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Crowned Dens Syndrome

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Background: Patients with crowned dens syndrome typically present with severe neck pain and have calcium deposits around the odontoid process of the axis on radiographs. To our knowledge, the cases of only thirty-five patients have been reported in the English-language literature and the clinical features remain unclear. The purposes of this study were to examine the clinical features of crowned dens syndrome, determine treatment outcomes, and propose diagnostic criteria.

Methods: Forty patients with severe neck pain had calcium deposition around the odontoid process on computed tomography scans, and they were thus diagnosed as having crowned dens syndrome. Data were collected in relation to these patients, including the date of onset of neck pain, the presence of inflammatory indicators (increased body temperature, C-reactive protein levels, and white blood-cell count), and treatment outcomes.

Results: The male-to-female ratio was 0.6, and two-thirds of the patients were more than seventy years of age. All patients had markedly restricted neck motion, particularly in rotation, and all had one or more positive inflammatory indicators. Calcium deposition was detected in all areas around the odontoid process, but chiefly behind the process. Pain was typically relieved by nonsteroidal anti-inflammatory drugs, prednisolone, or both. A combination of both appeared to be the most effective.

Conclusions: We believe that crowned dens syndrome is more common than previously recognized, especially in elderly patients. It is diagnosed on the basis of acute and severe neck pain; marked restriction of neck motion, particularly in rotation; the presence of inflammatory indicators, such as an elevated C-reactive protein level; calcium deposition around the odontoid process detected by computed tomography; no history of trauma; and the exclusion of other inflammatory diseases and tumors. Prednisolone and nonsteroidal anti-inflammatory drugs in combination are the recommended treatment for symptom relief.

Level of Evidence: Therapeutic Level IV. See Instructions to Authors for a complete description of levels of evidence.

Calcium deposition around the odontoid process of the axis can be associated with neck pain and even cervical myelopathy when the deposition builds up and compresses the spinal cord[1-7]. Ossifications around the dens may also be asymptomatic[2,8]. In 1985, Bouvet et al. proposed crowned dens syndrome as a clinical and radiographic entity characterized by acute neck pain and evidence of calcium deposits around the odontoid process on radiographs[1]. The calcium deposits are thought to be composed of calcium pyrophosphate dihydrate or hydroxyapatite crystals, although we found only three cases in which the deposits had been examined histologically[1-5].

General orthopaedists and spine surgeons may not be aware of this entity as articular chondrocalcinosis, such as pseudogout which involves calcium pyrophosphate dihydrate deposition, may be present elsewhere[9]. To our knowledge, the cases of only thirty-five patients with crowned dens syndrome from fourteen institutions have been reported in the English-language literature[1-7,10-16]. Of these cases, nine were referred to by different names, such as calcification of the transverse ligament, calcification of the alar ligament, or calcium pyrophosphate crystal deposition at the craniovertebral junction[4,5,12,14], and the largest of these reported series consisted of only eight patients[6]. The terminology of calcium deposition

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disease around the dens is confusing, and the clinical features of crowned dens syndrome are unclear. Once we recognized the disease entity, we realized that we had treated many patients with crowned dens syndrome. Hence, we performed a retrospective review of the clinical data on our patients in order to clarify the clinical features of crowned dens syndrome, including the computed tomographic findings and treatment outcomes, and to propose diagnostic criteria.

Materials and Methods

A total of 2023 patients who were at least twenty years of age visited Senboku Kumiai General Hospital from June 2003 to November 2005, with the chief symptom of neck pain. Of these patients, 1225 were diagnosed as having cervical radiculopathy, spine injuries, destructive spondyloarthropathy, inflammatory disorders, tumors, intracranial disorders, or retropharyngeal tendinitis[3,17]. The remaining 798 patients had only neck pain, and a normal or degenerative cervical spine was noted on plain radiographs. Among these patients, sixty-two had severe pain with a score of >70 mm on a visual analog scale (ranging from 0 to 100 mm, with 0 indicating no pain and 100 indicating unbearable pain). These patients underwent computed tomographic examination, and forty were diagnosed as having crowned dens syndrome on the basis of calcium deposition around the odontoid process. These forty patients are the subjects in the current study. This study was approved by the Ethics Committee of Senboku Kumiai General Hospital, and all patients gave detailed informed consent before treatment.

