

# Crowned Dens Syndrome Occurred in a Patient after Simple Drainage for Chronic Subdural Hematoma: A Case Report

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Crowned dens syndrome (CDS) is a rare disease which presents with neck pain and rigidity. A 74-year-old man with right chronic subdural hematoma (CSDH) underwent hematoma drainage. After the operation, he complained of neck pain and laboratory test revealed elevation of C-reactive protein (CRP) and white blood cell (WBC). Suspecting localized infection, wound irrigation was performed. Neck pain relieved after irrigation, but we could not find the source of infection. CDS was diagnosed by computed tomography (CT). CDS is frequently misdiagnosed as meningitis and localized infection. CT is useful for diagnosis. Neurosurgeons need to be aware of CDS after operation.

**Keywords:** chronic subdural hematoma; crowned dens syndrome; neck pain

## Introduction

Crowned dens syndrome (CDS) is a relatively rare orthopedic disease that typically causes severe neck pain and limitation of neck rotation in the elderly due to deposition of calcium pyrophosphate dehydrate (CPPD) crystals or hydroxyapatite at cervical spine.<sup>1)</sup> The clinical manifestation of CDS is similar to that of meningitis and thus may result in misdiagnosis.<sup>2)</sup> CDS is not familiar to neurosurgeons. There are some previous reports about poststroke CDS, but no report on a patient with CDS after neurosurgical procedure has been reported so far.<sup>3)</sup> We report herein a rare case of a patient with CDS that occurred after simple drainage for chronic subdural hematoma (CSDH).

## Case Description

### Onset

A 74-year-old Asian male complained of a headache that had lasted for a month and was performed a medical checkup. Head Magnetic resonance imaging (MRI) showed right CSDH. He had no previous history of head trauma. Neurological examination at the time of initial presentation revealed no other apparent abnormal findings.

### Preoperative radiological and laboratorial findings

CT imaging showed iso density CSDH with a thickness of 13 mm on his right convexity, and a midline shift (MLS) of 3 mm (Fig. 1A) in consequence.

Laboratory testing showed C-reactive protein (CRP) of 0.49 mg/dl and white blood cell (WBC) count of 7,800/ $\mu$ l. Although the hematoma did not seem to compress the brain severely, his clinical manifestation of headache was severe, and the patient preferred to undergo surgical treatment. Thus, an operation was planned.

