Crowned dens syndrome misdiagnosed as polymyalgia rheumatica, giant cell arteritis, meningitis or spondylitis: an analysis of eight cases

A. Aouba, V. Vuillemin-Bodaghi¹, C. Mutschler² and M. De Bandt²

Background. The crowned dens syndrome, related to microcrystalline deposition in the peri-odontoid articular and abarticular structures, is mainly responsible for acute or chronic cervical pain.

Patients. We report eight cases of crowned dens syndrome with atypical presentations mimicking giant cell arteritis, polymyalgia rheumatica, meningitis or discitis. The clinical and radiological aspects of these cases are presented and discussed.

Results. For all patients, fever, cervical stiffness, headaches and biological inflammatory syndrome were reported. For three patients, impairment of general condition, occipito-temporal or mandible pain and weakness with inflammatory pain of the shoulder girdle was suggestive of giant cell arteritis and/or polymyalgia rheumatica, leading to temporal artery biopsy and/or long-term steroid treatment. Recurrence of clinical symptoms when tapering steroids was noted. In two cases, previous breast carcinoma led to the initial diagnosis of metastatic spondylitis. For three patients with vomiting, nausea and Kernig’s and/or Brudzinski’s sign, the first diagnosis was meningitis, leading to unhelpful lumbar puncture. In all cases, diagnosis of crowned dens syndrome once evoked, was confirmed by cervical CT scanning and dramatic improvement with non-steroidal anti-inflammatory drugs or colchicine.

Conclusion. This under-recognized entity must be considered as a differential diagnosis of meningitis and discitis, but also of giant cell arteritis and polymyalgia rheumatica, as well as a possible aetiology for fevers of unknown origin. CT scanning is necessary for diagnosis. Clinicians should be aware of such misleading clinical presentations.

KEY WORDS: Crowned dens syndrome, Polymyalgia rheumatica, Giant cell arteritis, Calcium pyrophosphate dehydrate crystals, Hydroxyapatite.
**Patients 2, 3, 4 and 5**  
These four older patients (mean age 79 yr) presented with long-lasting and remitting cervical and shoulder pain and/or neck pain, with fever, weight loss, pain in the temporal (patients 3 and 4) or mandible (patients 2 and 3) area and morning stiffness (Table 1). Patient 3 had decreased pulsation in the temporal areas but no local inflammatory signs. All had elevated ESR (mean 105 mm in the first hour, range 78–150) and CRP (mean 120 mg/l, range 80–300, normal value <10). A working clinical diagnosis of GCA with PMR was considered for patients 2 and 3 but was not confirmed on histological examination of both temporal arteries. Both patients had received steroid therapy in expectation of the temporal biopsy. Patients 4 and 5 had a past history of breast and lung cancer (2 and 1 yr previously). A working diagnosis of cervical metastatic spondylitis was made, but X-rays and MRI of the cervical spine were normal. Tumour markers were within normal values. Patients 3 and 5 had a previous history of acute arthritis of the knees and wrist.

Joint X-rays showed tendinous calcific deposits of both shoulders in patient 2, a typical aspect of CCPD on both wrists in patient 3 and the knees in patient 5. All causes of arm weakness were ruled out (shoulder tendonitis, muscle disease, CPPD of the shoulders, diffuse osteoarthritis, etc.). In all cases, cervical CT scanning that was focused on C1/C2 allowed the diagnosis of CDS, showing a tiny linear calcification behind the odontoid process and diffuse cervical degenerative joint disease (Fig. 2). Spectacular efficiency of NSAID infusion (ketoprofen 200 mg daily) appeared within 2 days, without relapse after 10 days of treatment. A second attack of CDS appeared 14 months after the first (patient 3), with rapid resolution under the same treatment, then switched for colchicine.

**Patients 6, 7 and 8**  
Table 2 summarizes some clinical and biological data for these patients. All these patients complained of headaches, fever, sicknesses or vomiting, with cervical or shoulder pains for 5–10 days. Initial clinical examination found cervical stiffness with Kernig’s and/or Brudzinski’s sign. In each case, infectious meningitis was the diagnosis made. A lumbar puncture, performed for patients 6 and 7, was normal.

