsy, mesial temporal sclerosis is an important one and it presents as intractable epilepsy. However, we describe here a case of intractable temporal lobe epilepsy with a rather rare etiology, calcifying pseudo neoplasm of neuraxis (CAPNON) syndrome. CAPNON is a rare benign lesion that can occur anywhere in the central nervous system. The thought process till date is to excise any intracranial space occupying lesion to relieve pressure and for a better prognosis, which is not questionable. However, we feel in case of CAPNON, wait and watch protocol can be used to a better effect with radiological and clinical follow-up. Above all surgical excision was primarily done due to imaging confusion over CAPNON and this article comes up with few key findings to clinch the radiological diagnosis of CAPNON.

Keywords: Calcifying pseudo neoplasm of neuraxis, cavernoma, epilepsy, pseudoneoplasm

INTRODUCTION

Calcifying pseudoneoplasm of the neuraxis (CAPNON) is a rare entity with only 90 cases being reported in literature till date. Among the reported 90 cases, 54 (60%) are intracranial and 36 (40%) are intraspinal lesions.^[1] It was first identified by Miller and erroneously reported as fibro-osseous lesion in 1922. It has been synonymously termed as brain stones, fibro-osseous lesions, and calcifying pseudo tumors.^[2] In 1978, it was reported as a distinct entity by Rhodes.^[2-4] If intracranial, it can be either intra or extra-axial in location.^[2,5] This entity mimics many calcifying intra-axial lesions like ganglioglioma, oligodendroglioma, cavernous malformation, and infection like tuberculosis. Hence, CAPNON should always be considered in the list of differential diagnosis for calcifying intra-axial lesions.^[6] Few patients have presented with hallucinations and partial seizures. Here, we illustrate a case presenting with intractable temporal lobe epilepsy and eventually diagnosed as CAPNON on imaging which was conservatively managed with a good clinical outcome.

CASE HISTORY

A male patient came to emergency room with five episodes of seizures since morning involving the right upper and lower

limbs with secondary generalization. He was a known case of seizure disorder for the past 5 years and on anti-epileptic medication which includes eptoin and valproate. Based on clinical symptoms, the patient was referred for CT examination.

Non-contrast CT scan revealed multiple thick amorphous calcifications involving the right gangliocapsular region involving the genu and posterior limb of internal capsule, lentiform nucleus, right hippocampus, and right peduncle of midbrain [Figure 1a and b]. The patient was further evaluated with MRI with intravenous Gadolinium

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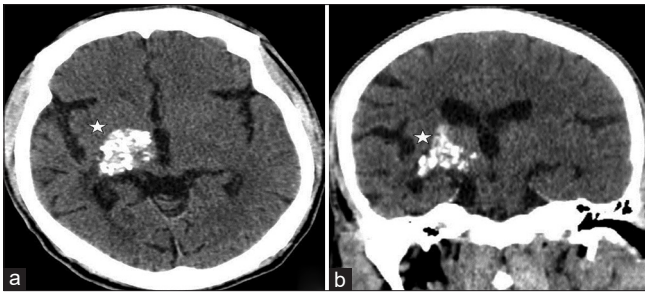


Figure 1: A man with seizures, (a) axial and (b) coronal sections of CT images shows multiple chunky parenchymal calcifications (star) in the right gangliocapsular region involving the genu and posterior limb of internal capsule, lentiform nucleus, right hippocampus, and right peduncle of midbrain

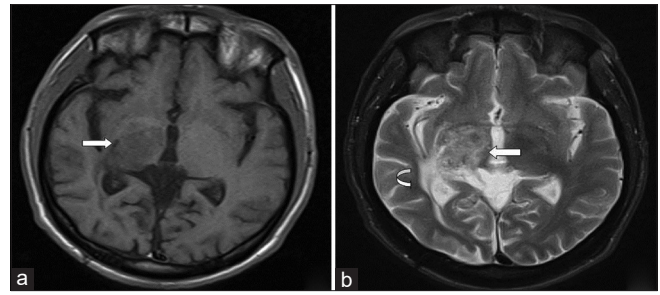


Figure 2: A man with seizures, (a) T1W and (b) T2W MR images shows a T1 hypointense (right arrow) and a T2 hyperintense (left arrow) lesion in the right thalamocapsular region with mild perilesional surrounding edema (right curved arrow)

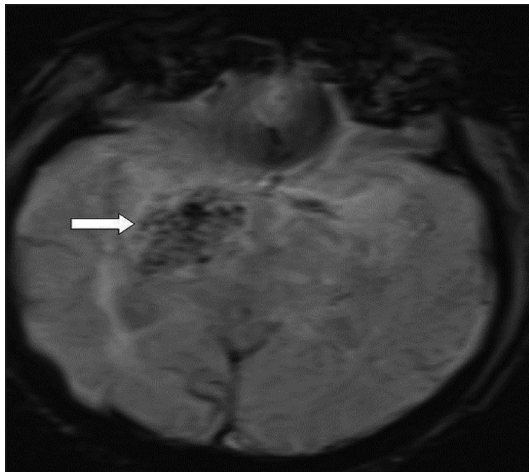


Figure 3: A man with seizures, gradient echo image shows multiple foci of blooming within the lesion (right arrow), suggestive of calcification

