

# Calcifying Pseudoneoplasms of the Neural Axis. Report of Three Cases

## Kalcifikující pseudoneoplazma nervového systému – tři kazuistiky

### Abstract

Calcifying pseudoneoplasms of the neural axis (CAPNONA) are rare lesions that have been reported in both intra/extra-axial locations intracranially, and in extra-axial locations in the spinal region. Central loss of signal on T1/T2-weighted images with enhancement following contrast administration is most commonly observed on magnetic resonance imaging, however, the histopathological composition of CAPNONA is diverse which may be reflected in the imaging findings. We present three cases of CAPNONA detected by MRI and confirmed by histological evaluation, and the largest review of the MRI characteristics of these lesions to date.

### Souhrn

Kalcifikující pseudoneoplazmata nervového systému (CAPNONA) jsou vzácné léze, které byly popsány v intra- i extraaxiálních lokalizacích intrakraniálně a v extraaxiálních lokalizacích ve spinální oblasti. Charakterickým znakem na magnetické rezonanci je centrální výpadek signálu na T1 a T2 vážených obrazech s postkontrastním zvýrazněním po aplikaci kontrastní látky. Nicméně histologická skladba může být různorodá, což se v zobrazení může odrazit. Předkládáme tři kazuistiky CAPNONA, které byly detekovány pomocí MR a potvrzeny histologicky. Kazuistiky jsou doplněny rozsáhlým přehledem literatury s MR charakteristikami těchto lézí, jak byly dosud popsány.

A. M. Rulseh<sup>1,2</sup>, J. Keller<sup>1,3</sup>,  
J. Klener<sup>4</sup>, J. Šroubek<sup>4</sup>,  
M. Syrůček<sup>5</sup>, I. Zemanová<sup>5</sup>,  
V. Dbalý<sup>4</sup>, J. Vymazal<sup>1,6</sup>

<sup>1</sup> Department of Radiology, Na Homolce Hospital, Prague, Czech Republic

<sup>2</sup> Department of Radiology, First Faculty of Medicine, Charles University, Prague, Czech Republic

<sup>3</sup> Department of Neurology, Third Faculty of Medicine, Charles University, Prague, Czech Republic

<sup>4</sup> Department of Neurosurgery, Na Homolce Hospital, Prague, Czech Republic

<sup>5</sup> Department of Pathology, Na Homolce Hospital, Prague, Czech Republic

<sup>6</sup> Department of Neurology, First Faculty of Medicine, Charles University, Prague, Czech Republic



**Prof. Josef Vymazal, M.D., D.Sc.**  
Na Homolce Hospital  
Roentgenova 2  
15030 Prague 5  
e-mail: Josef.Vymazal@homolka.cz

Accepted for review: 31. 12. 2010  
Accepted for print: 4. 4. 2011

### Key words

calcifying pseudoneoplasm – CAPNONA –  
fibro-osseous lesion – calcification

### Klíčová slova

kalcifikující pseudoneoplazma –  
CAPNONA – fibro-oseozní léze –  
kalcifikace

### Support

This work was supported by grant NS 9654-4 from the Ministry of Health of the Czech Republic and by Ministry of Education Research Programs MSM 0021620849 and MSM 0021620816.

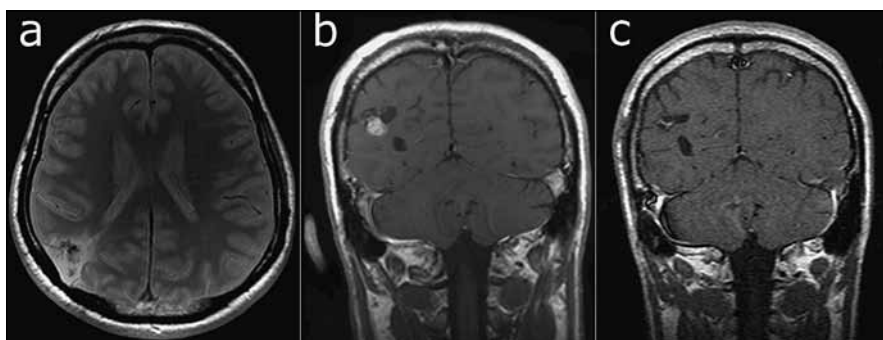


Fig. 1a. T2-weighted TSE sequence. A hypointense nodule is visible within a wedge-shaped lesion in the angular region.

