The Role of MRI in Diagnosing Calcifying Pseudoneoplasms of the Neuraxis (CAPNON): A Rare Case Report of CAPNON with Corpus Callosum Agenesis and a Lipoma

Jeton Shatri[1,2], Kreshnike Dedushi Hoti[2], Ardita Kafexholli[1], Jeton Kafexholli[3], Veton Kurti[2], Diar Kabashi1, Dijon Musliu[1]*

[1]*Institute of Anatomy, Faculty of Medicine, University of Prishtina, Kosovo*

[2]*Clinic of Radiology and Nuclear Medicine, University and Clinical Center of Kosovo*

***Correspondence:** Dijon Musliu, MD, PhDC, Institute of Anatomy, Faculty of Medicine, University of Prishtina, Kosovo

: dijon.musliu@uni-pr.edu

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AUTHORS' CONTRIBUTIONS
Jeton Shatri – radiologic scanning and evaluation, writing case report
Kreshinke Dedushi Hoti – radiologic scanning and evaluation, writing case report
Ardita Kafexholli – Discussion writing
Veton Kurti – data analysis, writing case report
Dijon Musliu – reviewing, submitting

DISCLOSURES
The authors declare no financial, personal, or competing interests that could influence the work reported in this manuscript.

CONSENT
Yes.

HUMAN AND ANIMAL RIGHTS
This study was conducted in compliance with the ethical standards of the institutional and national committees on human experimentation and in accordance with the Helsinki Declaration of 1975, as revised in 2000.

ETHICAL STATEMENT
This study was conducted in accordance with the ethical standards of the institutional and national research committees and with the 1975 Helsinki Declaration, as revised in 2000. Written informed consent was obtained from the patient for the publication of this case report. No experiments were conducted on animals in this study.

ABSTRACT

Calcifying pseudoneoplasms of the neuraxis (CAPNON) are rare, benign lesions of the central nervous system, often misdiagnosed due to their nonspecific clinical and radiological presentations. We report a unique case of a 59-year-old male with persistent headaches, found to have a calcified mass in the left cerebral hemisphere associated with corpus callosum agenesis and an interhemispheric lipoma. MRI revealed a hypointense lesion with surrounding edema and compressive effects, complemented by CT findings of pronounced calcifications. The coexistence of CAPNON with corpus callosum agenesis and a lipoma is exceedingly rare, with few similar cases documented. This report underscores the critical role of MRI in diagnosing CAPNON and highlights its potential to mimic other central nervous system pathologies. Early recognition is essential to avoid misdiagnosis and guide appropriate management.

CASE REPORT

BACKGROUND

Calcifying pseudoneoplasms of the neuraxis (CAPNON) are rare, non-neoplastic, fibro-osseous lesions of the central nervous system (CNS) that can mimic a variety of neoplastic and infectious conditions. While their exact pathogenesis remains unclear, they are believed to result from chronic inflammation, prior trauma, or an abnormal reparative process. CAPNON lesions are often heavily calcified and can occur in both intra-axial and extra-axial locations, making their differentiation from tumors such as meningiomas, oligodendrogliomas, and chondrosarcomas crucial.

The significance of this case lies in its unusual presentation, as CAPNON is observed alongside corpus callosum agenesis and an interhemispheric lipoma—an exceedingly rare combination. To our knowledge, only one similar case has been reported in the literature. This report expands the clinical and radiological spectrum of CAPNON and reinforces the importance of recognizing its distinct imaging characteristics on MRI and CT. By accurately identifying CAPNON, unnecessary surgical interventions and misdiagnoses can be avoided. This case contributes to the growing body of knowledge on CAPNON, highlighting its potential coexistence with other congenital anomalies and emphasizing the need for a multidisciplinary diagnostic approach in complex neuroimaging findings.

CASE PRESENTATION

A 59-year-old male patient presents with a six-month history of persistent headaches, occasionally accompanied by dizziness. Neurological examinations reveal no deficits. The patient is referred for a cranial CT scan, which reveals a calcified mass approximately 53x25 mm in the left cerebral hemisphere, associated with an interhemispheric lipoma and agenesis of the corpus callosum. Subsequently, the patient undergoes MRI with intravenous contrast.

The MRI shows a supratentorial, irregular formation located in the parasagittal white matter of the left hemisphere, measuring around 52x25 mm (AP x LL). On non-contrast images, the mass appears hypointense, while post-contrast sequences reveal heterogeneous contrast enhancement. The described mass appears as a homogenous hyperdensity on the CT scan. It is associated with surrounding edema and pronounced compressive effects.