The following data were collected from the medical records of the patients: gender; age; duration between the onset of neck pain and the initial visit to the hospital; score on the visual analog scale for pain; range of motion of the neck; neurological abnormalities; inflammatory indicators such as fever, white blood-cell count, and C-reactive protein level; and whether there had been a history of a pseudogout attack.

Lateral and open-mouth anteroposterior radiographs of the cervical spine were examined to detect calcium deposition around the odontoid process. Anteroposterior radiographs of the wrists, knees, and pelvis were investigated for evidence of articular chondrocalcinosis elsewhere[9]. The calcium depositions on computed tomography scans at the C1-C2 joint were characterized according to their localization and extent with respect to the odontoid process.

Treatment approaches were divided into three groups: nonsteroidal anti-inflammatory drugs alone, prednisolone alone, or both. The treatment selected for each patient and the duration from the onset to the resolution of symptoms in each treatment group were established from the medical records. Resolution of symptoms was defined as a decrease in the pain score on the visual analog scale to <10 mm with no restriction of neck motion. Relapse of symptoms was also recorded.

Statistical analysis was performed with use of a Mann-Whitney U test, with a p value of <0.05 considered significant.

TABLE I Summary of Clinical Data on the Patients in the Present Study and Patients in Previous Reports

	Present Study (N = 40)	Previous Reports[1-7,10-16] (N = 35)
Gender		
Male	15	11
Female	25	24
Average age (range) *(yr)*		
Male	69 (48-83)	73 (37-89)
Female	75 (60-75)	62 (26-89)
Onset of symptoms *(no. of patients)*		
Acute	40	27
Chronic or subacute		8
Positive inflammatory sign*	40	25
Negative sign		2
No description available		8
Other articular chondrocalcinosis	26	23
No articular chondrocalcinosis		11
No description available		1

*A positive inflammatory sign included evidence of inflammation according to the C-reactive protein level, erythrocyte sedimentation rate, or fever.

Results

A summary of the data on the forty patients is shown in Table I. The male-to-female ratio was 0.6. Twenty-six patients (65%) were more than seventy years of age and only one man was less than fifty years of age. All patients visited the hospital within one day of the onset of pain. The pain spread mostly from the suboccipital region to the posterior part of the neck on both sides, and most patients had no specific points of tenderness. The score on the visual analog scale for pain averaged 83 mm (range, 75 to 94 mm). All patients had markedly restricted neck motion, with almost no rotational motion. No neurological abnormalities were detected.

All patients had at least one inflammatory indicator: a high temperature in ten patients, an increased C-reactive protein level in all patients (average, 4.2 mg/dL; range, 0.3 to 14.1 mg/dL; normal range, ≤0.2 mg/dL), and a slightly elevated white blood-cell count in thirteen patients (average, 11,000 cells/μL [11×10^9/L]; range, 9700 to 14,700 cells/μL [9.7 to 14.7 $\times 10^9$/L]; normal range, 3500 to 9100 cells/μL [3.5 to 9.1 $\times 10^9$/L]). The levels of the inflammatory indicators did not correlate with the severity of pain. Twenty-two patients had a history of pseudogout attacks, and twenty-six patients had articular chondrocalcinosis, which involved the knee (eight patients), wrist (eight), ligamentum flavum of the cervical spine (six), and pubic symphysis (six).