Fig. 1b. Post-contrast T1-weighted SE sequence shows an area of nodular enhancement attached to a hypointense lesion.

Fig. 1c. Post-contrast T1-weighted SE sequence six months after resection of the lesion. Minimal enhancement was detected in the resected region.

## Introduction

Calcifying pseudoneoplasms of the neural axis (CAPNONA), or fibro-osseous lesions as they are alternatively known, are rare, tumor-like lesions that have been reported to occur in both intra-axial (IA) and extra-axial (EA) locations intracranially, and in EA locations in the spinal region. Due to their rarity, CAPNONA are seldom considered in the differential diagnosis of atypical brain or spinal lesions. These lesions carry an excellent prognosis and are generally curable by surgical resection. We present three cases of CAPNONA detected with magnetic reso-

nance imaging (MRI) and confirmed by histological evaluation.

## Case Reports

### Case 1

A 46-year-old male with a three-year history of headache suffered a skull fracture at the age of eleven in the right parietal region following a blow to the head with a sharp metal object. Current neurological examination did not reveal any abnormalities. MRI at 1.5T showed a wedge-shaped lesion in the right angular region with increased signal intensity and central hypointense areas on proton-density- (PD)

and T2-weighted images (Fig. 1a). The wedge-shaped region had significantly decreased signal intensity on T1-weighted images, suggestive of a pseudocystic lesion. Internal nodular enhancement was observed after contrast agent administration (Fig. 1b). Due to the ambiguous character of the clinically symptomatic lesion, microsurgery with navigation was recommended. During the surgery a bony fragment approximately 8 × 10 × 20 mm was observed as the plate of the skull was lifted off. The fragment was adherent to and extended 20 mm into the brain parenchyma. Gross and intraoperative microscopic inspection of the MRI-enhancing nodule revealed a fibrous stroma filled with numerous small calcifications. Histological examination showed a chondromyxoid matrix in a nodular pattern, calcification and osseous metaplasia, scattered psammoma bodies, palisading spindle cells, foreign-body-like reaction with giant cells and a fibrous stroma (Fig. 2). The postoperative course of the patient was uneventful. The patient reported the cessation of his headaches and MRI six months after surgery demonstrated total removal of the lesion (Fig. 1c).

### Case 2

A 43-year-old female presented with recurrent pain in the lumbar region with radiation to both legs along the dorsal aspect. She had undergone surgery of the lumbar spine two years previously for isthmic spondylolisthesis and a herniated disc at L5/S1. MRI at 1.5T showed a small nodular extradural lesion at the level of L3. The lesion was hypointense on T1-, T2- and PD-weighted images (Fig. 3). No contrast agent was administered. During the microsurgery a small, calcified nodule approximately 5 mm in diameter was removed. The nodule was attached to one of the nerve roots accompanied by small tortuous vessels supplying a vascularized fibrous stroma. Histological examination revealed a calcified lesion consisting of primitive bone trabeculae and islets of choroid tissue in a moderately cellular matrix, scattered psammoma bodies and a fibrous stroma. The pathologist confirmed the diagnosis of CAPNONA. The postoperative course of the patient was uneventful. MRI ten months after the surgery showed no evidence of recurrence.