Additionally, an interhemispheric, well-circumscribed formation is observed in the region of the corpus callosum and splenium on the left side, extending into the left choroid plexus, measuring approximately 33x16x25 mm (AP x LL x CC). This formation is hyperintense on T1 and T2-weighted MRI sequences and FLAIR, without contrast enhancement, suggesting a lipoma. Furthermore, there are supratentorial lesions with hyperintensity on FLAIR and T2 sequences without restricted diffusion on DWI and ADC, localized in the cerebral white matter, likely representing gliotic lesions within the context of advanced cerebral vascular encephalopathy.

The imaging findings from the native and contrast-enhanced brain MRI, complemented by CT, are suggestive of a **Calcified Pseudoneoplasm of the Neuroaxis (CAPNON)**, accompanied by corpus callosum agenesis and an interhemispheric lipoma. Figure 1 and 2 shows the MRI and CT imagery findings and description of these findings.

DISCUSSION

Calcifying pseudoneoplasms of the neuraxis (CAPNON) are rare, benign lesions of the central nervous system (CNS), thought to develop from past trauma, infections, or inflammatory conditions [1,2]. Also known as fibro-osseous lesions, these slow-growing, solitary masses predominantly appear in the supratentorial compartment but can also occur intra-axially. Due to their distinctive histopathological features, CAPNONs were likely misdiagnosed before their identification in recent decades [3,4].

The clinical presentation of CAPNON is variable and depends on lesion size and location, with symptoms ranging from headaches to seizures and motor deficits. Some cases are asymptomatic and discovered incidentally during imaging. Radiologically, CAPNONs present as heavily calcified meningeal (extra-axial) or parenchymal (intra-axial) pseudo-masses, showing low signal intensity on T1 and T2-weighted images and dropout on gradient sequences (T2* and SWI). Unlike tumors such as chondrosarcoma, which have fine stippled calcifications, CAPNONs exhibit bulky, amorphous calcifications. Surgical resection of symptomatic lesions is typically curative, with rare recurrence and no malignant potential [1,5,6].

Although CAPNONs are rare, with only about 100-150 cases reported, recognizing them is crucial to avoid misdiagnosis as more common brain tumors [1,5]. Their exact cause remains unknown, but they are believed to result from a reaction to trauma, infection, inflammation, or neoplasms, inferred from their benign behavior and histopathological characteristics [1,2,7].

CAPNONs can occur in various CNS locations, including the spine and skull base. They are most often extra-axial but can also be intra-axial. CAPNONs show no predilection for sex or age and are sometimes found near other CNS lesion [1,7,8]. The histogenesis of CAPNON remains uncertain, possibly representing an unusual form of tumoral calcinosis or a unique healing response. Histologically, CAPNONs are characterized by a fibrillary chondromyxoid matrix, surrounded by spindle to epithelioid cells, osseous metaplasia, psammoma bodies, fibrous stroma, and giant cells [6,9,10].

In this case report, we present a rare instance of Calcifying pseudoneoplasms of the neuraxis (CAPNON) coexisting with corpus callosum agenesis and a lipoma. Notably, only one similar case has been previously documented in the literature, described by Inukai et al., where calcifications and a lipoma were identified surrounding the corpus callosum through various imaging modalities, mirroring our findings [11]. CAPNON is an infrequently encountered benign tumor that predominantly affects males and manifests across a broad age range from 2 to 90 years. The clinical presentation of CAPNON is often marked by non-specific symptoms such as headaches, seizures, and focal neurological disturbances [1,7,11].

The pathophysiological basis in our case likely involves an inflammatory response initiated by the lipoma. This inflammatory cascade is hypothesized to trigger macrophage infiltration, M2 macrophage activation, angiogenesis, and cell proliferation, eventually leading to the calcification processes that typify CAPNON. Such inflammatory mechanisms have

been implicated in similar neuro-pathological conditions, suggesting a consistent pattern across different cases [11].

The literature elucidates various manifestations of CAPNON, further emphasizing its diagnostic complexity. Reports have documented cases associated with acute hydrocephalus, progressive cranial nerve palsy, and signs of brainstem compression, all consistently showing perilesional edema in preoperative MRI scans [12]. A particularly intriguing case by Li et al. describes a misdiagnosis of oligodendroglioma due to the lesion's pronounced calcification and strong enhancement on MRI, which was later histopathologically confirmed as CAPNON, thus highlighting the critical role of comprehensive tissue analysis in establishing the correct diagnosis [13].

