Original article

Eighteen cases of crowned dens syndrome: Presentation and diagnosis

Syndrome de la dent couronnée : à propos de 18 cas

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A B S T R A C T

Background and purpose. – Crowned dens syndrome is an ill-known etiology of acute neck pain.
Methods. – We carried out a retrospective study of 18 cases of patients with crowned dens syndrome, assessing clinical and radiological features.
Results. – The results of our study are comparable to data from the literature. The clinical presentation of acute febrile neck pain, occipital headache and multidirectional stiff neck especially affects women aged over 60. No predisposing factor was recognized. However, a history of peripheral joint chondrocalcinosis may reinforce the diagnosis. In more than 50% of cases, laboratory tests showed a marked inflammatory syndrome. The diagnosis was obtained with cervical CT-scan focusing on the C1/C2 joint. This gold standard test was able to show a calcification of the cruciform ligament in connection with deposits of calcium pyrophosphate crystals in almost 80% of cases. Other imaging tests provided little information, including standard radiographs of the cervical spine. MRI can eliminate some differential diagnoses such as infections or neurological emergencies. Complications are infrequent. The standard treatment is based on anti-inflammatory drugs (NSAID, colchicine) or corticosteroids. These treatments are highly effective: a dramatic full recovery of cervical mobility may be observed within 48 hours. In over half of cases, a different diagnosis was initially made, responsible of unnecessary additional tests and treatment.
Conclusion. – A comprehensive consultation, a complete clinical examination and a precise analysis of the imaging will avoid certain investigations and rule out differential diagnoses.

R É S U M É

Introduction. – Le syndrome de la dent couronnée est une étiologie peu connue des cervicalgies aiguës fébriles. L'objectif de ce travail est de rapporter les caractéristiques clinique et radiologique de ce syndrome à partir d’une série.
Méthodes. – Il s’agit d’une étude rétrospective monocentrique regroupant les cas de patients hospitalisés ou ambulatoires pour lesquels le diagnostic de syndrome de la dent couronnée a été retenu.
Résultats. – Dix-huit cas sont recensés et analysés. Cette atteinte touche préférentiellement les femmes (11/18), et les sujets d’âge avancé (supérieur à 75 ans : 11/18). Le tableau clinique complet, cervicalgies aiguës fébriles, céphalées occipitales et raideur cervicale multidirectionnelle, est retrouvé dans près de 70 % des cas. Il n’existe pas de facteur prédisposant particulier ; il est cependant nécessaire de rechercher la présence d’une maladie métabolique sous-jacente. Dans plus de 50 % des cas, la biologie objective un syndrome inflammatoire marqué. Le diagnostic de certitude est obtenu grâce au scanner cervical centré sur la charnière C1/C2, qui permet dans tous les cas de mettre en évidence une calcification du ligament cruciforme évocatrice de dépôts de cristaux de pyrophosphate de calcium dans près de 80 % des cas. Les autres examens d’imagerie sont peu contributifs, notamment les radiographies standards du rachis cervical. L’IRM permet d’éliminer certains diagnostics différentiels infectieux ou neurologiques urgents.

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1. Introduction

Crowned dens syndrome was first described in 1974 by Dirheimer and Wackenheim and represents an ill-known etiology of febrile acute neck pain [1]. This syndrome can cause misleading symptoms, which leads to diagnostic difficulties. The clinical presentation includes febrile neck pain, headache and neck stiffness. Neurological complications are rare. With appropriate treatment, symptoms may improve very quickly, leading to a full recovery of cervical joint mobility. The aim of this study was to describe the clinical presentation of the syndrome and how imaging was used to diagnose it in a single center series.

2. Methods

We carried out a retrospective observational study of patients who were treated in our department between 1990 and 2011, and whose final diagnosis was “crowned dens syndrome”.

3. Results

Eighteen cases were recorded and are summarized in Table 1. Most of the patients affected (61% of all cases) were women (11 women to seven men) and were aged over 60 on average (median 75.5 years, range: 53–88). Examinations found a history of chondrocalcinosis in only one case.

Clinically, the diagnostic triad (neck pain, stiff neck and headaches) was present in 13 out of 18 cases (72%). In one patient these clinical features were not found. In four patients, neck pain had been present for several months or more than a year and in seven patients fever was absent (38.8%).

