



Calcifying Pseudoneoplasms of the Neuraxis (CAPNON): Report of Four Cases and Review of the Literature

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Introduction

Calcifying pseudoneoplasms of the neuraxis (CAPNON) are rare lesions occurring anywhere in the central nervous system (CNS). Since their description by Rhodes and Davis, only 45 cases have been reported. We present the largest series reviewing their imaging features, histology and potential origins.

Methods

Four patients with histopathologically verified CAPNON are presented. Subsequently, we review all reports published with respect to study type, number of patients, clinical presentation, anatomical area (intracranial, spinal, or both), radiological features, therapy, histopathologic features, duration of follow-up, complications, and outcome. Moreover, current management of CNS CAPNON are discussed.

Results

Four patients with histopathologically verified diagnosis of CAPNON are presented between 46-73 years-old. Three of them were located in the spinal cord (levels C3,D2 and L2) and one intracranial (left atrium). The spine ones were diagnosed due to radicular pain, paraparesis and numbness in lower limb, the intracranial because of intense headache. The differential diagnosis included cavernous malformation, in the case of the lumbar CAPNON this suspicion put back the surgery six months. All cases were surgically treated with complete resection. No recurrence showed at the 12- month follow-up.

Cranial cases

No	Age	Sex	Presentation	Localization	Treatment	Follow-up (months)	Recurrence
1	48	F	headache	Left Atrium	GT	18	No
2	56	M	headache	Right cerebello-pontine angle	ST	6	No
3	27	F	headache	Right frontal lobe	ST	84	No
4	58	M	dizziness and vomiting	corpus callosum	GT	N/a	No
5	44	M	facial pain	Trigeminal ganglion region	GT	N/a	No
6	31	M	jugular foramen syndrome	Left jugular foramen	ST	156	Yes
7	48	M	Right XI paralysis	Right cerebellar tonsil	GT	228	No
8	32	M	seizures	Frontal lobe	GT	360	No
9	58	M	decreased hearing	Jugular foramen	ST	N/a	N/a
10	22	F	seizures	Right Parietal lobe	GT	96	No
11	32	F	incidental	Left Temporal lobe	GT	12	No
12	33	F	developmental delay	Left Temporal lobe	GT	31	No
13	49	M	tinnitus	Clivus	GT	60	No
14	47	F	seizures	parasagittal frontal region	GT	72	No
15	6	M	seizures	Left temporal medial region	ST	6	No
16	16	M	incidental	Right temporal horn	GT	N/a	No
17	35	M	seizures	Right temporal lobe	GT	N/a	N/a
18	49	F	seizures	Left hippocampus	GT	N/a	N/a
19	59	M	Left arm numbness	Right parietal lobe	GT	N/a	N/a
20	67	F	seizures	Right inferior colliculus	GT	18	N/a
21	48	M	seizures	Right temporobasal	GT	N/a	No
22	36	M	headache and irritability	Left cerebello-pontine angle	ST	7	No
23	46	M	seizures	Right Parietal lobe	ST	10	No
24	55	F	hysteriomania	Left frontoparietal lobe	ST	22	No
25	49	F	headache	corpus callosum	GT	N/a	No
26	24	M	migraine	Right temporo-occipital lobe	GT	12	No
27	36	F	facial numbness	bilateral frontoparietal	GT	96	No

ST Subtotal remove; GT Gross total remove

A total of 21 retrospective articles were identified and selected for review: 7 case series (33.3 %) and 14 reports of single cases (66.6 %). The 21 articles and our additional cases added up to a total of 22 patients with spinal CAPNON and 27 patients with intracranial CAPNON. All patients were treated surgically. A follow-up, provided in 34 patients, showed no signs of recurrence in 32 of 34 patients.

CAPNON are unusual lesions,they don ´t have a predilection for sex, age, or location.The patient age ranges from 12 to 83 years. Most reported lesions have been extra-axial.