Additional reports from Greco et al. and Gamblin et al. explore the differential diagnostic considerations prompted by the radiological ambiguities of CAPNON, ranging from ependymoma to schwannoma and metastasis, indicating the broad spectrum of radiological features CAPNON can mimic [1,14]. Furthermore, the challenges in diagnosing such lesions were exemplified by Yangi et al., where contrasting diagnoses were proposed by different centers, reflecting the necessity for meticulous radiological assessment and possibly, multi-center review when encountering unusual or complex cerebral calcifications [15].

Magnetic Resonance Imaging (MRI) plays a pivotal role in the diagnosis of Calcifying pseudoneoplasms of the neuraxis (CAPNON), as highlighted by multiple case studies in the literature [1,7,11,12,15]. In cases of CAPNON, MRI can reveal varied and distinctive radiological features that are critical for accurate diagnosis. For instance, calcified masses typically appear as low-intensity on T1-weighted images (T1WI) and T2-weighted images (T2WI), while associated lipomas present as high-intensity on both sequences due to their fat content. Notably, gadolinium enhancement can delineate the periphery and interior of calcifications, suggesting active pathological processes, such as inflammation or vascular proliferation [11]. Moreover, MRI can detect extensive vasogenic edema in surrounding white matter, indicative of the tumor's impact on adjacent brain tissues. In cases with ambiguous or complex presentations, MRI serves as an indispensable tool for differential diagnosis, ruling out other potential pathologies such as oligodendroglioma, ependymoma, or schwannoma, which may present with similar imaging characteristics [7]. Thus, the comprehensive imaging capabilities of MRI are essential for not only identifying CAPNON but also for assessing its extent, evaluating associated anomalies, and guiding therapeutic strategies

Our contribution to the literature through this case report is significant as it underscores a unique combination of CAPNON with corpus callosum agenesis and a lipoma, expanding the clinical and radiological spectrum of this rare condition. This case serves as an important reminder of the varied presentations of CAPNON and the essential role of a multidisciplinary approach in its diagnosis and management, encouraging ongoing vigilance and collaboration in the neurology and neurosurgery communities.

TEACHING POINT

Calcifying pseudoneoplasms of the neuraxis (CAPNON) are rare, benign lesions of the central nervous system characterized by extensive calcifications and hypointensity on T2-weighted MRI with signal dropout on gradient echo sequences. Recognizing these imaging features is crucial to differentiate CAPNON from neoplastic or infectious pathologies, preventing unnecessary interventions.

QUESTIONS

1. What is the most characteristic imaging finding of Calcifying Pseudoneoplasms of the Neuraxis (CAPNON) on MRI?

A) High signal intensity on T2-weighted imaging

B) Peripheral ring enhancement with gadolinium

C) Low signal intensity on T2-weighted imaging with blooming artifact on susceptibility-weighted imaging (applies)

D) Diffusion restriction on DWI

E) Hyperintensity on FLAIR with cystic components

Explanation: CAPNON presents as a calcified mass that appears hypointense on T2-weighted MRI and demonstrates signal dropout on gradient echo or susceptibility-weighted imaging due to its mineralized nature. Unlike tumors or infections, it typically lacks significant enhancement or diffusion restriction.

2. Which differential diagnoses should be considered when encountering a CAPNON lesion on imaging?

A) Meningioma (applies)

B) Oligodendroglioma (applies)

C) Chondrosarcoma (applies)

D) Lipoma

E) Glioblastoma

Explanation: CAPNON can mimic meningiomas due to its extra-axial location and calcifications. Oligodendrogliomas and chondrosarcomas may also present with calcifications but exhibit different imaging characteristics, such as a more heterogeneous enhancement pattern. Lipomas do not show calcifications, and glioblastomas typically exhibit necrosis and peritumoral edema.

3. What is the most likely etiology of CAPNON?

A) Genetic mutations in CNS tumor suppressor genes

B) Long-term inflammation, trauma, or previous infections (applies)

C) Primary vascular malformation

D) Congenital malformation associated with corpus callosum agenesis

E) Metastatic calcification

Explanation: CAPNON is considered a reactive lesion rather than a neoplasm, arising from chronic inflammation,

trauma, or infection, leading to fibro-osseous metaplasia. It is not associated with congenital malformations or metastases.

4. What is the recommended management approach for CAPNON?

A) Immediate surgical resection in all cases

B) Chemotherapy and radiotherapy

C) Observation in asymptomatic cases and surgical removal if symptomatic (applies)

D) Corticosteroid therapy

• E) Antibiotic treatment

Explanation: CAPNON is benign and does not require treatment unless symptomatic. Surgery is considered only if the lesion causes mass effect or neurological symptoms. It does not respond to chemotherapy, radiation, or pharmacological therapies.