No neurological or cardiac abnormalities were found in our patients. Two patients presented vestibular dizziness and nausea. In six cases (33%) these symptoms were associated with bilateral shoulder pain and occipital neuralgia.

From a biological point of view, a marked inflammatory syndrome was observed in over half the cases (11 out of 17.64%, data missing for one patient) with a mean CRP levels of 82.3 mg/L (23–275) at admission. In six cases inflammation was absent. Neutrophilic leukocytosis was observed in two cases. Blood cultures positive for Staphylococcus aureus that was sensitive to methicillin were observed in one case. For this patient there was a specify CT-scan and a normal cervical spinal MRI.

A cervical bone CT-scan, in axial and sagittal views, confirmed the suspected diagnosis in 100% of cases. In two cases, MRI revealed the calcifications. The test (MRI) was conducted because cervical spondyloarthrosis was suspected.

In 44% of cases (8 out of 18) calcifications were located on the transverse ligament of the atlas and the alar ligament (Fig. 1). In three cases (16.6%) calcifications were mottled, and located at the postero-lateral surface of the dens (Figs. 2–4). In two patients, calcifications were also found in the knee menisci and in the ligament of the corpus.

Calcifications of calcium pyrophosphate were seen in 83% of the cases (15 of 18). A deposit of hydroxyapatite was found in three cases before the appearance of mottled heterogeneous calcification that disappeared on radiological imaging exams.

In over half of the cases (12 out of 18) other diagnoses were initially considered, such as: infectious cervical spondylodiscitis (n = 4), cervical spondylosis (n = 3), occipital neuralgia (n = 1), impingement syndrome (n = 1), Horton’s disease (n = 1), polymyalgia rheumatica (n = 2).

Looking at the data, we observed that a single treatment by oral corticosteroids was administered in two patients, and anti-inflammatory drugs (NSAID) and level 1 and 2 analgesics in six cases. One patient underwent cervical infiltration before admission.

Concerning treatment during hospitalization, 14 out of 18 patients received treatment with colchicine, associated with systemic NSAID or corticosteroids in five cases. Two patients were treated with NSAID alone.
Table 1
Series of 18 cases collected between 1990 and 2011.
Série de 18 cas colligés entre 1990 et 2011.

