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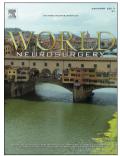
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Calcifying pseudoneoplasm of the neuraxis (CAPNON): a case illustrating natural history over 17 years of radiological surveillance

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Abstract

Calcifying pseudoneoplasm of the neuraxis (CAPNON) is a rare central nervous system (CNS) lesion that can be found in both the brain and the spine. Although this entity is poorly understood, radiological and histological features have been identified. We describe a unique case of a 31-year-old patient who was managed with antiepileptic medication for 17 years before requiring neurosurgical intervention for tumour progression. T2weighted MRI revealed hyperintensity within the tumour with extensive associated vasogenic oedema, which is not normally associated with CAPNON. Resection was successful with no complications.

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Introduction

Calcifying pseudoneoplasm of the neuraxis (CAPNON) was first described by Rhodes and Davis in 1978.¹ This rare central nervous system (CNS) lesion is known to follow an indolent course, and can present as either an intracranial or spinal mass. These entities were initially thought to represent foci of atypical bony metaplasia,¹ however it is now thought that these lesions may reflect a reactive proliferative process.² Approximately 100 cases of CAPNON have been reported in the literature, and the associated radiological and histological features have been characterized.³ The radiological differential diagnosis includes meningioma, oligodendroglioma, and astrocytoma with calcifications, among others.⁴

Intracranial CAPNON can present with seizures, headaches, visual loss, weakness, or developmental delay.^{2,3,5,6} CAPNON of the spinal cord may present with back pain,⁷ sciatica,⁸ or less commonly, acute cauda equina syndrome.⁹ A small number of cases have also been discovered incidentally at autopsy.¹ Once a radiological diagnosis of CAPNON has been made, treatment typically involves total surgical resection, which confers an excellent prognosis.

Previous cases have described a 4-8 year monitoring period prior to progression warranting neurosurgical intervention.^{4,10,11} To the best of our knowledge, this is the first case report to describe the natural history of CAPNON in a patient over an extended (17 year) period of frequent radiological surveillance prior to surgical intervention. We report a 31-year-old patient who was diagnosed as an adolescent with a perirolandic mass that was initially managed with anti-epileptic medication and regular monitoring by serial imaging. She subsequently underwent total surgical resection after interval imaging demonstrated progression.

Case Report

A 14-year-old female with no significant past medical history presented with intermittent left facial numbness which was diagnosed retrospectively as simple partial epilepsy. She was found to have a mass in the right perirolandic area, initially thought to be a cavernoma, which was followed with serial imaging for several years. Her seizure frequency and severity continued to progress and intrusive complex partial seizures with occasional generalisation were treated with multiple anti-epileptic drugs without remitting. Imaging at the time of her most recent surgical presentation, at which time she had more than daily seizures despite triple therapy, revealed an intrinsic mass containing solid and cystic components with a surrounding area of diffuse T2 hyperintensity

consistent with vasogenic oedema and/or infiltration. Although the lesion showed irregular contrast enhancement, it also contained several foci of susceptibility artefact consistent with calcification (**Figure 1**, **Figure 2**). Functional magnetic resonance imaging (fMRI) performed at the time of presentation confirmed superficial co-location with sensory cortex and involvement of motor pathways in deep white matter, where activation was seen with left finger tapping tasks. The location of the lesion was consistent with the patient's seizure semiology, which typically involved left hemianesthesia, a distorted sense of sound and an altered perception in three-dimensional space, often progressing to generalized tonic-clonic activity.

The patient was monitored with serial imaging (over a period of 17 years) prior to surgical intervention in view of the eloquent location of the mass, the presumed diagnosis of low-grade glioma, and the maintenance of relatively good seizure control. Follow-up magnetic resonance imaging (MRI) every 1-2 years demonstrated slow but progressive enlargement of the mass. In 2017, at the age of 31, further tumour enlargement and increase in extent of surrounding oedema was seen (**Figure 1**). Given this, together with worsening seizure control, the patient agreed to undergo right-sided awake craniotomy and resection of the mass.

During the surgery, stimulation of the cortical surface overlying the tumour did not elicit motor activity, and thus a corticectomy was made at this point to gain access to the mass. The lesion was found to be heavily calcified at the core necessitating piecemeal resection with pituitary rongeurs, microdissection and the use of an ultrasonic aspirating tool. Resection was carried out until normal-appearing brain was reached.