5. What imaging feature differentiates CAPNON from oligodendroglioma?

• A) Presence of hemorrhage

• B) Enhancement on contrast-enhanced MRI

• C) Hyperintensity on T1-weighted imaging

• D) Bulk amorphous calcifications rather than fine calcifications (applies)

• E) Presence of cystic components

Explanation: Oligodendrogliomas typically show fine, stippled calcifications, whereas CAPNON presents with large, amorphous calcifications. Additionally, oligodendrogliomas may enhance and have cystic or necrotic components, which are not features of CAPNON.

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FIGURES

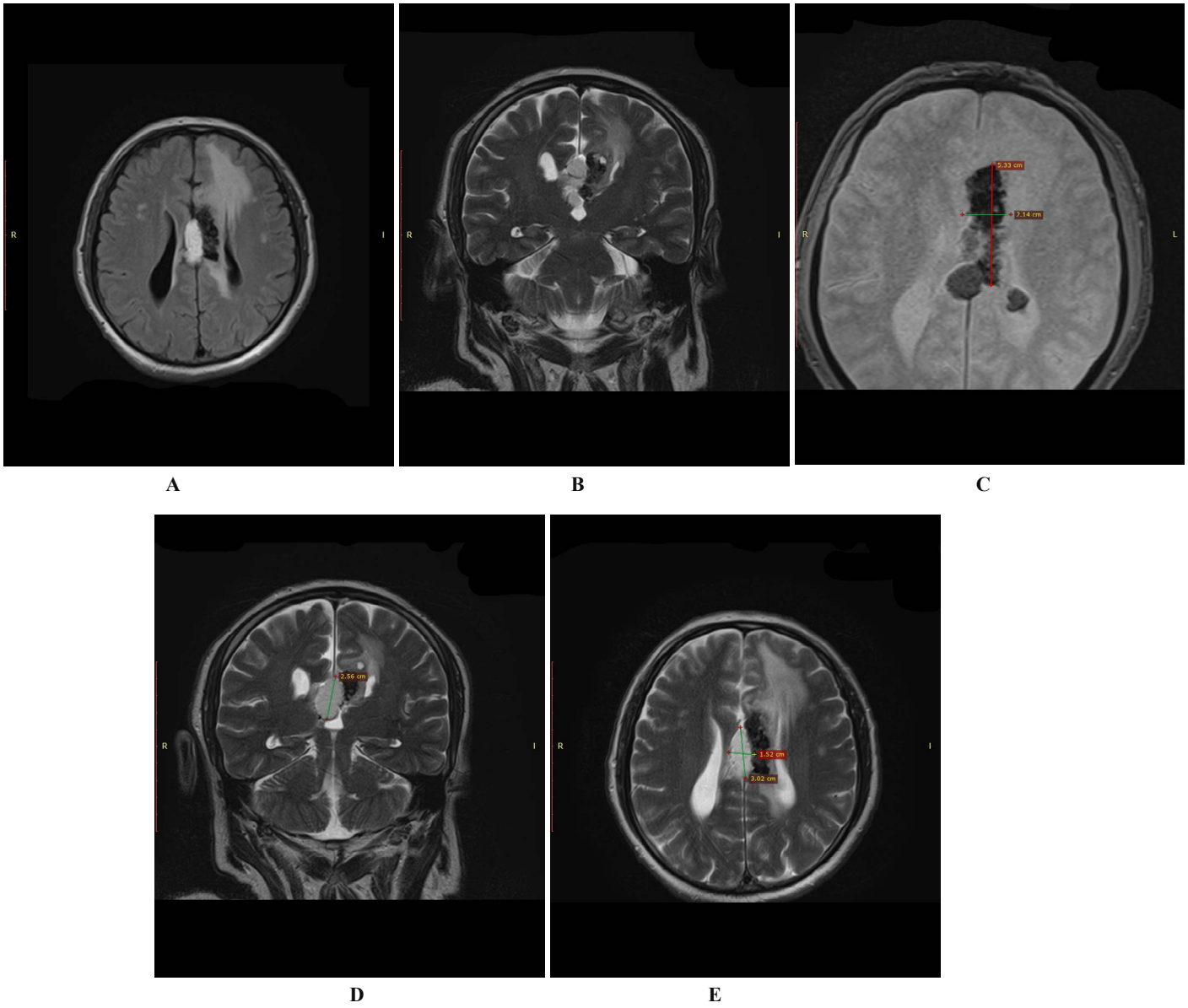
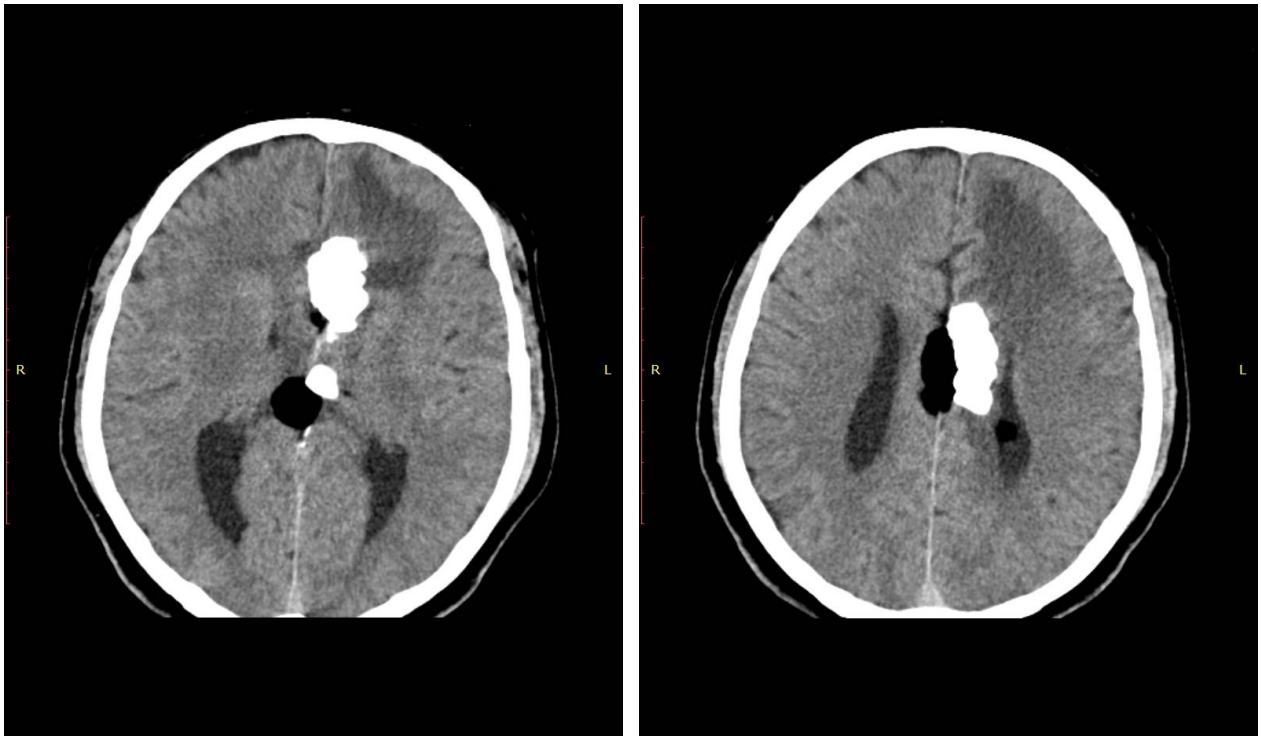