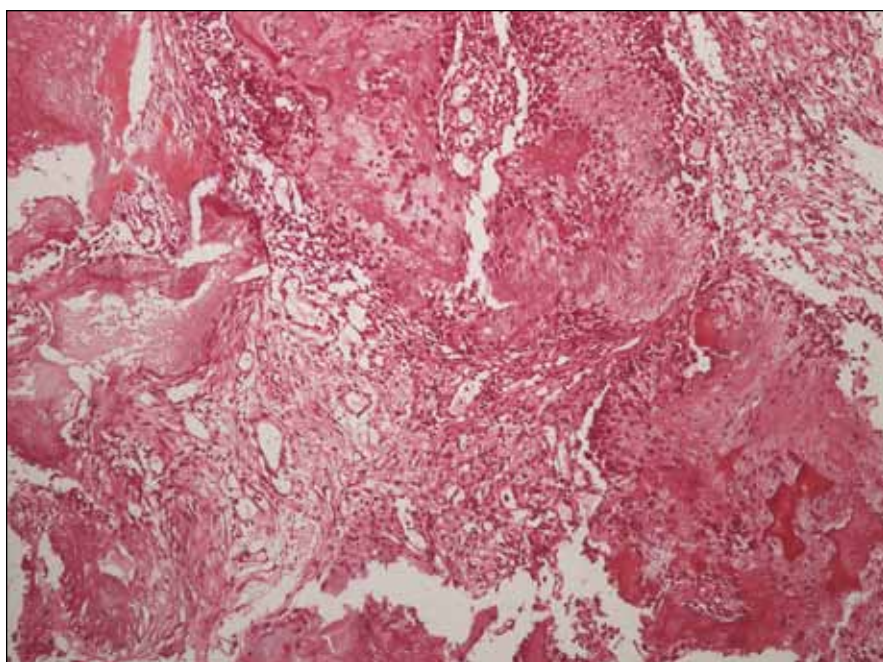


Fig. 2. Calcifying pseudoneoplasm of the neural axis: fibrillar tissue, amorphous stroma with partial calcification.



Fig. 3a. T1-weighted TSE sequence showing a small hypointense nodule at the level of L3 (arrow).

Fig. 3b. T2-weighted TSE sequence showing the same hypointense nodule (arrow).

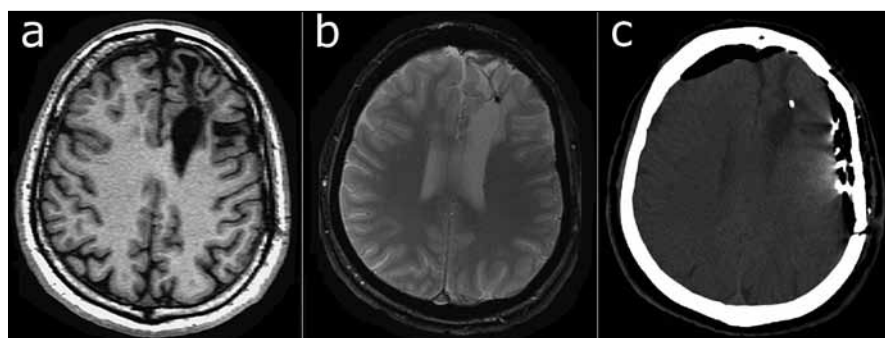


Fig. 4a. T1-weighted MP-RAGE sequence. A large post-surgical defect may be seen in the left frontal region.

Fig. 4b. T2\*-weighted FLASH sequence reveals a small hypointense lesion within the defect.

Fig. 4c. CT image acquired one day later, following application of subdural strip and grid electrodes. A calcified (500 HU) lesion in the left frontal region is visible. Strip and grid electrodes are visible on the left, and pneumocephalus may be seen anteriorly.

### Case 3

A 23-year-old male was referred due to refractory epilepsy. At ten years of age he suffered an episode of serous meningitis and developed epilepsy two years later. At the age of fifteen a pial AVM in the left frontal region was diagnosed and resected. Following the surgery the patient has been seizure-free for four years, after which time the seizures recurred. CT and MRI examination revealed a calcified lesion in the post-resection area with surrounding gliosis (Fig. 4). MRI was acquired at 1.5T and revealed a hypointense lesion on T1-, T2- and PD-weighted images. No contrast agent was administered. During the surgery a large area of gliosis was identified and resected. The resected area included calcified tissue measuring approximately 7 × 6 × 2 mm, in contact with the frontal horn of the left lateral ventricle. Histological examination of the calcified tissue showed palisading spindle to epithelioid cells, calcification and osseous metaplasia with primarily adipose bone marrow, and a fibrous stroma. The pathologist confirmed the diagnosis of CAPNONA. The patient had two seizures during the first week following the surgery, and since that time (11/2009) is seizure-free.