Post-operatively, the patient complained of intermittent sensations of numbness and paraesthesiae in the left forearm and hand, but was otherwise symptom free and had no neurological deficits. She recovered fully and was discharged home within several days. She has been monitored radiographically for 9 months without recurrence. Clinically she has experienced a greater than 50% reduction in her seizure frequency, but remains on anti-epileptics.

Histologically, the tumour was characterized as a fibro-osseous lesion consisting of deposits of amorphous granular to fibrillary material and fibrous tissue, with variable calcification and foci of irregular osseous metaplasia. Numerous calcified foci of variable size were seen with central areas of dense mineralisation/ossification and radially arranged spicules at the peripheries. A population of cytologically bland

and uniform (apparently meningothelial) cells was also noted, composed of cells with oval nuclei, indistinct nucleoli and little to no mitotic activity. A variable chronic, inflammatory infiltrate was also present, including a mixed population of small lymphocytes (CD20+ and CD3+), plasma cells, macrophages (CD68+) and occasional foreign-body-type giant cells. No necrosis, microvascular proliferation or mitotic activity was observed. The meningothelial-type cells expressed epithelial membrane antigen (EMA), but not pancytokeratin, and to a variable extent progesterone receptor (PR). The constituent cells were negative for neuronal and glial markers. The Ki67 proliferation index was low (5-10%), a proportion of which represented inflammatory cell infiltrates. This histological appearance (**Figure 3**) was felt to be consistent with a calcifying pseudoneoplasm of the neuraxis (CAPNON).

Discussion

CAPNON is a rare CNS lesion that is poorly understood. The aetiology is yet to be established, but the cellular element is postulated to be of arachnoid or fibroblastic origin. CAPNONs have been discovered in a variety of intra – and extra-axial locations within the CNS, including the foramen magnum,¹² skull base,¹³ inferior colliculus,¹⁴ and spine,² among others. **Tables 1**, **2** and **3** summarize over 100 cases of CAPNON that have been reported in the literature. Among these reported cases, the average age at presentation was 47 ± 17 years, and a slight male predilection was seen (53.4% male, 44.7% female, 1.9% no data). 1,1015,1617,182,1920–2223,24253,264,13,14,27–356,7,36–435,44,45 211,20,4621,29,47–547,55–639,64,65 Seizures (31.1%) and

headaches (31.1%) appear to be the primary clinical presentation of intracranial CAPNON (most likely due to localised mass effect), while spinal CAPNON most typically present with axial neck or back pain (57.1%) and radicular symptoms (35.7%). Kerr et al⁴ reported a case in which headaches were associated with enlargement of a CAPNON at the right cerebellomedullary angle and hypothesised that headache occur due to growth of the mass itself or a reactive process in the surrounding brain tissue. Surgical resection is the treatment of choice, and due to the benign nature of the process, post-operative chemo-radiotherapy is not recommended.

The radiological features of CAPNON documented in numerous case reports are relatively inconsistent. While a predominant hypointense signal intensity on both T1 & T2-weighted imaging is typically seen (corresponding to dense calcification on computed tomography [CT]), the enhancement and oedema patterns associated with CAPNON appear variable, as does the axial location of the mass. The majority of reported CAPNON cases display a minimal peripheral enhancement pattern,^{3,5,12,18} although in some cases a heterogenous or serpiginous

internal enhancement pattern is seen.^{3,12,18} Mild surrounding T2 hyperintensity suggestive of oedema has been reported in several cases,^{5,18} but not in others.^{3,12} In the majority of reported cases, intracranial CAPNON is an intra-axial mass, but both Shrier et al.¹⁸ and Wiśniewski et al.¹² have described cases in which CAPNON is seen as an extra-axial mass near the foramen magnum. Spinal CAPNON are most commonly extradural (**Table 3**). The imaging findings in our case are distinguished by a prominent T2 hyperintense component suggestive of cyst formation around the calcified nodule, as well as extensive surrounding oedema in the later stages of progression (Figure 1G), which has not previously been described.