Figure 1: The MRI reveals a multilobulated, calcified mass in the left frontal lobe, encircled by edema. **A)** Axial Fluid Attenuated Inversion Recovery (FLAIR) sequence; **B)** Coronal T2-weighted Magnetic Resonance Imaging (MRI) sequence; **C)** Axial Gradient Echo sequence; **D)** Coronal T2-weighted sequence displaying an interhemispheric lipoma. **E)** Axial T2-weighted MRI demonstrating a multilobulated lesion in the left frontal lobe with central hypointense foci suggestive of calcification, measuring approximately 3.02 × 1.52 cm. The mass is associated with surrounding hyperintense signal indicating vasogenic edema and exerts mild mass effect on the adjacent frontal horn of the lateral ventricle.



A **B**

Figure 2: CT imaging presentation. A-B) Corpus callosum agenesis together with a tubulo-nodular lipoma

KEYWORDS

Calcifying pseudoneoplasm; CAPNON; brain calcifications; fibro-osseous lesion; CNS tumors

ABBREVIATIONS

CAPNON = CALCIFYING PSEUDONEOPLASM OF THE NEURAXIS
MRI = MAGNETIC RESONANCE IMAGING
CT = COMPUTED TOMOGRAPHY
FLAIR = FLUID-ATTENUATED INVERSION RECOVERY
DWI = DIFFUSION-WEIGHTED IMAGING
ADC = APPARENT DIFFUSION COEFFICIENT
SWI = SUSCEPTIBILITY-WEIGHTED IMAGING