Further evaluation for patient 6 showed a diffuse radiological aspect, characteristic of chondrocalcinosis. In patient 7, the...
The diagnosis of CDS was made, associated with idiopathic CPPD with peripheral joint deposit (patients 6 and 8) and haemochromatosis (patient 7). Cervical CT scanning confirmed the radiological crowned dens in each case (a tiny half-hoop-shaped calcification surrounding the odontoid process; calcifications on the top of the odontoid, subchondral geodes and prominent cysts on the dens) (Figs. 3, 4 and 5).

In all cases, treatment with ketoprofen infusions for 3 days followed by oral administration for 10 days led to a dramatic clinical improvement in 3 or 4 days, with normalization of biological parameters in 10–14 days, allowing a definitive diagnosis of CDS.

**Discussion**

We report on eight patients with an atypical clinical presentation of CDS.

The diagnosis of CDS is based on the association of clinical, biological, radiological and therapeutic signs: acute periodic attacks of cervico-occipital stiffness and feverish pains with biological inflammatory syndrome, radiological identification of periodontal calcifications due to microcrystalline deposits on the retro-odontoid ligament and dramatic resolution of symptoms under treatment with NSAIDs or colchicine. This typical description was reported for the first time in 1980 [1, 2, 3, 4]. Authors have described acute attacks of pain in high cervical areas that are different from the chronic or subacute manifestations of microcrystalline deposition in the lower part of the cervical or dorsal and

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**Table 2. Clinical and biological manifestations of patients 6, 7 and 8 (meningitis)**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (yr)/sex</th>
<th>Fever (°C)</th>
<th>ESR (mm 1st h)</th>
<th>CRP (mg/l)</th>
<th>Leucocytes/mm³</th>
<th>Lumbar puncture</th>
<th>Other signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>6</td>
<td>74/M</td>
<td>38.5</td>
<td>45/71</td>
<td></td>
<td>11 500</td>
<td>Normal</td>
<td>Wrist arthritis</td>
</tr>
<tr>
<td>7</td>
<td>67/M</td>
<td>38.8</td>
<td>65/165</td>
<td></td>
<td>4 500</td>
<td>Not done</td>
<td>Hamochromatosis</td>
</tr>
<tr>
<td>8</td>
<td>87/M</td>
<td>39.0</td>
<td>50/227</td>
<td></td>
<td>14 000</td>
<td>Normal</td>
<td>Knee arthritis</td>
</tr>
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</table>
lumbar spine, which are often confused with common degenerative processes.

The triad of headaches, fever and cervical morning stiffness evokes either infectious meningitis leading to lumbar puncture, or of cervical metastatic spondylitis in the case of a medical history of cancer. As illustrated by our patients, CDS can mimic and lead to misdiagnosis of meningitis or cervical spondylitis.

When cervical stiffness is associated with pain in the shoulder girdle and jaw claudication, CDS can mimic PMR and/or GCA. Some authors [5] have reported chondrocalcinosis among a wide spectrum of rheumatic diseases that can mimic PMR because of constitutional symptoms, but not especially in the form of CDS.

Our three patients fulfilled at least three diagnostic criteria out of the Bird criteria for PMR. This and the lack of histological diagnostic criteria for PMR explain why this diagnosis was considered initially. But our patients also fulfilled the ACR 1990 criteria [6], allowing us to perform temporal artery biopsy for a strict diagnosis of GCA. Authentic biopsy-negative GCA [7] is recognized by the ACR 1990 criteria. For this biopsy-negative group, Gonzales-Gay et al. [8] found that PMR and headaches were the most frequent manifestations. In contrast, jaw claudication, abnormal temporal artery, visual complications and constitutional syndrome were less common than in biopsy-proven GCA, except for the presence of constitutional syndrome, as in our patients. Two of our patients fulfilled criteria for biopsy-negative GCA: this raises the question: are some biopsy-negative GCAs really GCAs or under-recognized CDS? The practical aspect of this is obvious, given the long-lasting treatment with steroids in the case of GCA. One answer would be to perform a CT scan of the cervical spine in the case biopsy-negative GCA, combined with test treatment with NSAIDs.