<table>
<thead>
<tr>
<th>Pat No/ date</th>
<th>Sex</th>
<th>Age</th>
<th>ATCD of CCA</th>
<th>Clinical signs: triad</th>
<th>Others clinical signs</th>
<th>PCR/ESR</th>
<th>Leuko/Neutro</th>
<th>Radiological signs</th>
<th>Suspected diagnosis</th>
<th>TTT/Evolution</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/ 2006</td>
<td>F</td>
<td>85</td>
<td>Yes (crowned dens Sd)</td>
<td>No neurological signs</td>
<td>242/96</td>
<td>17.5/ 89%</td>
<td>MRI: thickening of the soft tissues periodontoidal CT-scan: axis CCA</td>
<td>Cervical spondylodiscitis</td>
<td>Hm: ANG and NSAID Hp: Colc.</td>
<td></td>
</tr>
<tr>
<td>2/ 2009</td>
<td>F</td>
<td>75</td>
<td>No</td>
<td>Right Arnold’s neuralgia</td>
<td>4/16 Ccal N</td>
<td>9.2/</td>
<td>CT-scan: dens posterior Lgt calcification Rx: CCA knees, carp, symphys</td>
<td>Arnold’s neuralgia</td>
<td>Hm: INF + Colch + CTC bolus and relay per os</td>
<td></td>
</tr>
<tr>
<td>3/ 2002</td>
<td>F</td>
<td>78</td>
<td>No</td>
<td>No neurological signs</td>
<td>145/56 Sterile BC</td>
<td></td>
<td>CT-scan: alar Lgt calcification</td>
<td>Impingement sd</td>
<td>Hm: ANG HP: NSAID</td>
<td></td>
</tr>
<tr>
<td>4/ 2001</td>
<td>F</td>
<td>61</td>
<td>No</td>
<td>No neurological signs, no endocarditis signs</td>
<td>64/70 No leuko</td>
<td></td>
<td>CT-scan: longitudinal Lgt calcification</td>
<td>MRI: osteochondroma</td>
<td>Hm: CTC Hp: Colch</td>
<td></td>
</tr>
<tr>
<td>5/ 2003</td>
<td>H</td>
<td>77</td>
<td>No</td>
<td>Subacute neck pain No fever Right cervical limitation Yes but afebrile</td>
<td>No pyramidal Sd Right shoulder pain 14/4 Hyper IgM</td>
<td>No leuko</td>
<td>Rx and CT-scan: anterior dens calcification SASM cervical spondylodiscitis</td>
<td>SASM cervical spondylodiscitis</td>
<td>Hm: ANG + infilt Hp: Colch</td>
<td></td>
</tr>
<tr>
<td>6/ 2010</td>
<td>H</td>
<td>74</td>
<td>No</td>
<td>No neurological signs No endocarditis signs</td>
<td>275/ BC + a SASM Sterile urine culture</td>
<td>No leuko</td>
<td>CT-scan: longitudinal Lgt calcification</td>
<td>MRI: osteochondroma</td>
<td>Hm: CTC Hp: Colch</td>
<td></td>
</tr>
<tr>
<td>7/ 2009</td>
<td>H</td>
<td>85</td>
<td>No</td>
<td>Yes lasting for 2 months Antalgic attitude in anteflexion or reducible No neurological signs</td>
<td>127/</td>
<td>9.1/</td>
<td>CT-scan: longitudinal Lgt calcification</td>
<td>MRI: osteochondroma</td>
<td>Hm: CTC Hp: Colch</td>
<td></td>
</tr>
<tr>
<td>8/ 1999</td>
<td>H</td>
<td>74</td>
<td>No</td>
<td>Neck pain since 1 year</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9/ 2010</td>
<td>F</td>
<td>78</td>
<td>No</td>
<td>Malaise, dizziness Normal neurological examination</td>
<td>48.8/ 50 No leuko</td>
<td></td>
<td>Rx: osteoarthritis CT-scan: periodontoid calcifications CT-scan: left periodontoid mottled calcifications (apat). Loss in CT-scan in a few months.</td>
<td>MRI: osteochondroma</td>
<td>Hm: NSAID</td>
<td></td>
</tr>
<tr>
<td>10/ 1990</td>
<td>F</td>
<td>62</td>
<td>No</td>
<td>No at the beginning then appear without fever</td>
<td>5/20</td>
<td>8.9/40%</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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<tr>
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<th>PCR/ESR</th>
<th>Leuko/Neutro</th>
<th>Radiological signs</th>
<th>Suspected diagnosis</th>
<th>TTT/Evolution</th>
</tr>
</thead>
<tbody>
<tr>
<td>11/2011</td>
<td>F</td>
<td>83</td>
<td>No</td>
<td>Yes</td>
<td>Normal neurological examination, shoulder pain</td>
<td>112/7.4</td>
<td>CT-scan: transverse Lgt of C1 calcifications</td>
<td>Hm: NSAID in intramuscular Hp: Colch</td>
<td></td>
<td></td>
</tr>
<tr>
<td>12/2009</td>
<td>H</td>
<td>73</td>
<td>No</td>
<td>Yes</td>
<td>Normal neurological examination</td>
<td>23/6 Sterile BC 4.4/2.3</td>
<td>CT-scan periodontoid calcifications Infectious cervical spondylodiscitis Hp: Colch</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>13/2007</td>
<td>F</td>
<td>83</td>
<td>No</td>
<td>No, neck pain for 1 year, afebrile</td>
<td>Shoulder pain, decrease left temporal pulse, heart murmur unknown</td>
<td>162/73 Sterile BC 4.7/2.0</td>
<td>CT-scan periodontoid calcifications Infectious cervical spondylodiscitis-complicated by infective endocarditis Hp: Colch</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>14/2003</td>
<td>F</td>
<td>53</td>
<td>No</td>
<td>No, old neck pain stiffly straight</td>
<td>Normal neurological examination</td>
<td>2.7/29 5.9/2.9</td>
<td>CT-scan: transverse Lgt of C1 calcifications Cervical spondylodiscitis Hp: CTC Hp: Colch + NSAID</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>15/2004</td>
<td>F</td>
<td>64</td>
<td>No</td>
<td>Yes for 1 year, afebrile, no headache</td>
<td>Dizziness, nausea</td>
<td>1/8 14.6/11.5</td>
<td>CT-scan: calcifications Lgt front and right side of C2 Cervical spondylodiscitis Hp: ATG Hp: cervical INF under CT + CTC bolus, Colch</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>16/2011</td>
<td>H</td>
<td>79</td>
<td>No</td>
<td>Yes, afebrile, no stiff neck</td>
<td>Normal neurological examination</td>
<td>9/38 4.9/2.4</td>
<td>CT-scan: calcifications longitudinal Lgt C1 Horton, Cervical spondylodiscitis Hp: ANG and CTC</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>17/2011</td>
<td>F</td>
<td>83</td>
<td>No</td>
<td>Yes</td>
<td>Normal neurological examination, shoulder pain, jaw claudication</td>
<td>157/80 7.4</td>
<td>CT-scan: periodontoid calcifications PR Hp: ANG and Colch</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>18/2011</td>
<td>H</td>
<td>88</td>
<td>No</td>
<td>Yes</td>
<td>Normal neurological examination</td>
<td>52/60 6.0</td>
<td>CT-scan: periodontoid calcifications PR Hp: Colch</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