Spinal cases

Number	Age (years)	Sex	Presentation	Location	Treatment	Follow-up (months)	Recurrence
1	50	M	neck pain	FM 6	RI	42	No
2	51	F	back pain	L2 la	RC	39	No
3	46	F	neck pain	C3to	RC	27	No
4	73	M	paraparesis	D2to	RC	12	No
5	23	M	back pain	Th10to	RI	N/a	No
6	58	M	parasthesias	C3to	RI	112	No
7	12	M	neck pain	C5to	RI	39	No
8	32	M	back pain	L4to	RI	83	No
9	33	F	back pain	Th6to	RI	N/a	No
10	68	F	sciatica	L4to	RI	16	No
11	20	F	incidental	C3to	RI	N/a	No
12	56	F	back pain	L4to	RI	N/a	No
13	49	M	sciatica	L3to	RC	N/a	No
14	59	M	tetraparesis	FMto	RC	24	No
15	59	M	tetraparesis	C1to	RC	46	No
16	60	M	neck pain	C2to	RI	24	Yes
17	58	M	back pain	Th10to	RI	46	No
18	63	M	tetraparesis	C3to	RI	60	No
19	40	M	thoracic pain	Th6to	RC	36	No
20	59	F	radiculopathy	C7to	RC	N/a	No
21	67	F	neurogenic claudication	L4to	Laminectomy	N/a	No

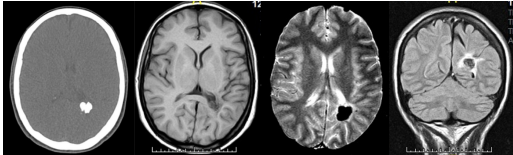
FM foramen magnum; Th Thoracic spinal segment; C Cervical spinal segment; L Lumbar spinal segment; N/a not available; e epidural; ie intradural extramedullary; id intradural; io intrasoosous

The lesions seem to be slow growing with symptoms related to local compression or irritation of adjacent tissues. Its underlying cause remains unknown.

Spinal case

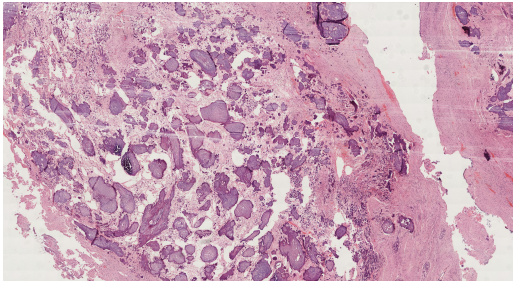


Intracranial case



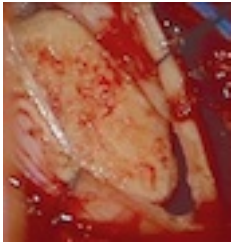
CT images of CAPNON typically show solid attenuated calcifications, and the MR imaging often shows a well-defined lesion that is uniformly hypointense on both T1- and T2- weighted images without surrounding edema.

Histopathologic Evaluation



The “classic” histopathologic features in CAPNON include: 1) typical chondromyxoid matrix in a nodular pattern; 2) palisading spindle to epi- thelioid cells; 3) variable amounts of fibrous stroma; 4) calcification, osseous metaplasia, and scattered psammoma bodies; and 5) foreign-body reaction with giant cells.

Surgical view of a spinal CAPNON



Conclusions

Calcifying pseudoneoplasms are rare benign lesions of yet unknown origin. They should be taken into consideration in the differential diagnosis of calcified lesions because an inaccurate diagnosis can result in potentially harmful and unnecessary therapies, as prognosis for these lesions is generally favorable.

References

- Lyapichev K, Bregy A, Shah AH, Shah K, Desai MB, Petito C, Komotar RJ. Occipital calcified pseudoneoplasms of the neuraxis (CAPNON): understanding a rare pathology. BMJ Case Rep. 2014 Dec 5;2014. pii: bcr2014206855. doi: 10.1136/bcr-2014-206855.
- Stienen MN, Abdulazim A, Gautschi OP, Schneiderhan TM, Hildebrandt G, Lücke S. Calcifying pseudoneoplasms of the neuraxis (CAPNON): clinical features and therapeutic options. Acta Neurochir (Wien). 2013 Jan;155(1):9-17. doi: 10.1007/s00701-012-1502-2. Epub 2012 Oct 3. Review.
- Mohapatra I, Manish R, Mahadevan A, Prasad C, Sampath S, Shankar SK. Calcifying pseudoneoplasm (fibro osseous lesion) of neuraxis (CAPNON) - a case report. Clin Neuropathol. 2010 Jul-Aug;29(4):223-6.
- Aiken AH, Akgun H, Tihan T, Barbaro N, Glastonbury C. Calcifying pseudoneoplasms of the neuraxis: CT, MR imaging, and histologic features. AJNR Am J Neuroradiol. 2009 Jun;30(6):1256-60. doi: