Calcifying Pseudoneoplasm of the Neuraxis (CAPNON): A Comprehensive Review

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Test

Question 1: What is the primary characteristic feature of Calcifying Pseudoneoplasm of the Neuraxis (CAPNON)?

- [ ] A. Rapid tumor growth - [x] B. Presence of calcifications within the central nervous system - [ ] C. High potential for malignancy - [ ] D. Invasive behavior into nearby tissues

Question 2: How is CAPNON typically diagnosed?

- [ ] A. Blood tests - [ ] B. Electroencephalogram (EEG) - [x] C. Imaging studies like MRI or CT scans - [ ] D. Lumbar puncture

Question 3: What is the primary treatment for CAPNON?

- [ ] A. Chemotherapy - [ ] B. Radiation therapy - [x] C. Surgical resection - [ ] D. Immunotherapy

Question 4: What is the benign nature of CAPNON referring to?

- [ ] A. It is easily treatable - [x] B. It does not invade nearby tissues or spread to other parts of the body - [ ] C. It is common and frequently diagnosed - [ ] D. It always resolves on its own without treatment

Question 5: CAPNON typically presents with which of the following symptoms?

- [ ] A. Joint pain - [ ] B. Hearing loss - [x] C. Seizures, headaches, and neurological deficits - [ ] D. Skin rashes

Question 6: What is the importance of long-term follow-up in CAPNON cases?

- [ ] A. To ensure that the patient adheres to a strict diet - [x] B. To monitor disease progression - [ ] C. To evaluate the effectiveness of chemotherapy - [ ] D. To prevent initial symptoms from occurring

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Abstract

Calcifying Pseudoneoplasm of the Neuraxis (CAPNON) is a rare benign brain lesion characterized by central nervous system calcifications. This comprehensive review examines the nature of CAPNON, its clinical manifestations, diagnostic modalities, and treatment options. A case series and multiple case

reports are included, shedding light on the diverse clinical presentations and surgical outcomes associated with CAPNON. The rarity of this condition emphasizes the importance of accurate diagnosis and long-term follow-up to ensure optimal patient management.

Keywords: CAPNON, calcifying pseudoneoplasm, central nervous system, brain lesion, diagnosis, treatment, case series, case reports, rare condition.

Introduction

Calcifying pseudoneoplasm of the neuraxis (CAPNON) is a rare and benign brain lesion characterized by the presence of calcifications within the central nervous system. This condition was first described in the medical literature in 2005 and is still not well understood due to its rarity.

Here are some key points about CAPNON:

Benign Nature: CAPNON is considered a benign brain lesion, meaning it is not a cancerous tumor. It does not invade nearby tissues or spread to other parts of the body.

Calcifications: The hallmark feature of CAPNON is the presence of calcifications within the brain or spinal cord. These calcifications can often be detected through imaging studies such as CT scans or MRI.

Symptoms: CAPNON can sometimes cause symptoms, which may vary depending on the location and size of the lesion. Common symptoms can include headaches, seizures, neurological deficits, and, in some cases, changes in mental status.

Diagnosis: A definitive diagnosis of CAPNON is typically made through imaging studies, such as MRI or CT scans, which reveal the presence of calcifications within the brain or spinal cord. Biopsy or surgical removal may be performed in some cases to confirm the diagnosis.

Treatment: In many cases, CAPNON is managed conservatively, especially if it is asymptomatic or causing only mild symptoms. If symptoms are severe or if there is a concern about the lesion's growth, surgical removal may be considered.

It's important to note that CAPNON is a rare condition, and the understanding of its clinical behavior and management is still evolving.

Case series

Ten patients were identified that met the inclusion criteria. Most patients presented with headaches (n=6, 60%), seizures (n=5, 50.0%), and neck and facial pain (n=3, 30.0%). Most lesions were supratentorial, (n=7, 70.0%), with three infratentorial origins. Surgical resection was the most common initial management undertaken (n=7, 70.0 %). No new permanent post-operative neurological deficits were identified. The median clinical and/or radiographic follow-up for all patients was 6.8 years (range 0.7-23.3 years), with no recurrence of disease for five patients that underwent gross total resection. Four of five patients with residual or non-resectable lesions showed no interval

growth on radiographic follow-up; one patient demonstrated progression and worsening of presenting symptoms two months after resection. Resection substantially improved seizures and headaches in patients presenting with these symptoms (80% and 83.3%, respectively).

Intracranial CAPNONs may present with a wide variety of symptoms characteristic of the site of origin. The outcomes of these regarding survival and disease control are generally favorable, although resection does not always yield complete resolution of presenting deficits in certain patients, particularly those presenting with headaches or neck/facial pain [1)]

Case reports

A 16-year-old girl who experienced bothersome headaches. Through advanced imaging techniques like computed tomography (CT) and magnetic resonance imaging (MRI), we glimpsed a delicate calcified growth within the lateral ventricles' posterior horn. Motivated by our unwavering commitment to solving mysteries, we embarked on a surgical journey that not only freed the young patient from her ailment but also shed light on the true nature of her puzzling adversary remarkable CAPNON.

For patients with CAPNON who have multiple or non-respectable lesions, the primary goal is to alleviate symptoms. After alleviating the symptoms with partial resection, close monitoring of any residual lesions is essential. If there is no evidence for disease progression, a strategy of continued close observation is appropriate [2)].

