Psammomatous Cavernous Malformation Presenting as Drug-Resistant Epilepsy: Case Illustration and Review of Literature

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Key words
- Cavernoma
- Drug-resistant epilepsy
- Electrocorticography
- Psammoma
- Vascular malformation

Abbreviations and Acronyms
AED: Antiepileptic drug
ECoG: Electrocorticography
EEG: Electroencephalography
MR: Magnetic resonance
PB: Psammoma body
QoL: Quality of life

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BACKGROUND: **Psammoma bodies (PBs) are whorled, laminated hyaline spherules containing calcium deposits. Intracranially, the presence of PBs is associated with variants of meningioma and pituitary lesions, as well as aging choroid plexus. Limited information exists on their presence in vascular malformation.**

RESULTS: **In this report, we describe a case of an adolescent male with drug-resistant epilepsy that was surgically managed at our regional epilepsy center. The epileptogenic focus was determined to be emanating from an indolent right insular lesion. Histopathologic evaluation showed the abundance of intravascular and perivascular PBs. Immunohistochemical evaluation confirmed the vascular origin using vascular markers. The unusual presence of PBs in a vascular lesion was unanticipated.**

CONCLUSIONS: **Based on our case, we present the clinicoradiologic characteristics, supplemented with intraoperative findings, for this unusual lesion. In addition, because of the unusual presence of PBs in vascular lesions, we provide the findings of a systematic literature review to show the association of PBs with intracranial vascular lesions.**

INTRODUCTION

Psammoma bodies (PBs) are whorled, well-circumscribed, laminated structures containing calcium deposits. The presence of PBs is strongly linked to neoplastic and nonneoplastic lesions, including, but not limited to, the thyroid, ovary, cervix, uterus, kidney, pancreas, and pleura.[1] Intracranial PBs are associated with benign tumors such as meningiomas,[2,3] pituitary lesions,[4] and aging choroid plexus.[5] Limited literature exists on the association of PBs with intracranial vascular lesions.[6,7]

In this report, we present a case of an adolescent boy with drug-resistant epilepsy attributable to an insular cavernous malformation. Histopathologic evaluation of the resected mass showed the unusual presence of PBs. With limited literature documenting association of PBs with intracranial vascular malformation, our case serves to revisit this rare association and describe clinicopathologic correlates and radiologic and intraoperative findings of this unusual disease. In addition, we provide the results of a systematic review of the literature on intracranial vascular diseases associated with the presence of PB.

METHODS

A systematic review of the literature for relevant, peer-reviewed articles up to 15 March 2016 using the electronic databases MEDLINE and Google Scholar was performed. Boolean operators, "AND" and "OR" using the search terms "psammoma", "vascular", "malformation", "hemangioma," and "cavernous" were used in various combinations to narrow the scope of the review. The resulting citations were examined in their entirety by 2 authors (K.S. and P.K) independently. To filter relevant articles, retrieved articles were screened by reviewing each article title, abstract, or full texts as available. Bibliographies of identified publications and articles citing them were also scrutinized. Criteria implemented for screening included 1) articles documenting the presence of PB within vascular malformations; 2) vascular malformations in intracranial locations; and 3) articles limited to humans. Exclusion criteria applied to article selection included articles documenting the presence of PBs in extracranial diseases and those not in English. Articles with intracranial association of PBs with tumors (meningioma and pituitary lesions), those describing aging choroid plexus, and other diseases not related to intracranial vascular lesions were also excluded. Any potential conflict arising from article selection was mitigated by discussion and mutual consensus. An algorithm of the literature search strategy is shown in Figure 1. Data from identifiable articles were synthesized. This included extraction of