As in our case, these lesions are consistently immunopositive for vimentin and EMA, but negative for S100 protein and glial fibrillary acidic protein (GFAP).^{4,6,11,14,21} The typical histological characteristics of CAPNON have been summarised by Qian et al²⁰ and include: (1) a chondromyxoid matrix in a nodular pattern; (2) palisading spindle to epithelioid cells; (3) fibrous stroma (4) osseous components containing birefringent lamellar bone, calcifications, and psammoma bodies; and (5) foreign-body reaction with giant cells. Not all features are present in every case.

The rarity of this entity and the absence of any pathognomonic imaging features makes it difficult to diagnose based on radiographic appearances alone. The radiological differential diagnosis includes intra-axial neoplasms which are also known to calcify (e.g. ganglioglioma and oligodendoglioma), vascular lesions (e.g. cavernomas), and infectious processes (e.g. tuberculomas). CAPNON may also develop extra-axially and mimic meningioma.²¹ Uniform T1 and T2 hypointensity without solid enhancement have been cited as key distinguishing features of CAPNON.³ However, as we have demonstrated CAPNON may produce a more complex radiographic appearance and be associated with extensive surrounding vasogenic oedema.

Conclusion

CAPNON is a rare, non-neoplastic lesion of the central nervous system that, although benign, may nevertheless exert mass effect and may show a progressive increase in size, mimicking an intraaxial or extraaxial tumour. The present case illustrates the long-term natural history of CAPNON prior to resection and highlights the variations in radiological appearances that may be associated with this poorly understood entity.

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Figure 1. MRI images (2005, top row; 2017, bottom row). Images depict progression in tumour size and associated oedema over 12 years of radiographic follow-up. (A, F) axial T2, (B, G) axial T2 FLAIR, (C, H) coronal T1 pre-contrast, (D, I) coronal T1 post-contrast, (E, J) axial T1 post-contrast sequences.

Figure 2. Axial susceptibility-weighted MRI demonstrating susceptibility artefact associated with the right frontoparietal mass, consistent with the dense calcification (later confirmed during surgery).

Figure 3. Histological appearance. (A) Low-power micrograph demonstrating areas of calcification (arrow) and metaplastic bone formation (arrowhead). (B) Medium-power image depicting increased cellularity and variable mineralisation/calcification. (C) Low power image. Radial spiculated arrangement around central mineralised area (arrow). (D) Immunohistochemistry for epithelial membrane antigen (EMA). EMA-positive cells are seen around the periphery of calcified nodules.

Case	Author	Year	Age (years)	Sex	Presenting Symptoms	Location	Contrast Enhancement	Oedema	Treatment	Post- operative Follow- up (months)	Clinical Outcome	Recurrence
1	Rhodes and Davis ¹	1978	27	F	Headache	Right frontal (IA)	ND	ND	Surgical resection	84	Improved	None
2			55	F	Autopsy finding	Cranial dura (EA)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)
3			60	М	Autopsy finding	Left cerebellum (EA)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)
4			74	F	Autopsy finding	Cranial dura (EA)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)
5			46	М	Autopsy finding	Choroid plexus, 4 th ventricle (EA)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)
6			62	М	Autopsy finding	Pineal region (EA)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)
7			83	М	Autopsy finding	Cranial dura (EA)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)
8	Jun et al. 10	1984	55	М	Headache, nausea	Corpus callosum (EA)	ND	ND	Surgical resection	ND	Stable	None
9	Maruki et al. ¹⁵	1984	43	F	Seizures	Right temporal (IA)	ND	ND	Surgical resection	ND	ND	None
10	Hashimoto et al.	1986	29	М	Seizures	Right temporal (IA)	ND	ND	Surgical resection	ND	Seizure free	None
11	Nitta et al. ¹⁷	1987	28	F	Vertigo	Right cerebellum/dentate nucleus (IA)	ND	ND	Surgical resection	ND	ND	ND
12	Garen et al. 18	1989	44	М	Facial pain	Right meckel's cave (EA)	ND	ND	Surgical resection	132	Pain resolved	ND
13	Bertoni et al. ²	1990	31	М	Headache, jugular foramen syndrome	Left jugular foramen, cerebellopontine angle (EA)	ND	ND	Surgical resection	156	ND	Recurred at 3 years
14			48	М	Right cranial nerve 11 palsy	Right cerebellar tonsil (IA)	ND	ND	Surgical resection	228	ND	None
15			32	М	Seizures	Frontal lobe (IA)	ND	ND	Surgical resection	360	ND	None
16			45	F	Autopsy finding; History of multiple cranial nerve palsies	Right skull base (sphenoid, occipital bone, sella, foramen magnum) (EA)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)	NA (autopsy)
17			58	М	Hoarseness, hearing loss	Jugular foramen (EA)	ND	ND	Surgical resection	Lost to follow-up	ND	ND
18	Tokunaga et al.	1995	72	F	Tinnitus, ataxia	Cerebellum (IA)	ND	ND	Surgical resection	ND	Improved	ND
19	Qian et al. 20	1999	33	F	Developmental delay	Left superior temporal gyrus (IA)	ND	ND	Surgical resection	31	Stable	None
20			47	F	Seizures	Frontal lobe (IA)	ND	ND	Surgical resection	36	ND	None
21	Shrier et al. 21	1999	32	F	Incidental	Left temporal (IA)	Minimal, peripheral	+	Surgical resection	12	Stable	None
22	Tsugu et al. 22	1999	22	F	Seizures	Right parietal (IA)	ND	None	Surgical resection	ND	Improved	None
23	Albu et al. ²³	2001	53	F	Headache, visual loss	Left fronto-parietal, right fronto-parietal (IA)	ND	ND	ND	ND	ND	ND