In most patients, calcification around the odontoid process was not apparent on plain radiographs. Calcium

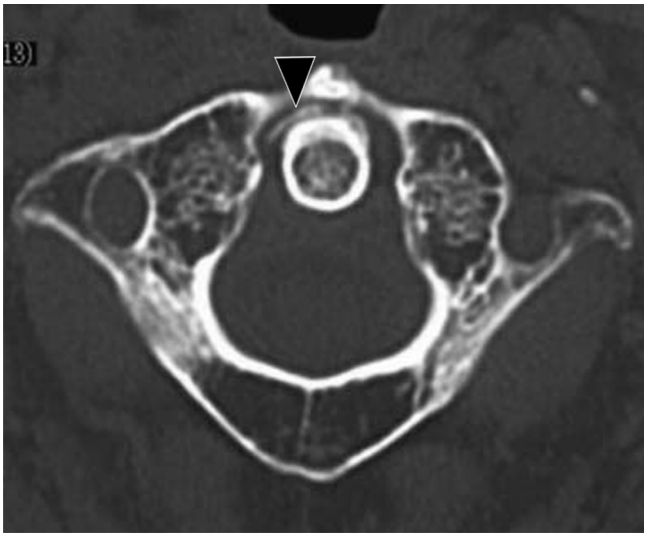

Fig. 1-A

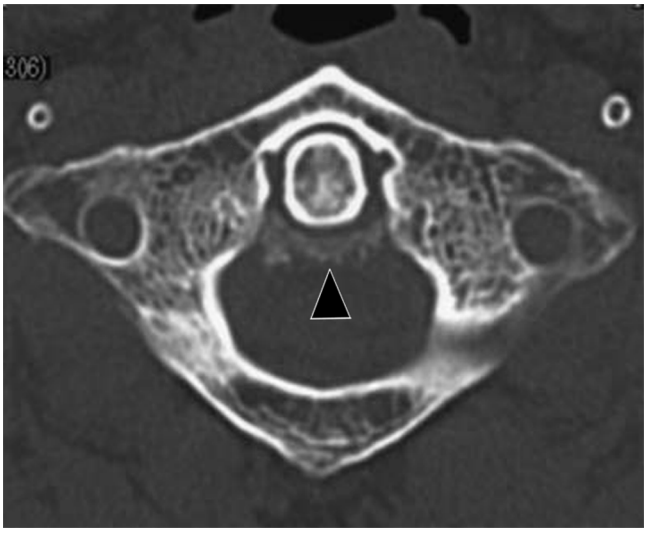

Fig. 1-B

Figs. 1-A through 1-E Patterns of calcium deposition according to localization and extent in relation to the odontoid process of the axis on computed tomography scans. **Fig. 1-A** Deposit (arrowhead) anterior to the odontoid process. **Fig. 1-B** Deposit (arrowhead) posterior to the odontoid process.

deposits were demonstrated on computed tomography scans, and most had symmetrical patterns of deposition around the odontoid process. Twenty patients had posterior deposits; eleven patients, semicircular calcification in the posterolateral aspect of the odontoid process; five patients, a circular pattern of deposition; two patients, anterior deposits; and two patients, tuberous calcification masses on both lateral sides of the odontoid process (Figs. 1-A through 1-E). Overall, calcium deposition occurred posterior to the odontoid process in thirty-six (90%) of the forty patients. In one of the four patients who had a subsequent computed tomography scan, the calcification disappeared about three months after the onset.

The patients were treated with a nonsteroidal anti-inflammatory drug, prednisolone, or both. The time required for the resolution of symptoms averaged four days (range, one to nine days). This period was significantly shorter for patients treated by nonsteroidal anti-inflammatory drugs and pred nisolone in combination compared with those treated with nonsteroidal anti-inflammatory drugs alone ($p = 0.002$) or prednisolone alone ($p = 0.006$). Six patients were initially treated with nonsteroidal anti-inflammatory drugs alone for four to six days and then with prednisolone, since they still had severe pain. After prednisolone treatment, the pain disappeared within two days. All patients were followed for at least