NSAID: non-steroidal anti-inflammatory; ATCD: antecedent; ANG: analgesic; CTC: corticosteroids; INF: corticosteroids infiltration; Physio: physiotherapy; TTT: treatment; Colch: colchicine; Ccal: corrected calcium; N: normal; BC: blood cultures; Lgt: ligament; CCA: chondrocalcinosis; Leuk: leukocytosis; Apat: hydroxyapatite crystals; Hm: treatment received at home; Hp: treatment received during hospitalization; PR: polymyalgia rheumatica; IE: infective endocarditis.
In all cases, clinical improvement was observed within a few days with complete recovery of cervical mobility.

4. Discussion

Crowned dens syndrome, which is rarely described in the literature (fewer than 50 cases), is a significant cause of acute or subacute neck pain with fever.

Presentation is clinical, biological and radiological, and prevalence is around 2% in patients with acute neck pain [2,3]. However, this condition seems to be underestimated, as many cases go undiagnosed. Women are more often affected than men, as seen in our study, with a mean diagnosis age of 60 years [4].

No risk factors have been found, but diagnosis is easier in patients with a history of chondrocalcinosis.

From a pathophysiological point of view, a deposit of calcium pyrophosphate is observed. Deposition of hydroxyapatite is also observed and is characterized by mottled calcification, disappearing on imaging.

Clinical examinations usually show a combination of diagnostic features including acute or sub acute neck pain, multidirectional neck stiffness and occipital headache [4]. As observed in our series, fever is present only in 75% of cases. According to some studies, rotation of only 45° or less suggests involvement of the joint C1/C2 [1]. Some authors believe that a limitation in flexion-extension is more characteristic of meningeal syndromes [5,6]. The rest of the clinical examination, including the neurological examination, does not show any unusual features, as we observed in our study. Very rare cases of spinal cord compression have been found in the literature causing walking difficulties, a tetra paresis or even tetraplegia that progress slowly, mimicking cervical spondyloptic myelopathy [7]. Cervical MRI performed before the first-line clinical setting show periodontoid mass hyposignal T2, leading to a differential diagnosis with neoplastic lesions in particular (Fig. 5). These calcium deposits may cause spinal stenosis due to their size or spinal cord compression due to intradural extension and dural erosion. Surgical treatment is then necessary and consists in a combined resection of the lesion. It is often associated with significant neurological sequelae [8–10].

In cases of acute inflammation associated with neurological signs, even when the results of imaging give cause for concern, it may be lawful to begin test treatment with colchicines (or NSAID) as first-line treatment before resorting to surgery on the C1/C2 joint. This is especially true in elderly patients for whom surgery carries particular risks. Some patients in our study were initially sent to a neurosurgical department for acute neck pain, since medical test treatment was effective and surgery was therefore not required. Surgical treatment can be done by a previous cervical access “opened mouth” with a microscope or by nose endoscope process. This latter approach seems better nowadays. A simple dens’ resection is often enough with the ablation of ossifications around the dens. Sometime it is useful to complete by a posterior cervical spine procedure because a painful instability may happen an occipital cervical spine’ fixation is then performed.

Biological assessments show an inflammatory syndrome with no other specific characteristics. Examinations should look for any underlying metabolic disease (hemochromatosis, hyperparathyroidism, Wilson disease, etc) especially in patients aged under 50 [3].