24	Tatke et al. 24	2001	6	М	Seizures	Left temporal (IA)	ND	ND	Surgical resection	6	Improved	None
25	Ghosal et al. 25	2007	26	F	Seizures	Trigone/temporal horn (EA)	Minimal, irregular	++	Surgical resection	6	ND	None
26	Rodriguez et al.	2008	67	F	Incidental	Right cerebellar hemisphere (IA)	Minimal, peripheral	+	Surgical resection	ND	Recovered well	ND
27	Aiken et al. 3	2009	16	М	Incidental	Right temporal horn (EA)	Minimal, irregular	None	Surgical resection	None	ND	ND
28			35	М	Seizures	Right temporal (IA)	Minimal, irregular	ND	Surgical resection	None	ND	ND
29			49	F	Seizures	Left hippocampus (IA)	None	Minimal	Surgical resection	None	ND	ND
30			59	М	Left arm numbness	Right parietal (IA)	Minimal, peripheral	ND	Surgical resection	None	ND	ND
31	Montibeller et al. 14	2009	67	F	Episodic dizziness	Right inferior colliculus (EA)	Strong, heterogenous	None	Surgical resection	18	Symptoms resolved	None
32	Mohapatra et al.	2010	48	М	Seizures	Right temporal (IA)	Minimal, irregular	ND	Surgical resection	6	Stable	None
33	Ghosal et al. ²⁸	2011	50	М	Headache, seizures	Posterior third ventricle/pineal region (EA)	Strong, heterogenous	ND	Surgical resection	6	Stable	None
34	Hodges et al. ¹³	2011	34	М	Headaches, left cranial nerve 12 palsy	Left cerebellopontine angle (EA)	Moderate, irregular	ND	Surgical resection	7	Stable	None
35	Rulseh et al. 29	2011	46	М	Headaches	Right parietal (IA)	Strong, heterogenous	+	Surgical resection	6	Improved	None
36			23	М	Seizures	Left frontal (IA)	None	Minimal	Surgical resection	ND	Improved	ND
37	Yan et al. 30	2011	45	М	Seizures	Right mesial temporal (IA)	None	ND	Surgical resection	48	Seizure free	None
38	Fletcher et al. 31	2012	19	М	Frontal headache, nasal discharge	Left ethmoid sinus, anterior skull base (EA)	Minimal, irregular	ND	Surgical resection	ND	ND	ND
39	Nonaka et al. 32	2012	56	М	Chronic right ear infections, right facial pain/numbness	Right infratemporal fossa (EA)	Minimal, irregular	None	Surgical resection	ND	Stable	ND
40			35	М	Occipital headaches, dizziness	Left clivus/posterior fossa (EA)	Minimal, irregular	Minimal	Surgical resection	6	Stable	None
41	Salim et al. ³³	2012	47	F	Headache, gait ataxia, cognitive decline	Right frontal, periventricular (IA)	ND	+	Surgical resection	ND	Stable	ND
42	Grabowski et al.	2013	49	F	Headaches	Corpus callosum (EA)	Minimal, irregular	+	Surgical resection	21	Improved	None
43	Kerr et al. ⁴	2013	56	М	Suboccipital headache	Right cerebellomedullary angle (EA)	Minimal, peripheral	Minimal	Surgical resection	6 months	Hoarseness, swallowing difficulty improving	None
44	Stienen et al. 35	2013	46	М	Seizures	Right parietal (IA)	Minimal, peripheral	+	Surgical resection	10	Seizure free	ND
45			55	F	Behavioral disorder	Left frontoparietal (IA)	Minimal, peripheral	+	Surgical resection	22	Temporary right brachiofacial hemiparesis, aphasia	ND