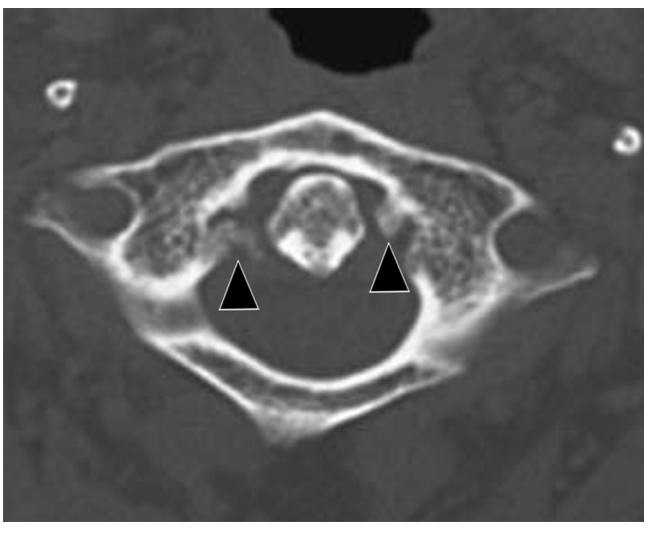

Fig. 1-C

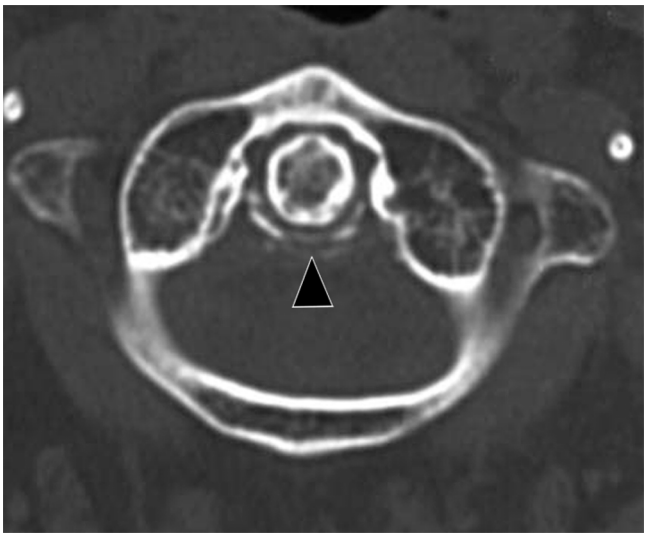

Fig. 1-D

Fig. 1-C Tuberous deposits (arrowheads) lateral to the odontoid process. **Fig. 1-D** Semicircular deposit (arrowhead) posterolateral to the odontoid process.

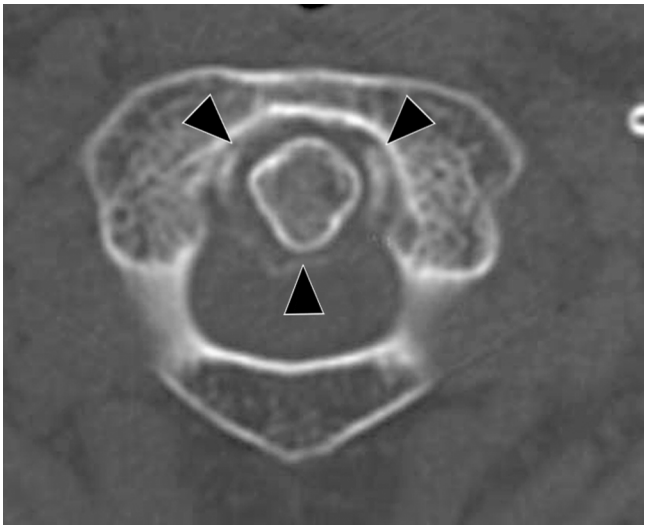

Fig. 1-E
Circular deposit (arrowheads) surrounding the odontoid process.

nine months after the initial attack (average, twenty-two months; range, nine to thirty-nine months), and nine patients (23%) had a relapse of symptoms with an average interval of nine months (range, 1.5 to eighteen months). The patients with a relapse were treated with nonsteroidal anti-inflammatory drugs and prednisolone in combination, and the symptoms completely resolved within seven days, with no further relapse for six months or more.