Radiological assessments include bone-window cervical CT-scan focusing on C1/C2 in axial and coronal cuts. Our series reinforces this test as the gold standard for visualizing the calcifications, confirming the diagnosis in all cases of our series [11]. The calcium deposits usually affect the transverse part of the cruciate ligament or longitudinal fibers at the side. The alar ligament may
also be the deposition site. The appearance of calcification varies between linear, arcuate, clumps or crown. Goto's radiological classification lists the possible locations of these deposits in relation to the dens: posterior (50%), posterolateral (27.5%), circular (12.5%) older (5%) or lateral (5%) [2,12].

Other radiological examinations including standard radiographs of the cervical spine are less useful, even as open mouth views, as calcification are not usually visible due to the radiological superposition of atlas and axis. MRI is not able to make a specific diagnosis, but enables us to eliminate differential diagnoses [7]. On the other hand, the discovery of radiological calcium deposits in peripheral joints (knees, wrists and pubic symphysis) can be useful in the search for a diagnosis.

The numerous differential diagnoses that often lead to additional and unnecessary therapeutic tests are a major problem in crowned dens syndrome [2]. Infectious etiologies such as acute bacterial meningitis, emergency diagnoses and therapy or cervical spondylitis should be raised first due to neurological risks. In this situation, the physical examination is characterized by a neurological examination that is confusing due to cerebral etiologies [13]. In this case, lumbar puncture seems difficult to avoid. The symptoms are usually resistant to anti-inflammatory drugs. Bacteriological samples are used to confirm the infectious nature of the disease and identify the causative organism. Spinal inflammatory diseases (rheumatoid arthritis) may lead to febrile cervical pain due to the pannus around the odontoid. The patient's medical history and imaging (erosions) can confirm the diagnosis [1,14]. In this context, dislocation C1/C2 is another possible diagnosis. Other types of inflammatory vasculitis that can mimic Horton's disease may be misleading due to the stiff neck pain triggered. Brain imaging may help the diagnosis in such cases. Lastly, neoplastic causes should be ruled out. Primary bone cancer is rare, and metastatic sites are more often associated with a significant risk of spinal cord complications. Many other anecdotal causes exist, such as calcification of the muscles along the neck for which a radiograph of the cervical spine is enough to confirm the diagnosis or Grisel's syndrome: acute febrile neck pain following a viral infection of the upper airway [4,7]. In practice, a full and properly conducted investigation, a thorough clinical examination, including neurological and radiological tests and a precise analysis of imaging helps us to avoid invasive investigations and can eliminate a number of differential diagnoses.

According to data in the literature, treatment first involves the use of non-steroidal anti-inflammatory drugs that have an amazing effect within days. Colchicines, systemic corticosteroids or oral therapy may be used as alternatives. In our series, treatment with colchicines was used as the first-line treatment (14 out of 18 patients, 77.7%), and was usually associated with another anti-inflammatory therapy (steroids or non-steroids). The results show a good clinical efficacy.

A cervical CT-scan, although not indicated systematically, can be useful retrospectively to determine the pathophysiological cause when calcification disappears in cases of hydroxyapatite deposition.

The recovery of cervical spinal mobility is complete without sequelae. However, there is a slight risk of late complications such as erosion, spondylolysis or subluxation. Any future complications haven't been found in our series, as patient follow-up could not be carried out.

Crowned dens syndrome is a frequent cause of acute febrile neck pain and is often raised by rheumatologists. Because it is not well known in other areas of medicine including neurosurgery, it can often lead to misdiagnosis or unnecessary surgery on vulnerable patients.

To retain

- Syndrome caused by a calcium pyrophosphate deposit in the majority of the cases.
- Association diagnoses characteristic, cervical pain, stiffness of nape of the neck and occipital headache.
- Biologically: inflammatory syndrome without specify. Need to investigate for a metabolic disease under unclaimed among patients of less than 50 years.
- Radiologically: diagnosis thanks to a cervical bone window CT scan centred on C1/C2. Axial and coronal slides allow the identification of calcifications on the cruciform ligament.
- Many differential diagnoses to evoke: infectious, inflammatory, tumour...
- Treatment: anti-inflammatory drugs avoiding steroids in first intention, in the event of failure the colchicines or corticoids yield spectacular effectiveness. Complete recovery of cervical spine mobility without long-term side effects.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References