46	Lyapichev et al.	2014	24	м	Seizures, progressive	Right temporo-occipital	None	Minimal	Surgical	1.5	Stable	ND
	U				visual loss	(IA)			resection	6		
47	Shimony et al.	2014	33	М	Headache, deteriorating vision	Corpus callosum/parafalcine (EA)	Minimal, peripheral	++	Surgical resection	10	Stable	ND
48			54	М	Incidental	Corpus callosum/parafalcine (EA)	Minimal, irregular	Minimal	Surgical resection	2.5	Post-operative hemorrhage, left leg weakness, hydrocephalus	ND
49			60	М	Incidental	Left frontal (IA)	Minimal, peripheral	Minimal	Surgical resection	15	Stable	ND
50	Hubbard et al. 37	2015	46	F	Headaches, left facial numbness	Bilateral fronto-parietal (IA)	Minimal, irregular	Minimal	Surgical resection	18	Headaches improved	ND
51	Krejči et al. ³⁸	2015	38	F	Seizures	Right perirolandic (IA)	Minimal, peripheral	X +)	Surgical resection	17	Mild left arm weakness	None
52	Lu et al. 39	2015	No data	No data	Suboccipital headaches	Right cerebellomedullary angle (EA)	Minimal, irregular	Minimal	Surgical resection	ND	ND	ND
53	Duque et al. ⁷	2016	48	F	Headaches	Left atrium (EA)	ND	+	Surgical resection	24	Stable	ND
54	Ghaemi et al. 40	2016	18	ND	Headache, left oculomotor nerve palsy	Left interpeducular cistern (EA)	Minimal, peripheral	None	Surgical resection	No data	ND	ND
55	Merola et al. 41	2016	39	F	Seizures	Left perisylvian (IA)	Minimal, peripheral	++	Surgical resection	1	Left frontoparietal infarct, right hemiparesis	ND
56	Tan et al. 42	2016	45	М	Headaches, diplopia, left cranial nerve 6 palsy	Superior medullary velum (EA)	Irregular/rim	Minimal	Surgical resection	3	Improved	ND
57	Zerehpoosh et al. 43	2016	25	М	Incidental	Left temporal (IA)	Minimal, irregular	Minimal	Surgical resection	36	Stable	ND
58	Brasiliense et al. ⁴⁴	2017	67	F	Seizures	(1) Ventral midbrain (IA), (2) left frontal (IA)	(1) None, (2) minimal, irregular	(1) None, (2) Minimal	Surgical resection (left frontal)	4	Stable	None
59	Higa et al. ⁵	2017	62	М	Headaches	Left cingulate gyrus (IA)	Minimal, peripheral	+	Surgical resection	14	Improved	Recurred at 14 months
60	Nussbaum et al.	2018	39	F	Right-sided hearing loss/tinnitus	Right jugular foramen (EA)	Minimal, irregular	None	Surgical resection	ND	Right-sided complete hearing loss	ND
61	Barber et al.	2018	14	F	Seizures	Right perirolandic (IA)	Minimal, irregular	+++	Surgical resection	9	Improved seizure frequency	None
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Table 1. Summary of reported cases of intracranial CAPNON in the literature. M, male; F, female; ND, no data; NA, not applicable; IA, intra-axial; EA, extra-axial