Discussion

Calcium deposition around the odontoid process of the axis is occasionally detected by computed tomography. Asymptomatic deposition may occur in adults affected by inflammatory rheumatic conditions such as psoriatic arthritis and ankylosing spondylitis[2]. Tumors, including chordoma, meningioma, aneurysmal bone cyst, and osteoblastoma, and acute calcific retropharyngeal tendinitis may also have calcium deposition around the dens[3]. A large deposition may compress the spinal cord, causing cervical myelopathy, and this condition has been designated as retro-odontoid massive calcium pyrophosphate crystal deposition, massive calcium pyrophosphate crystal deposition in the craniovertebral junction, foramen magnum syndrome from pseudogout, and tophaceous pseudogout[3-5,18-22].

As Malca et al. pointed out, however, crowned dens syndrome should be distinguished from calcification around the dens, which is only a radiographic entity without cervical pain[2]. Crowned dens syndrome should be defined by characteristic clinical and radiographic findings[1]. The typical clinical features of patients with crowned dens syndrome have been described as an acute onset of severe neck pain, restricted range of neck motion (particularly of rotation), and positive inflammatory indicators[1,2,6], all of which were seen in our patients. Several patients, however, have been described in reports as having subacute pain or chronic symptoms, which were atypical for crowned dens syndrome[6,7]. Neck rotation is often markedly restricted, and this is likely due to inflammation around the odontoid process[23]. There is doubt as to whether severe neck pain can actually originate from calcification around the odontoid process. Wu et al.[7] showed enhancement of the odontoid process and adjacent soft tissues by magnetic resonance imaging, and we also confirmed a high-signal intensity change on T2-weighted images around the dens in three patients (Fig. 2); these findings suggest inflammation around the odontoid process[24].

The forty patients with crowned dens syndrome in this study were among 2023 patients who visited our hospital over a period of 2.5 years with neck pain as the chief complaint. They accounted for two-thirds of those with severe neck pain who had a visual analog scale score of >70 mm on a 100-mm scale and 5% of the patients who had neck pain alone with a normal or degenerative cervical spine on plain radiographs. Therefore, crowned dens syndrome appears to be a relatively common cause of severe neck pain. The condition is most commonly seen in elderly women. In the thirty-five patients described in previous reports, the male-to-female ratio was 0.46, the mean age at the time of onset was seventy-three years in men and sixty-two years in women, and half of the patients were more than seventy years old (Table I)[1-7,10-16]. Similar trends were apparent in our series of patients. The incidence of calcium deposition disease probably increases with age, although the exact prevalence is unknown[3,8,25]. It is of note that the geographic area in which the current study was conducted has a

Fig. 2
A T2-weighted magnetic resonance image of a patient, showing a high-signal intensity change around the dens (arrowhead).

large elderly population, with people over sixty-five years old accounting for about 30% of the local population; this might explain why we encountered a particularly high frequency of patients with crowned dens syndrome.

Calcification can develop anywhere around the odontoid process, including the synovial membrane, articular capsule, transverse ligament, and transverse cruciate and alar ligaments[2,6,12,14,26], although 90% of the deposits in our study were posterior to the odontoid process.

Patients with crowned dens syndrome typically have a good prognosis, and symptoms usually subside within a few weeks[1,2]. Calcium deposits may resorb, as observed in one of our patients[14]. However, treatment with nonsteroidal anti-inflammatory drugs, colchicine, or steroids has been reported to lead to quicker resolution of the symptoms[1,6,15]. We found that nonsteroidal anti-inflammatory drugs and prednisolone in combination were most effective and thus are the recommended treatment. ■

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