### Discussion

First described by Rhodes and Davis in 1978 [1], only 45 cases of CAPNONA have been reported [1–18]. CAPNONA have been reported more in males than females (1.5:1) and in ages ranging from 6–83 years (27 male, 18 female; mean age at diagnosis 47.4 years, SD 17 years) (Table 1).

The histopathological characteristics of CAPNONA are diverse. Aiken et al. [2] summarized the typical findings as the variable appearance of 1. a typical chondromyxoid matrix in a nodular pattern; 2. palisading spindle to epithelioid cells; 3. variable amounts of fibrous stroma; 4. calcification, osseous metaplasia, and scattered psammoma bodies; 5. and foreign-body reaction with giant cells. Only one of our cases demonstrated all five findings. Calcification, osseous metaplasia, scattered psammoma bodies, and a variable amount of fibrous stroma were seen in all three of our cases.

MRI findings in CAPNONA have been previously reported [2,4,7–15] and may

Table 1. CAPNONA – Reported Cases.

References	Case No.	Age	Sex	Location	T1WI	T2WI	PDWI	Contrast	Previous Trauma/ /Surgery
Rhodes & Davis, 1978	1	27	F	R Frontal lobe, IA	*	*	*	*	No
	2	55	F	Cranial dura, EA	*	*	*	*	*
	3	60	M	L cerebellum, EA	*	*	*	*	*
	4	74	F	cranial dura, EA	*	*	*	*	*
	5	46	M	choroid plexus, 4th ventricle	*	*	*	*	*
	6	62	F	pineal leptomeninges, EA	*	*	*	*	*
	7	83	M	cranial dura, EA	*	*	*	*	*
Jun & Burdick, 1984	8	55	M	corpus collosum, EA	*	*	*	*	yes, trauma
Garen et al., 1989	9	44	M	R trigeminal cave, EA	*	*	*	*	*
Bertoni et al., 1990	10	31	M	skull base, EA	*	*	*	*	*
	11	50	M	skull base, EA	*	*	*	*	*
	12	48	M	R cerebellar tonsil, IA	*	*	*	*	*
	13	23	M	T10, EA	*	*	*	*	*
	14	58	M	C2–C3, EA	*	*	*	*	*
	15	32	M	frontal lobe, IA	*	*	*	*	*
	16	45	F	skull base, EA	*	*	*	*	*
	17	58	M	skull base, EA	*	*	*	*	*
	18	12	M	C6, EA	*	*	*	*	*
	19	32	M	L4–L5, EA	*	*	*	*	*
	20	33	F	T9, EA	*	*	*	*	*
	21	68	F	L4–L5, EA	*	*	*	*	*
	22	20	F	C2, EA	*	*	*	*	*
	23	56	F	L4–L5, EA	*	*	*	*	*
	Moser et al., 1994	24	68	M	C7–T1, EA	iso	hyper	*	rim C+
Smith & Berry, 1994	25	48	M	L2–L3, EA	*	*	*	no C+	yes, surgery
Shrier et al., 1999	26	32	F	L temporal lobe, IA	hypo	hypo	*	rim C+	*
	27	59	M	foramen magnum, EA	hypo	hypo	*	rim & heterogeneous C+	*
Qian et al., 1999	28	33	F	L temporal lobe, IA	*	*	*	*	*
	29	49	M	upper cervical/Clivus, EA	*	*	*	*	*
	30	59	M	C1–C2, EA	*	*	*	*	*
	31	47	F	frontal region, IA	*	*	*	*	*
Tsugu et al., 1999	32	22	F	R parietal lobe, IA	*	*	*	*	no
Chang et al., 2000	33	60	M	C2, intraosseous	hypo	hypo	*	rim C+	*
Mayr et al., 2000	34	58	M	T10–12, EA	hypo	hypo	*	rim C+	*
	35	63	M	C3–C4, EA	hypo	hypo	*	*	*
Tatke et al., 2001	36	6	M	L temporal lobe, IA	*	*	*	*	*
Liccardo et al., 2003	37	40	M	T8–T9, EA	hypo	hypo	*	no C+	*
Rodriguez et al., 2008	38	67	F	R cerebellar hemisphere, IA	hypo	hypo	*	rim C+	*
Park et al., 2008	39	59	F	C7–T1, EA	iso	iso	*	rim C+	*
Aiken et al., 2009	40	16	M	R temporal horn, IV	hypo	hypo	*	internal linear C+	*
	41	35	M	R temporal lobe, IA	hypo	hypo	*	internal linear C+	*