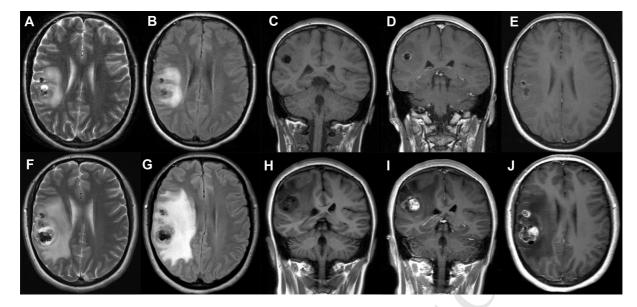
Case	Author	Year	Age (years)	Sex	Presenting Symptoms	Location	Contrast Enhancement	Oedema	Treatment	Post- operative Follow- up (months)	Clinical Outcome	Recurrence
1	Bertoni et al. ²	1990	50	М	Right neck pain, occipital headache	Right foramen magnum, extradural	ND	ND	Surgical resection	42	ND	None
2			23	М	Back pain	T10, extradural	ND	ND	Surgical resection	Lost to follow-up	ND	ND
3			58	М	Incidental	C2/3, extradural	ND	ND	Surgical resection	112	ND	None
4			12	М	Neck pain	C6, extradural	ND	ND	Surgical resection	39	ND	None
5			32	М	Back pain	L4/5, extradural	ND	ND	Surgical resection	84	ND	None
6			33	F	Back pain	T9, extradural	ND	ND	Surgical resection	Lost to follow-up	ND	ND
7			68	F	Right hip pain	L4/5, extradural	ND	ND	Surgical resection	16	ND	ND
8			20	F	Incidental	C2, extradural	ND	ND	Surgical resection	Lost to follow-up	ND	ND
9			56	F	Back pain	L4/5, extradural	ND	ND	Surgical resection	Lost to follow-up	ND	ND
10	Moser et al. ⁴⁶	1994	68	М	Left arm radicular pain	Left C7/T1, extradural	Minimal, peripheral	None	Surgical resection	ND	Improved	ND
11	Smith and Berry ¹¹	1994	48	М	Left leg radicular pain	L2/3, intradural/extramedullary	None	ND	Surgical resection	ND	Improved	ND
12	Qian et al. 20	1999	49	М	Weakness	C1/clivus/foramen magnum, extradural	ND	ND	Surgical resection	90	ND	None
13			59	М	Shuffling gait	C1/2, extradural	ND	ND	Surgical resection	46	ND	None
14	Shrier et al. 21	1999	59	М	Neck pain, shuffling gait	Left foramen magnum, extradural	Solid, heterogenous	+	Surgical resection	24	Stable	None
15	Chang et al. ⁸	2000	60	М	Neck pain	C2 odontoid process, intraosseous	Heterogenous	None	Surgical resection	24	Improved	Local recurrence at 24 months
16	Mayr et al. ⁴⁸	2000	58	М	Back pain, myelopathy	T10-12, extradural	Minimal, peripheral	None	Surgical resection	48	Improved	None
17			63	М	Bilateral upper extremity numbness, myelopathy	C3/4, extradural	ND	None	Surgical resection	60	Improved	None
18	Liccardo et al.	2003	40	М	Thoracic radicular pain, myelopathy	T8/9, extradural	None	ND	Surgical resection	36	Improved	ND
19	Sharma et al. 50	2005	55	М	Low back pain, bilateral leg pain	L3, intradural/filum terminale	None	None	Surgical resection	ND	Improved	ND
20	Park et al. 51	2008	59	F	Neck pain, left C8 radicular pain	Left C7-T1, extradural	Minimal, peripheral	Minimal	Surgical resection	ND	Improved radicular pain	ND
21	Apostolopoulos et al. 52	2009	53	М	Low back pain, left leg pain	Conus, Intradural/extramedullary	Strong, heterogenous	ND	Surgical resection	ND	Improved	
22	Tong et al. 53	2010	67	F	Lower back pain	Left L4/5, extradural	No data	ND	Surgical resection	ND	ND	ND
23	Ozdemir et al. 54	2011	53	М	Left facial pain, left leg weakness	Left foramen magnum, intradural/extramedullary	Strong, heterogenous	+	Surgical resection	ND	ND	ND
24	Rulseh et al. 29		43	F	Low back pain, bilateral leg pain	L3, extradural	ND	None	Surgical resection	10	Stable	None