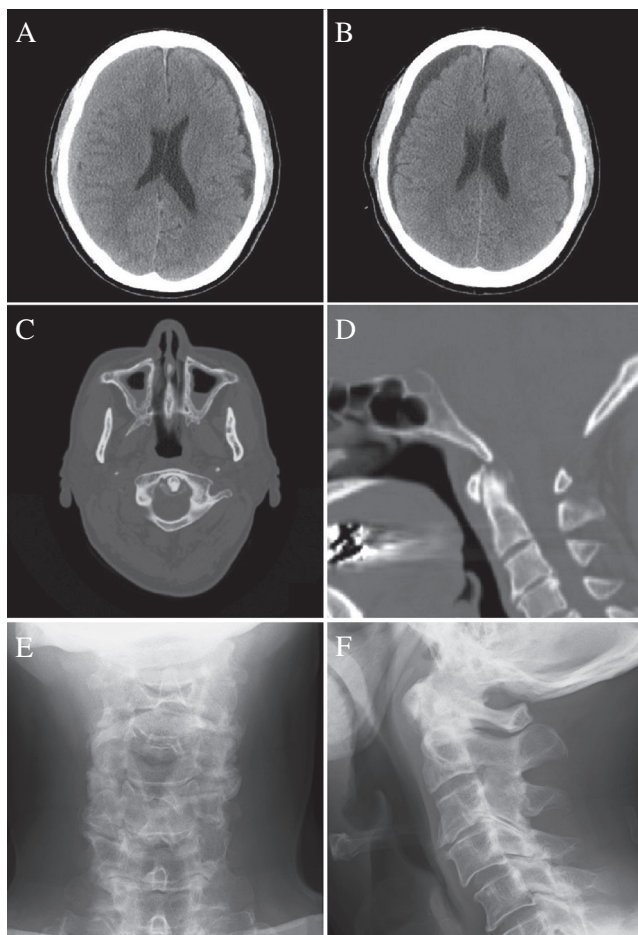
### Operation and postoperative course

We performed simple drainage to the patient (one burr hole procedure without irrigation, using a closed drainage system.<sup>4)</sup> A drainage tube was placed into the hematoma cavity for a day. The hematoma was drained (200 ml) and computed tomography (CT) performed the day after the operation showed that 80% of the hematoma was removed. The MLS was improved (Fig. 1B).

After the operation, his headache relieved. However, one day after the operation, the patient complained of neck pain. Physical examination showed that the mobility of his neck was restricted especially in the horizontal direction. The symptom was initially not severe and was observed conservatively. However, a laboratory testing performed 3 days after the operation showed CRP of 15.5 mg/dl and WBC count of 10,700/ $\mu$ l. The blood level of procalcitonin was under the threshold of meningitis. The symptom continued to get worse, and 5 days after the operation, the ranges of motion of his neck and shoulder became severely restricted. Slight redness at the wound site was observed at the same time. He had no fever and did not experience lassitude.

Although meningitis was not completely denied, lumbar puncture was not performed, because it has a risk of bringing bacterium inside the body as well as a risk of exacerbating subdural hematoma. At the same time, we had a slight impression that this symptom was not caused by meningitis because his neck rigidity was strong in the horizontal direction, which was not typical for meningitis. The patient did not complain of lassitude either. Instead, because a slight redness was seen at the wound site, we decided to open the surgical site again to search for surgical site infection. The same wound was re-opened, the burr hole cap was removed, and the surgical site was irrigated. No signs of infection were identified during the operation.

His neck pain was partially relieved immediately after the operation. The elevation of serum WBC and CRP did not improve immediately. MRI of head and neck was performed in search for subdural abscess and other infections, but no abnormal finding was detected. At this point, CDS was suspected. Non-enhanced CT scan of the neck showed a high-density area in a crown- or halo-like distribution around the



**Fig. 1** (A) Axial plain CT scan at the time of admission shows right chronic subdural hematoma with thickness of 13 mm and MLS of 3 mm. (B) Axial plain CT scan after the operation shows the improvement of MLS. (C) Axial cervical bone CT scan shows calcifications behind the odontoid process suggesting CDS. (D) Sagittal cervical bone CT scan shows calcifications of the transverse ligament of the atlas at C1/2 level. (E) Calcification at C1/2 level could not be detected from frontal cervical radiograph. (F) Calcification at C1/2 level could not be detected from lateral cervical radiograph.

odontoid process that demonstrated atlantoaxial synovial calcifications (Fig. 1C, D). Although not specific, this image was compatible with the image of CDS. Calcification at C1/C2 levels could not be detected from the frontal and lateral radiographs. (Fig. 1E, F). In conjunction with clinical findings, we made a diagnosis of CDS. Nonsteroidal anti-inflammatory drug (NSAID) was administered, and the symptom improved gradually. The patient was discharged 10 days after the initial simple drainage without any neurological sequelae.

## Discussion

CDS is a relatively rare clinical disease typically revealed with acute onset of neck pain and stiffness of the cervical spine.<sup>5</sup> CDS was named by Bouvet et al. in 1985,<sup>6</sup> and some other cases of CDS have been reported since then.<sup>5,6</sup> CDS usually occurs in patients over 60 years of age and has a female predominance.<sup>1</sup> The underlying reason for CDS is still controversial. One theory is that chondroid cell in

fibrous cartilage structures in transverse ligament around the odontoid process causes the disease. As for differential diagnoses, infectious or other inflammatory diseases such as abscess, meningitis, giant cell arteritis (GCA), polymyalgia rheumatica (PMR), calcific tendinitis of the longus colli and vertebral osteomyelitis should be considered from their symptom of acute neck pain in conjunction with elevated WBC and CRP.<sup>1,2,7-10</sup>

