Clinical Neurology and Neurosurgery 143 (2016) 116–120

Contents lists available at ScienceDirect

Clinical Neurology and Neurosurgery

journal homepage: www.elsevier.com/locate/clineuro

Calcifying pseudoneoplasms of the neuraxis: Report on four cases and review of the literature

Sara García Duque*, Diego Medina Lopez, Ana Ortiz de Méndivil, Jorge Diamantopoulos Fernández

Madrid Norte Sanchinarro Hospital, Oña 10, 28050 Madrid, Spain

ARTICLE INFO

Article history:
Received 30 December 2015
Received in revised form 4 February 2016
Accepted 18 February 2016
Available online 23 February 2016

Keywords:
Calcifying pseudoneoplasms of the neuraxis
CAPNON
Neuroaxis
Fibrocalcifying
Calcifying pseudotumour

ABSTRACT

Objectives: Calcifying pseudoneoplasms of the neuraxis (CAPNON) are rare lesions occurring anywhere in the central nervous system (CNS). Since their description, only 55 cases have been reported. We present the largest series reviewing their imaging features, histology and potential origins. Patients and 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. Autopsy patients were excluded.
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. A total of retrospective 30 articles were selected: 10 case series (33.33%) and 20 reports of single cases (66.66%). The 30 articles and our additional cases added up to a total of 27 patients with spinal CAPNON and 32 patients with intracranial CAPNON. All patients were treated surgically. A follow-up, conducted in 48 patients, showed no signs of recurrence in 46 of the 48.
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.

1. Introduction

Calcifying pseudoneoplasms of the neuraxis (CAPNON) are rare, non-neoplastic, calcified lesions of the central nervous system (CNS). Since the original pathologic description by Rhodes and Davis [1], approximately 55 cases have been reported. Little is known about the aetiology, natural history or course of the disease in surgically treated patients. It is important to recognise this entity, as it is a slow-growing lesion with a good prognosis; doing so will allow avoiding further examinations and inappropriate treatment [2,3]. Although the pathogenesis of this lesion is uncertain, it appears to be reactive rather than neoplastic [4,5].

We present four additional cases of calcifying pseudoneoplasm, the largest series of non-autopsy cases in the literature, reviewing the imaging features, histology, and potential origins.

1.1. Patients and methods

The case histories of four patients with histopathologically verified diagnoses of CAPNON are presented. Moreover, we reviewed all cases of patients with CAPNON published so far. For this, we searched for the abstracts and titles of all articles in MEDLINE (between 1977 and December 2014) with the following keywords: calcifying pseudoneoplasm; calcifying pseudotumour; and

* Corresponding author.
E-mail addresses: saragduque83@hotmail.com (S.G. Duque), diegoneuro@gmail.com (D.M. Lopez), aomendivil@yahoo.es (A.O. de Méndivil), jorgediaman@yahoo.es (J.D. Fernández).

http://dx.doi.org/10.1016/j.clineuro.2016.02.025
0303-8467/

fibro-osseous lesion. Only studies and reports about patients with CAPNON in English and Spanish were included; and autopsy reports were excluded. From the qualifying articles; the following parameters were collected: study type; number of patients; anatomical area (intracranial; spinal or both); clinical and radiological presentation; therapy; follow-up; incidence and type of complication and outcome. In the review; intracranial and spinal CAPNON were analysed separately. Furthermore; current recommendations for the management of spinal and intracranial CAPNON are discussed.

1.2. Case report 1

A 48-year-old female patient presented with a 1-year history of progressive, worsening headaches. Computed tomography (CT) scans ordered by her neurologist showed a densely calcified mass in the left atrium. Magnetic resonance imaging (MRI) revealed a hypointensity lesion on both T1- and T2-weighted images, with no vasogenic oedema, indicating a cavernoma (Fig. 1). Given the accessibility of lesion and the possibility of bleeding, we performed a complete resection. The lesion was successfully resected via an occipital craniotomy without any complications. Intraoperatively, the lesion was noted to have a significant calcified component. Pathological examination of the mass revealed a predominantly hypocellular basophilic calcified matrix, surrounded by palisading epithelioid cells. A chondroid matrix was present along with abundant fibrovascular stroma and a focal area of osseous metaplasia. Postoperatively, the patient recovered well without any neurological deficits at the 24-month follow-up.