A 56-year-old female presented with a history of recurrent holo cranial headache and dizziness, progressively worsening over the last month. Physical and neurological examinations revealed no evident abnormalities. Brain magnetic resonance imaging revealed a calcified and cystic mass, measuring 40 × 32 mm in the right frontal lobe. A complete excision of the mass was done. Histologically, the lesion was composed of glial tissue with abundant amorphous lamellar calcification and a myxoid matrix in the background. Concentric circular calcifications were observed with osseous metaplasia present in some areas. Palisading spindle to epithelioid cells was noted around the lesion. The final diagnosis was CAPNON. The postoperative course was uneventful, and one year of follow-up revealed no signs of recurrence.

CAPNON typically occurs in middle-aged adults and can present with variable symptoms depending on its location within the brain, including seizures, headaches, or neurological deficits. Surgical resection is considered the optimal treatment for CAPNON. Raising awareness and understanding of this rare entity is necessary for accurate diagnosis and management of patients affected by this condition [3)].

A 31-year-old woman was admitted to our hospital because of a high-density mass in the left frontal lobe, detected by computed tomography (CT) during a physical examination. She had a 3-year history of obsessive-compulsive disorder. We describe the imaging, histopathological, and molecular characteristics of the patient. This is the first report describing MA combined with CAPNON. We reviewed the literature on MA and CAPNON over the last decade and summarized the points for differential diagnosis and treatment. It is difficult to preoperatively distinguish between MA and

CAPNON. However, this coexisting condition should be considered when intra-axial calcification lesions are observed on radiological imaging. Accurate diagnosis and appropriate treatment are likely to benefit this patient group [4].

A 53-year-old man with a history of an untreated brain mass was taken to Toyama Prefectural Central Hospital by emergency transport. Computed tomography revealed an intracranial hypo-attenuated lesion exhibiting a mass effect. Several calcified foci were observed around the lesion. His radiographical diagnosis was meningioma with calcification and edema. He suddenly showed tonic seizure after admission; therefore an emergency craniotomy was performed. However, he unfortunately died due to advanced cerebral edema. Microscopic findings of the surgically obtained materials were consistent with neurenteric cyst (NC). Intracranial hard masses were found adjacent to NCs, and the masses were composed of a fibrous cartilage-like matrix with extensive linear calcification and the presence of surrounding round-to-oval epithelioid cells. Thus, calcifying pseudoneoplasm of the neuraxis (CAPNON) associated with NC was considered the most appropriate diagnosis of the present case. To the best of our knowledge, this is the first report of such a case. The present case suggests that delay of treatment might cause a poor outcome, at least in CAPNON associated with NC. Careful investigations, including the underlying pathology, may be essential when considering the etiology of CAPNON and its treatment strategies [5].

A 36-year-old woman with a 20-year history of left-sided hearing loss that had recently progressed. Computed tomography and magnetic resonance imaging showed a heterogeneous calcified lesion with vasogenic edema and a perilesional cyst in the left middle cerebellar peduncle. Although it is a rare radiographic feature of CAPNON, vasogenic edema should be included as a possible feature of this uncommon tumefactive lesion [6].

A 37-year-old woman with hypopituitarism was found to have an atypical sellar mass with slow growth on interval imaging. The lesion was debulked via a microscopic endonasal transsphenoidal approach and found to be a calcifying pseudo neoplasm of the neuraxis (CAPNON).

Lessons: CAPNON is a rare disease entity that may affect the sellar region. CAPNON should be on the differential diagnosis for sellar masses that are associated with T1 and T2 hypointensity on magnetic resonance imaging with minimal enhancement. Although CAPNON is not at risk for malignant progression, these benign lesions can continue to grow after a subtotal resection and require follow-up [7]

Paolini presented the case of a male 17-year-old who presented with new-onset seizures. MRIs revealed a 2 cm extra-axial solid-cystic mass, arising at the left temporo-occipital junction and abutting the dura with marked surrounding parenchymal vasogenic edema. The solid components demonstrated dense calcification and avid enhancement. Gross total surgical resection was performed. Histopathological examination revealed central regions showing characteristic features of CAPNON. Toward the periphery, the CAPNON was intimately associated with and sharply demarcated from a meningioma, which showed up to five mitoses per 10 high-power fields and had invasion into

the adjacent brain parenchyma, warranting a WHO grade II designation. This is the first report of CAPNON arising in association with a meningioma. The coexistence of these two tumors raises the possibility of a reactive/dysplastic etiology for CAPNON [8].

Duchesne et al., present a case that had the peculiarity to occur in a pregnant woman. At 32 weeks of gestation, a 26-year-old woman was hospitalized to explore nocturnal epigastralgia. During the hospitalisation, the patient presented generalised seizures. As an eclampsia had been suspected, a caesarean delivery was performed. Post-operatively, the patient harboured memory disorders and neuro-imaging explorations were done. They showed an intracerebral calcified mass located in the left frontal lobe and surrounded by an oedema. A complete surgical resection was performed. Histological examination of the surgical specimen showed a calcified tissue containing a fibrillary or granular material. A dense and hyalinised eosinophilic material focally surrounded the calcifications and contained regular fusiform cells of fibroblastic type. Foci of lipomatous and osseous metaplasia were present. Immunohistochemical staining for EMA and STAT6 was negative. There was no associated meningioangiomatosis nor tumour proliferation. Forty-five months after surgery, the patient did not present any seizures and had no sequelae. CAPNON are rare lesions occurring at any age. Their location in the central nervous system is ubiquitous and they can be intra or extra axial. The treatment is surgical and the prognosis excellent. CAPNON must be recognized and distinguished from the other calcified lesions, tumoural or non-tumoural, to avoid an inadequate and potentially harmful treatment [9].

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