References	Case No.	Age	Sex	Location	T1WI	T2WI	PDWI	Contrast	Previous Trauma/ Surgery
	42	49	F	L hippocampus, IA	hypo	hypo	*	no C+	*
	43	59	M	R parietal lobe, IA	hypo	hypo	*	rim C+	yes, surgery
Montibeller et al., 2009	44	67	F	R inferior colliculus, EA	iso	hypo	*	homogeneous C+	*
Tong et al., 2010	45	67	F	L4–L5 EA	*	*	*	*	*
present report	46	46	M	R angular region, IA	hypo	hypo	hypo	nodular C+	yes, trauma
	47	43	F	L3, EA	hypo	hypo	hypo	*	yes, surgery
	48	23	M	L frontal region, IA	hypo	hypo	hypo	*	yes, surgery

\* Not reported

CAPNONA: Calcifying Pseudoneoplasms of the Neural Axis; IA: intra-axial; IE: extra-axial; IV: intraventricular; T1WI: T1-weighted images; T2WI: T2-weighted images; PDWI: proton-density-weighted images; hypo: hypointensity; hyper: hyperintensity; iso: isointensity; C+: contrast enhancement; T: trauma; S: surgery; HET: heterogeneous; HOM: homogeneous; NOD: nodular

be summarized as 1. central low signal intensity on T1/T2/PD-weighted images consistent with calcification, and 2. variable post-contrast enhancement, most often seen as peripheral rim enhancement. There may be a single focus or multiple foci of low signal intensity reflecting the heterogeneous nature of calcification reported in these lesions.

Some exceptions to these typical findings have been reported. Moser et al. [10] reported central T1 isointensity with a margin of slightly increased signal, and central T2 hyperintensity. At surgery the lesion was found to be fluid-filled. Park et al. [11] reported central isointensity on T1/T2-weighted images; the pathology report indicated that the mass was semi-solid and contained only scattered calcification. Montibeller et al. [9] also presented a case with central T1 isointensity; the lesion was shown to be of low cellularity and only partially calcified. All three of the cases we present showed central hypointensity in all imaging sequences.

When a contrast agent has been administered, peripheral rim enhancement is most often reported and reflects the presence of a well-developed fibro-vascular stroma [2,8,11,13,14]. Internal enhancement has been reported to occur as homogeneous [9], heterogeneous [14], or linear [2], and lesions showing no enhancement have been reported as well [2,7,15]. A contrast agent was only administered in one of our patients and

showed a nodular pattern of internal enhancement.

The differential diagnosis of calcified intra- and extra-axial lesions is broad, but can be significantly narrowed by reviewing the medical history of the patient and by careful observation of the location and imaging characteristics of the lesion. To the best of our knowledge, CAPNONA have never been reported to occur within the spinal cord. It would appear from the limited amount of data available that CAPNONA occur in three spaces (IA-Intracranial, EA-Intracranial, EA-Spinal), and within each of these three spaces with approximately equal frequency (Table 1).

Primary neoplasms such as oligodendrogliomas, choroid plexus papillomas, ependymomas, astrocytomas, meningiomas, and craniopharyngiomas may all present as calcified intracranial masses [19] and should be included in the differential diagnosis. Other calcifying intracranial processes may also be considered such as chronic hematomas, vascular malformations, and tuberculoma. In the spine, the differential diagnosis of a calcified mass should include herniation of calcified disc material, calcified synovial cyst and psammomatous meningioma, followed by less frequently calcifying processes such as epidural abscess, chronic hematoma and tuberculoma.

CAPNONA are often considered to be a reactive, metaplastic response to injury [1,3,12,13,16]. A history of trauma or surgery was only reported in five cases previ-

ously, being positive in three [2,6,15] and negative in two [1,18]. All three of our patients had a history of previous trauma (case 1) or surgery (cases 2 & 3), and in two of these cases this directly impacted the region where CAPNONA later manifested. In one of our cases the previous surgery was at the level of L5/S1 and CAPNONA later manifested at the level of L3. Thus, our cases lend further support to a reactive hypothesis.

In conclusion, CAPNONA are rare, benign lesions that have been reported in IA and EA locations intracranially and in EA locations in the spinal region. The histopathological composition of these lesions is diverse, which may be reflected in the imaging findings. Finally, considering the history of trauma or surgery in the cases presented here and in those reported previously in the literature, it would appear that in at least some cases CAPNONA are a reactive, metaplastic response to injury.

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## Vysoké štátne vyznamenanie prof. Danielovi Bartkovi

Prezident Slovenskej republiky udelil vysoké štátne vyznamenanie RÁD LUDOVÍTA ŠTÚRA prof. MUDr. Danielu Bartkovi, PhD., DrSc, FAAN, FRSM, FAHA, FESO, čestnému členovi American Academy of Neurology, American Heart Association and American Stroke Association (Premium Professional Member) a ďalších 11 významných zahraničných odborných spoločností, čestnému členovi Slovenskej, Českej i Československej lekárskej spoločnosti a ďalších za mimoriadne zásluhy a výsledky pre rozvoj vedeckého poznania v oblasti medicínskych vied a osobitne neurológie.

Z predložených 212 návrhov bolo toto vyznamenanie udelené 15 navrhovateľom, z toho trom in memoriam a dvom v oblasti medicíny.

Srdečne blahoželáme

### Prof. Bartko: Úprimne ďakujem všetkým.

Vysoké vyznamenanie považujem za ocenenie všetkých mojich bývalých aj súčasných spolupracovníkov, mojej alma mater, minulej aj terajšej, celej našej slovenskej, českej aj bývalej československej neurológie a príbuzných odborov, mojej rodiny a priateľov doma i v zahraničí. Sme takí, akí sme, a preto niekedy veríme aj v zázraky. Takým zázrakom je nevídaný rozvoj neurológie a neurologických vied všeobecne. Sme svedkami toho, že najvýznamnejšie výsledky techniky exploituje najmä neurológia a neurovedy, že niektoré oblasti neurologických vied, ktoré boli pred pár desiatkami rokov na pokraji vedeckého záujmu neurológov, sa stali pre nich neatraktívnejšími a prinášajú nové, progresívne, provokujúce revolučné výsledky tak v oblasti vedeckého poznania, ako aj ich implemen-

tácie pre klinickú prax. To je mohutný zdroj permanentnej stimulácie nášho mozgu a jeho perfektnej plasticity. Nič však nemožno dosiahnuť bez boja. Boj je otcom i kráľom všetkého, z jedných robí bohov, z druhých ľudí, z jedných otrokov a z druhých slobodných (Herakleitos). Kto chce niečo dosiahnuť, a naozaj to chce, kto bojuje, pomáha mu všetko, celý vesmír, aby to dosiahol. Nech sa nám a našej neurológii naďalej darí, nech sú doterajšie výsledky mohutnou a mnohostrannou stimuláciou nášho mozgu. To umožní, aby nám nervové bunky naďalej dobre fungovali a žľazy endokrinné nevysychali. To prajem nám všetkým, našej neurológii, našej medicíne, nášmu zdravotníctvu aj našej spoločnosti. Ďakujem Vám, priatelia. Som rád, že som medzi Vami, že som s Vami, že som tu.

D. Bartko