Although the cerebral fluid examination is the most reliable method to exclude meningitis, symptoms and radiographic images are also useful. Patients of meningitis often complain of stronger lassitude than patients of CDS. Neck rigidity of meningitis is generally identified in an anteroposterior direction, but in contrast, neck rigidity of CDS is identified in a horizontal direction. Gadolinium enhanced MRI sometimes shows thickening of meninges in patients with meningitis.<sup>2,8</sup> As for PMR, the pain usually involves breech and crural area. In contrast, CDS may involve pain at the arm. While CDS can be treated with NSAIDs, this treatment is not effective for PMR, which requires steroid for treatment. In addition, PMR takes a relatively chronic clinical course in contrast to CDS. PMR is sometimes complicated with GCA, which causes temporal headache. However, abnormal calcification at C1/C2 levels does not appear in this disease.<sup>2,9,10</sup> Calcific tendinitis can be excluded from its location of calcification, which is seen on the lateral tendon of longus colli.<sup>9,10</sup> As for vertebral osteomyelitis, it rarely occurs at the upper cervical portion, and MRI shows abnormal intensity at the vertebral bodies.<sup>2,9,10</sup> Among the various diagnostic tools, CT scan is the most useful in detecting calcification of CDS at the C1/C2 region. Calcification is seen on the transverse, apical and alar ligament, but it should be noted that the finding is not specific to the clinical entity.<sup>7</sup> In this case, neck CT should have been performed before irrigating surgical site.

We tend to misdiagnose CDS with meningitis and localized infection especially after the brain surgery. It easily makes us perform an unnecessary lumbar puncture, but it may be avoided by excluding meningitis from the patient's symptoms. As for our case, we considered that meningitis was not likely, because the patient did not complain of lassitude and the neck rigidity appeared in the horizontal direction.

In addition, procalcitonin level was negative in this case. Serum procalcitonin level may be an useful tool to help the diagnosis of CDS, because it can be used to rule out infectious diseases.<sup>7</sup> Neck pain, increased WBC and CRP, and fever in patients with CDS are usually relieved within 1 to 3 weeks by the use of NSAIDs or a combination of NSAIDs and prednisolone.<sup>1</sup> In addition, CT sometimes shows disappearance of periodontoid calcifications. The prognosis of CDS is good when improvement of symptoms is drastic.<sup>3,6</sup> There are few previous reports on poststroke CDS, but CDS after neurosurgical procedures like our case has not been previously reported.<sup>5</sup> In general, CDS is often caused after invasive procedures. Although it has not been previously reported, such invasive procedures may include neurosurgical procedures and operative body position.

While these factors may have had an influence in our case, they may not be the only cause of CDS since the symptom improved at an early stage after reoperation; although it is merely an assumption, we believe that stimulus from foreign materials such as the burr hole cap and sutures may have played a role. CDS is caused by the deposition of CPPD crystals or hydroxyapatite to the damaged arthroge-nous cartilage. When they fall into the arthroge-nous space, the protein complex inflammasome activates caspase I, which then produces interleukin-1 and mobilizes leuco-cytes. This inflammatory process is caused by the innate immune system.<sup>11)</sup> In our case, the inflammatory process may have been aggravated by the stimulus from foreign materials such as the burr hole cap and sutures because the symptom immediately improved after irrigation and removal of the cap.

### Conclusion

CDS may be an unfamiliar disease for the neurosurgeon. Its symptoms of stiff neck and elevated inflammatory markers after operation often lead to misdiagnosis of meningitis. We should make the correct diagnosis by radiographic imaging and avoid unnecessary lumbar puncture. In addition, there are no reports of CDS that relieved immediately after reoperation. We believe that we have experienced a rare case and clinical course of CDS.

### Conflicts of Interest Disclosure

None of the authors have any disclosure to report.

### Patient Consent

The patient has consented to the submission of the case report for submission to the journal.

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