25	Jentoft et al. 55	2012	26	F	Left low back pain	L1/2, intradural	Strong	None	Surgical resection	ND	Improved back pain	ND
26	Kwan et al. 56	2012	48	М	Left thoracic radicular pain	T9/10, extradural	None	None	Indomethacin	3	Improved	Complete resolution
27	Muccio et al. 57	2012	55	F	Neck pain	Right foramen magnum, intradural/extramedullary	None	None	Surgical resection	14	Stable	None
28			57	М	Back pain, paraparesis	T10/11, dorsal extradural	None	None	Surgical resection	2	Improved paraparesis	None
29	Naidu and Patel	2012	43	М	Low back pain, left leg pain	Left L4, intradural/extramedullary	Strong, heterogenous	None	Surgical resection	ND	ND	ND
30	Bartanusz et al.	2013	2	F	Neck pain, torticollis	Left C1/2, extradural	Minimal, peripheral	None	Surgical resection	12	Improved neck pain and torticollis	None at 8 months
31	Reinard et al. 60	2015	44	М	Lower back pain, left leg pain	L4, extradural	Minimal, irregular	None	Surgical resection	48	Improved	ND
32	Song et al. 61	2015	77	F	Lower back pain, bilateral leg pain	T12, extradural	ND	ND	Surgical resection	5	Pain resolved	None
33			67	F	Right leg pain	L2/3, extradural	ND	ND	Surgical resection	ND	Pain resolved	ND
34			78	F	Lower back pain	L1, extradural	ND	ND	Surgical resection	ND	Pain resolved	ND
35	Wiśniewski et al. 62	2015	29	М	Headaches, impaired head rotation	Right foramen magnum, intradural/extramedullary	Minimal, peripheral	Minimal	Surgical resection	24	Stable	ND
36	Alshareef et al.	2016	59	F	Incidental	Right cerebellomedullary angle/foramen magnum, intradural/extramedullary	Strong	+	Surgical resection	12	Stable	None
37	Duque et al. ⁷		51	F	Lower back pain, bilateral leg pain	L2, dorsal extradural	ND	None	Surgical resection	12	Improved pain	None
38			46	F	Neck pain	C3, intraosseous	ND	None	Surgical resection	27	Improved pain	None
39			73	F	Paraparesis	T2, intradural/extramedullary	ND	ND	Surgical resection	12	Improved	None
40	Lopes et al. 9	2016	72	F	Cauda equina syndrome	L2, intradural	None	None	Surgical resection	No data	Improving paraparesis	ND
41	Singh et al. ⁶⁴	2016	90	F	Myelopathy	C7/T1, intradural/extramedullary	ND	Minimal	Surgical resection	2	Improved	ND
42	Wu et al. 65	2017	39	F	Sacrococcyeal pain	S2, intradural	Strong, irregular	None	Surgical resection	36	Improved	None

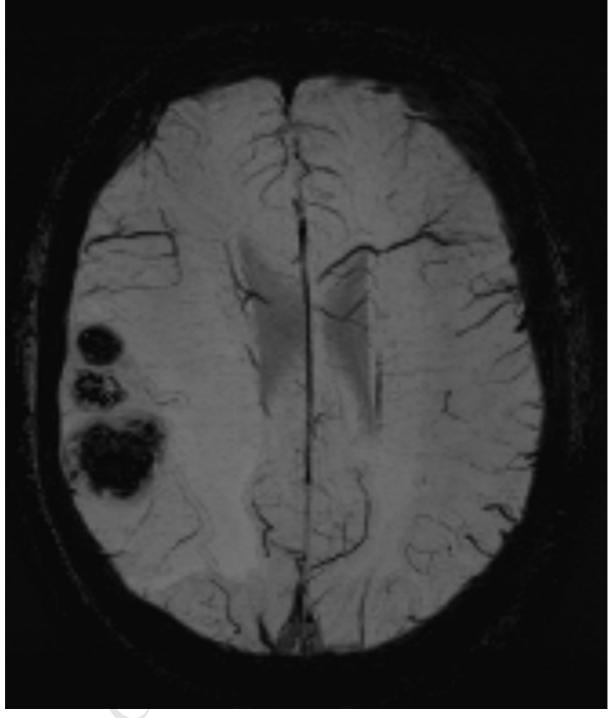
 Table 2. Summary of reported cases of spinal CAPNON in the literature. M, male; F, female; ND, no data