As this study shows, some atypical cases of CDS can be difficult to distinguish from PMR/GCA. We did not find in the literature any clinical sign allowing us to distinguish between CDS and PMR (or GCA): pain [9–12], neck stiffness [1, 2, 13], mandible signs [14–16], constitutional symptoms [8, 17] and the presence of peripheral arthritis [18] were not helpful in discriminating the two diseases. The only sign we did not observe in CDS was painful stiffness of the pelvic girdle suggesting that this could be discriminating between CDS and PMR.

X-rays may show densities in the odontoid process area, but are most often normal or do not allow the determination of which anatomic structure is involved. X-rays of the knees and wrists should be performed systematically; such calcifications remain asymptomatic but help in the clinical approach [19].

CT scanning focusing on C1/C2 is the gold standard of the diagnosis. This makes it possible to identify the anatomical substratum of CDS. If the typical tiny half-loop form of calcification behind the dens corresponds with certainty to the transverse ligament of the atlas (Figs 1 and 2), calcifications of other anatomical structures surrounding the top and the sides of the odontoid process are described in association with the clinical picture of CDS [3, 4]. According to some authors [1, 20, 21, 22], the definition of CDS should be extended to all the calcifications involving the synovial membrane, the articular capsule, the occipito-transverse ligament and/or the transverso-axial ligament that surrounds the dens. In our experience, CT scanning performs better than MRI in assessing calcifications of the dens area. Another useful aspect of CT scanning is the ability to rule out differential diagnoses of CDS such as unrecognized odontoid fractures or cervical cord compression, but it is important to keep in mind that CT scanning may fail to detect true CDS. This occurs when CT scanning is performed tardily after the acute attack, because the calcifications may be resorbed; hence the interest in repeating it [4, 17, 20].

We observed a dramatic clinical improvement within 5 days under NSAID or colchicine treatment. This allowed us to make the definitive and positive diagnosis of CDS.

The time that elapses before complete clinical recovery occurs is difficult to determine from the literature, as the data are often imprecise. Some authors have described clinical recoveries within 4 days to 3 weeks [4, 23], but the time of the initiation of NSAID treatment was not always given. So it appears that the delay before total recovery under treatment may be longer than in our patient. The dramatic initial improvement, followed by a second phase of slower but persistent improvement, seems more important than the total duration of improvement itself. Thus, it seems that, as for others microcrystalline joint involvement, NSAIDs appear to be the gold standard treatment of CDS; colchicine may also be prescribed, but it usually seems not as effective as in acute CPPD and its effects seem less predictable [24]. As rapid improvement with full recovery in a few days under steroid treatment is criteria for PMR, the test with NSAIDs could offer advantages over steroids in case of doubt. A positive (even incomplete) effect of NSAID treatment could be minor criteria for CDS. Long-term treatment must be discussed if their are recurrent attacks or subacute disease; low doses of oral colchicine have been shown to decrease the number of attacks [24]. Steroid therapy may be prescribed at low doses [24] in cases of severe diseases unresponsive to NSAIDs.

The occurrence of relapses in PMR patients could be the opportunity for clinicians to look for CDS, if it was not detected at first presentation. Clinicians should also consider the diagnosis of relapse of PMR only in the presence of at least one clinical sign and elevation of the acute-phase proteins in order to avoid unnecessary prolonged steroid treatment in elderly people suffering from other diseases than PMR, such as CCPD/CDS.

Conclusion

The spectrum of CDS is widening. We would like to emphasize that other manifestations, such as meningism (evoking meningitis or spondylitis), cervicobrachial pain (with shoulder weakness and stiffness) and occipital and temporal headaches (evoking atypical polymyalgia rheumatica and/or GCA) must be added to the classical acute feverish cervical pain. Some cases of prolonged evolution with relapses can mimic fever of unknown origin. The rapid diagnosis of CDS, based mainly on a cervical CT scan, can prevent invasive, expensive and useless investigations as well as a long course of potentially dangerous and inadequate treatments, notably with cortisone, or prolonged hospitalization in old patients.

<table>
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<tr>
<th>Rheumatology</th>
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<td>The crowned dens syndrome (microcrystalline deposition in the peri-odontoid articular structures) is responsible for cervical pain.</td>
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<td></td>
<td>Some atypical presentations are misleading and can mimic meningitis, polymyalgia rheumatica or giant cell arteritis.</td>
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The authors have declared no conflicts of interest.

References