1.3. Case report 2

A 51-year-old female patient with previous sacroiliitis presented with more than three months' history of lower back pain radiating to both legs. MRI showed a hypointensity lesion on both T1- and T2-weighted images, with no vasogenic oedema in the posterior epidural space at L2. A CT scan was performed and it was confirmed as a calcified lesion. After a laminectomy of L2, a gross total resection was done. The mass was firm and adherent to the dura mater. Histologically, epitheloid cells in a granuloma-like pattern, fibrocellular stroma with spindled fibroblastic cells, and calcified materials were found. These pathological findings confirmed a CAPNON. Postoperatively, back pain and radicular pain were resolved. A follow-up MRI at 12 months after surgical resection confirmed no recurrence, and symptoms continued to be almost completely resolved at that time.

1.4. Case report 3

A 46-year-old female patient presented with a one-year history of posterior neck pain. CT scans of the cervical spine showed a prominent central-to-right side calcified intraosseous mass with a distinctly increased density at the body of C3. Laminectomy of C3 and total removal of the mass were performed. After mass resection, fixation was performed from C2 to C4. Pathological findings indicated a typical chondromyxoid matrix in a nodular pattern with palisading spindles and epithelioid and scattered psammoma bodies, all of them consistent with a CAPNON lesion. Symptoms resolved after surgical resection, and there was no recurrence at 27-month follow-up.

1.5. Case report 4

A 73-year-old male patient with six-month history of progressive paraparesis consulted the department of neurology. A cervicodorsal MRI scan was performed revealing a well-circumscribed intradural extramedullary hypointensity lesion on both T1- and T2-weighted images in the posterior part of T2. In the subsequently performed CT scan, we confirmed the existence of calcium in the lesion. Because of the progressive nature of the symptoms, we performed a T2 laminectomy with a total resection of the lesion. Histopathological examination revealed a chondroid matrix with abundant fibrovascular stroma and a focal area of osseous metaplasia, which confirmed the existence of a CAPNON. Postoperatively, the motor weakness regressed completely and there was no recurrence at 12-month follow-up.

2. Results of literature review

We identified 364 possibly relevant publications. After application of the inclusion criteria, a total of 30 articles were selected: 10 case series (33.33%) and 20 reports of single cases (66.66%). The 30 articles and our additional cases added up to a total of 27 patients with spinal CAPNON and 32 patients with intracranial CAPNON. All patients were treated surgically. A follow-up, conducted in 48 patients, showed no signs of recurrence in 46 of the 48.

Intracranial CAPNONs have been documented in the literature in 32 cases, including those in this paper (Table 1). The ages of the affected individuals show a wide range (from 6-to 67-years-old), and there were 19 male (59.38%) and 13 female patients (40.63%). Twenty cases (62.50%) were located supratentorially, and of these, 8 cases (25%) affected the temporal lobe. The CAPNONs of 12 patients (37.50%) were located in the posterior fossa.

The mode of presentation included epileptic seizures in 9 (28.12%) headache in 9 (28.12%), and cranial nerve affection in 5 (15.62%). Further symptoms reported included vomiting, papilloedema, pituitary dysfunction and developmental delay, dizziness, tinnitus, hallucinosis, decreased hearing, limb paresis, cranial nerve affection, and jugular foramen syndrome. Complete resection of the lesion was possible in 22 of the 32 cases (68.75%). Incomplete resection was performed in ten cases (31.25%). Revision surgery was necessary in one patient with near complete removal of the lesion in the left cerebellopontine angle. The follow-up period was reported in the cases of 22 patients (68.75%), with a median of 57.9 months (range: 2–360 months). At the last follow-up, 21 patients (95.45%) were recurrence-free. The condition was stabilised in the cases of 13 patients who underwent complete resections and in the cases of 8 who underwent incomplete resections, however, one incomplete resection led to disease recurrence and subsequent debulking surgery three years after the initial surgery.

Spinal CAPNONs have been reported in 27 cases: fifteen male (55.55%) and twelve female patients (44.45%) with a mean age of 53.96 years (range: 12–90 years) were available for analysis (Table 2). The most common location of the spinal CAPNONs reported was the cervical spine (9 cases, 33.33%). The spinal CAPNONs were located epidurally in 22 cases (81.48%), intradurally in 4 cases (14.81%), and intraosseously in 2 cases (7.41%). The clinical presentation included diffuse neck or back pain (or both) in 13 cases (48.15%) and muscular paresis in 7 cases (25.92%). Complete excision of the lesion was performed in 13 cases (48.15%); in one patient (3.70%), a single level laminectomy for an epidural CAPNON was performed, but the authors do not specify the degree of resection. The median follow-up for those cases in which one was reported (16 of the 27 cases, 59.25%) was 38.43 months (range: 2–112 months). While 15 of those16 patients were reported recurrence-free (93.75%), one patient with an incomplete excision showed disease recurrence with local progression at the 24-month follow-up.

3. Discussion

CAPNONs are rare lesions: only 59 cases have been reported, and this is the longest series of cases in the literature. Six