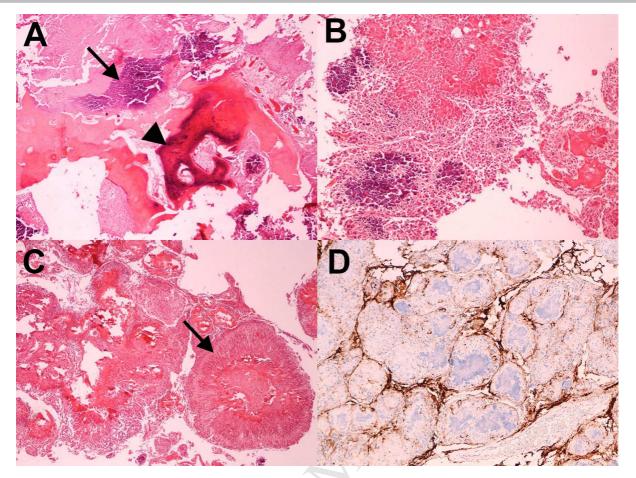
N		103					
Male	e	55 (53.4%)					
Fema	le	46 (44.79	,				
No da	ita	2 (1.9%)				
Age)	47 ± 17 ye	,				
Cran	ial	Spinal					
Ν	61 (59.2%)	N	42 (40.8%)				
·			Y				
	1	Location					
Supratentorial	38 (62.3%)	Lumbar	15 (35.7%)				
Infratentorial	15 (24.6%)	Cervical	11 (26.2%)				
Pineal region	4 (6.6%)	Thoracic	8 (19%)				
No data	3 (4.9%)	Foramen magnum	7 (16.7%)				
Infratemporal fossa	1 (1.6%)	Sacral	1 (2.4%)				
Intraaxial	33 (54.1%)	Extradural	27 (64.3%)				
Extraaxial	28 (45.9%)	Intradural/extramedullary	13 (31%)				
		Intraosseous	2 (4.7%)				
	Present	ting Symptoms					
Headache	19 (31.3%)	Axial pain (e.g. back, neck)	24 (57.1%)				
Seizures	19 (31.1%)	Radicular symptoms	15 (35.7%)				
Autopsy finding	7 (11.5%)	Myelopathy/weakness	9 (21.4%)				
Incidental	6 (9.8%)	Incidental	3 (7.1%)				
Cranial nerve palsy	6 (9.8%)	Cauda equina syndrome	1 (2.4%)				
Dizziness/vertigo	3 (4.9%)						
Facial pain	3 (4.9%)						
Visual loss	3 (4.9%)						
Hearing loss	2 (3.3%)						
Ataxia	2 (3.3%)						

Table 3. Characteristics of patient demographics, presenting symptoms and lesion location for all intracranial and spinal CAPNON reports in the literature. M, male; F, female; ND, no data



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CERTER

- Calcifying pseudoneoplasm of the neuraxis (CAPNON) is a rare central nervous system (CNS) lesion that can be found in both the brain and the spinal cord.
- Several consistent radiographic features have been previously described, such as T1 and T2 hypointensity, and calcification on CT, although the axial location of the mass and the degree of enhancement/oedema are variable.
- Histologically, CAPNON consist of a variable combination of nodular chondromyxoid matrices with palisading spindle to epithelioid cells, a fibrous stroma, osseous components, and foreign-body reactions with giant cells.
- We describe a case of a CAPNON in a 31-year-old patient monitored with serial imaging for 17 years prior to surgical resection and definitive diagnosis. Over this time period a progressive and extensive surrounding oedema developed, a radiographic finding which has not been described previously.

CER MAR

- CAPNON, calcifying pseudoneoplasm of the neuraxis
- CNS, Central nervous system
- MRI, magnetic resonance imaging
- fMRI, functional magnetic resonance imaging
- EMA, epithelial membrane antigen
- PR, progesterone receptor
- CT, computed tomography
- GFAP, glial fibrillary acidic protein

Declaration of Interests

The authors of the enclosed manuscript have no conflicts of interest – financial or otherwise - to disclose.