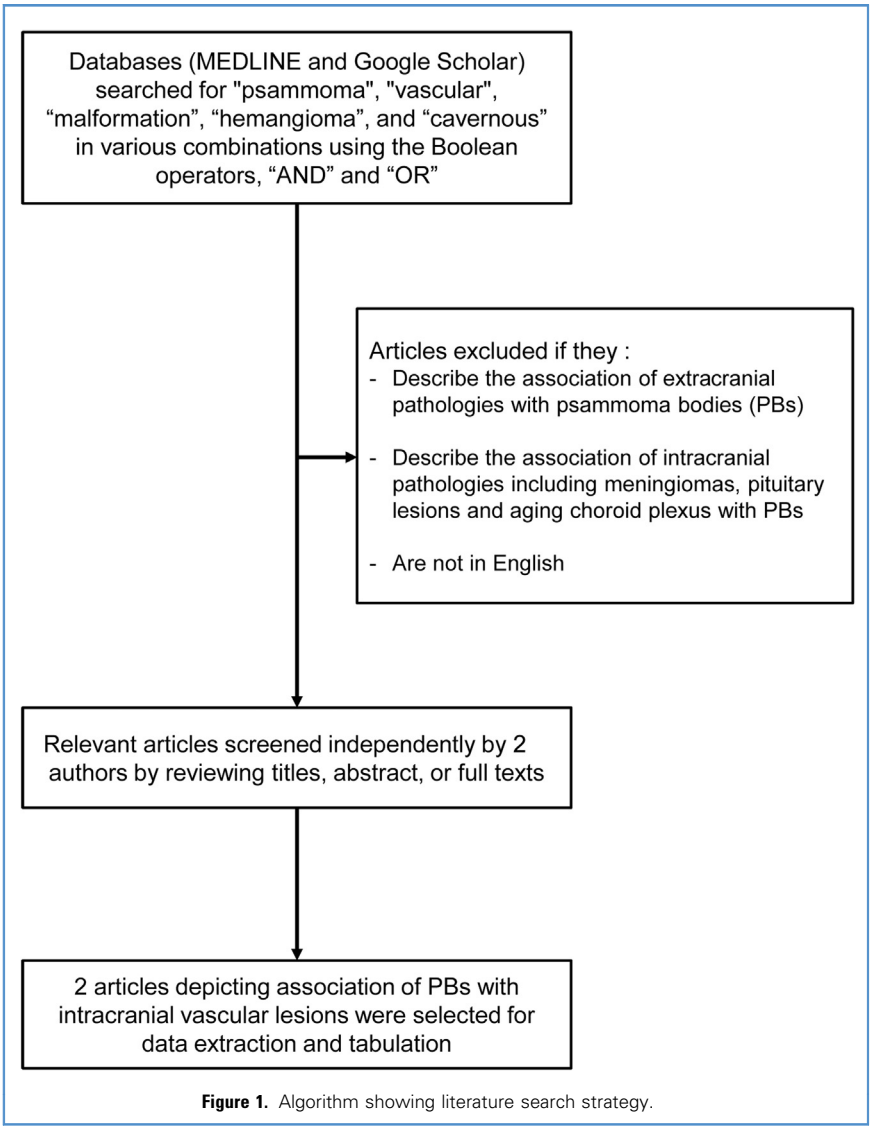

Figure 1. Algorithm showing literature search strategy.

patient characteristics including age, gender, clinical presentation, vascular pathology, radiologic and histologic findings, management, and outcome. Our review is summarized in Table 1.[6,7]

CASE ILLUSTRATION

History and Examination

A 15-year-old African American, right-hand-dominant boy presented to our regional epilepsy center at the Louisiana State University, Shreveport, Louisiana, USA, for evaluation and management of drug-resistant epilepsy. At age 12 years (2011), the patient was alleged to have experienced his first seizure episode while playing video games at home. The seizure semiology was suggestive of a complex-partial type, starting with a brief aura lasting less than a minute with impaired consciousness. The patient was unresponsive to verbal or tactile stimuli and experienced head shaking while staring at the ceiling, with unilateral blinking of his right eye and fixed left eye glare. The episode lasted over 5 minutes, followed by loss of postural tone. Postictally, the patient reported tiredness and confusion. No facial grimacing, tongue biting, upward eye rolling, fever, or jerky limb movements were noted. The seizure episode recurred 2 days later with similar presentation, after which the patient was evaluated by a neurologist. Past medical history was unremarkable except for occasional migraine headaches. No documented delay in his developmental history or milestones was noted. Family history was negative for seizure disorders. Video electroencephalographic (EEG) studies at that time determined the epileptogenic focus to be emanating from the anterior and central region of the right temporal lobe. Frequent high-amplitude spikes were seen over the right frontotemporal areas, maximal over F8 and T4. These epileptiform abnormalities had a broad field and spread to T4 and T6 and occasionally to C4 and P4. A contemporary computed tomographic scan of the head showed an area of calcification, measuring 8 mm in maximal dimension in the insular region, adjacent to the sylvian fissure (Figure 1A). The spherical lesion was confirmed on magnetic resonance (MR) imaging (Figure 1B and C), and was contrast enhancing (Figure 1D). However, MR angiography proved inconclusive to distinguish if the lesion represented an aneurysm or a vascular malformation. At this point, medical management with oxcarbazepine 300 mg orally twice a day was initiated for seizure control; observation of the insular lesion with serial imaging on follow-up was deemed prudent.

Over the course of the next 3 years (2012–2014) since the onset, the patient was prescribed antiepileptic drugs (AEDs) in augmented dosages to curtail intermittent seizure episodes that initially averaged 5 or 6 times quarterly (2012) and progressed to every 2–3 days (2014). The seizure progression necessitated frequent emergency room visits with subsequent hospitalizations, negatively affecting quality of life (QoL) and predisposing to missed school days. No signs of cognitive decline or poor academic performance were noted. Potentially recognizable precipitating factors for seizure induction included stimulus overload (video games) and stress. Follow-up computed tomography (mid-2012) at 1 year since the first seizure depicted the presence of preexisting calcification, albeit with slight increment in size to 10 mm (Figure 1E). By mid-2014, the AED regimen comprised levetiracetam 1000 mg twice a day, extended release oxcarbazepine 2100 mg/day, and