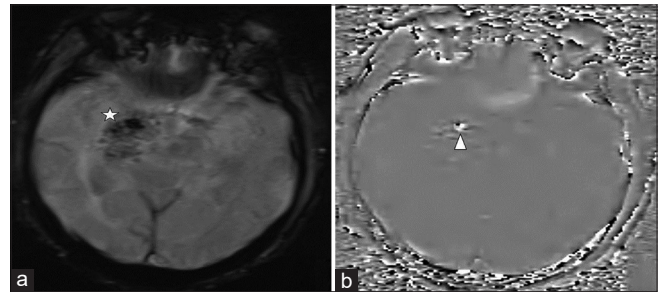


Figure 4: A man with seizures, Magnitude (a) and Phase (b) sequences of SWI images shows the lesion with blooming (star) in magnitude sequence and bright spot (Arrow head) in phase sequence, thus denoting areas of calcifications within the lesion

contrast. The MRI scan revealed an ill-defined intra-axial predominantly T2 hyperintense/T1 hypointense lesion involving the right thalamo capsular region, right medial temporal lobe, and the right peduncle of midbrain [Figure 2a and b]. Multiple discrete areas of blooming noted within the lesion on GRE [Figure 3] with magnitude and phase sequences of SWI showed features suggestive of calcification [Figure 4a and b]. Post-contrast T1-weighted axial and sagittal images shows minimal ill-defined discrete enhancement [Figure 5a and b].

Based on the imaging findings, a diagnosis of CAPNON was made and the patient was advised to continue the anti-epileptic drugs with regular follow-up. On follow-up, EEG was done and found normal. The patient was symptomatically better now. Hence, surgical plan was withheld.

DISCUSSION

CAPNON is a rare tumor of the central nervous system. A hypothesis has been suggested that CAPNON may develop as a healing response to a wide range of eliciting factors, which could explain the variations in histopathologic features. Even though the histopathologic features of CAPNON are not

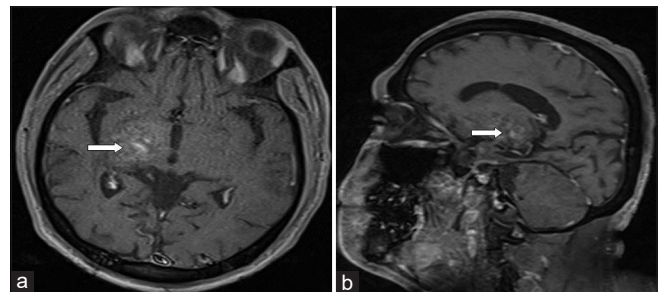


Figure 5: A man with seizures, T1 post-contrast (a) axial and (b) sagittal MR images shows the lesion with areas of minimal enhancement upon contrast administration (right arrow)

clearly understood, a reactive process over a hamartomatous process has been favored.^[2,7,8] Interestingly, this reactive process involved in the calcifying pseudoneoplasm is not only constrained to the neuraxis. It can also occur at different other sites, including pleura, breast, and mediastinum.^[9]

No sex or age predominance has been mentioned in most of the published articles, however the lesions can occur at any age between 6 and 83 years.^[2,6,8] The most common symptoms in intracranial lesions are headache and seizure with only few cases being discovered incidentally.^[2,8] Symptoms, when present, are variable and are related to local mass effect, rather than invasive growth. Seizure was the commonest symptom in supratentorial CAPNON (17/36 cases; 47.2%). The seizure types were generalized tonic-clonic seizure (GTCS) in 7 cases (41.1%), focal

impaired awareness seizure (FIAS) in 4 cases (23.5%), focal aware seizure (FAS) in 2 cases (11.8%), and not described in 4 cases.^[1] The lesions are typically sporadic, with few exceptions that have occurred in association with meningoangiomatosis in patients with type 2 neurofibromatosis^[9,10] and low-grade glioma.^[10]

On CT, generally CAPNON shows an area of solid calcification while on MR imaging the lesions appear hypointense on T1- and T2-weighted sequences.^[8] Post-contrast the lesions shows minimal linear internal or rim enhancement.^[8] Contrary in our case, the lesion showed areas of T2 hyperintensity, probably because of the presence of chondromyxoid matrix within the areas of calcifications. Perilesional vasogenic edema is not commonly reported and is unexpected in benign lesion.^[8] Grabowski *et al.*, in his study, reported vasogenic edema could be because of seizures rather than CAPNON.^[8] Similarly, in our case, perilesional edema was present secondary to seizures rather than CAPNON as proposed by Grabowski *et al.*

The typical histopathological features of CAPNON are (1) presence of palisading spindle to epithelioid cells; (2) presence of chondromyxoid matrix in a nodular pattern; (3) amorphous calcification, psammoma bodies, and osseous metaplasia; (4) foreign-body reaction with giant cells surrounding the calcified region; (5) fibroblastic proliferation.^[2,3,11] These pathological presentations are not evident in each case and some lesions may not show all of the above features.^[2,11] In our case, the patient was conservatively managed with anti-epileptic medications and is on regular follow-up. Earlier treatment has prevented the patient suffering from future complications like deterioration of memory.

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Conflicts of interest

There are no conflicts of interest.

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