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Crowned Dens Syndrome Triggered by Dental Treatment

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ABSTRACT

Crowned dens syndrome (CDS) is an important yet often overlooked cause of fever and neck pain, frequently leading to unnecessary examinations and treatments and misdiagnosis as infectious diseases or rheumatic diseases. The mechanism of an acute attack of CDS is not clarified completely, while it is considered that severe systemic stress can trigger inflammation caused by calcium pyrophosphate crystals. We describe a case of CDS triggered by localized physical stress to the neck and emphasize the importance of considering this condition in cases of fever and neck pain following dental treatment.

1 | Introduction

Crowned dens syndrome (CDS) is a subtype of acute calcium pyrophosphate crystal deposition disease (CPPD) [1]. This syndrome has two characteristic features: arthritis in the cervical spine, which responds remarkably well to NSAIDs, and crown-like calcification around the odontoid process on computed tomography (CT) scanning [2]. It has been reported that the prevalence of CPPD is approximately 4%–7% of the adult populations of Europe and the United States [3, 4]. However, this prevalence is estimated based on the frequency of radiographically detected cartilage calcification, making it difficult to assess the actual prevalence. As joint aspiration required for diagnosing CPPD is difficult to perform in the case of CDS, it is crucial for identifying CDS to suspect the condition based on typical presentation and prove the presence of crown-like calcification surrounding the odontoid process on CT imaging. Evidence indicates that CDS is associated with severe systemic stress, such as acute disease, trauma, and surgical stress, and that CDS causes fever and neck pain as the initial symptoms [1]. Some reports indicate that focal stress to the neck from trauma or surgery can also cause CDS [1]. We report a rare case of CDS triggered by dental treatment.

2 | Case Presentation

2.1 | Case History/Examination

An 80-year-old woman was referred to our outpatient clinic with fever and neck pain accompanied by a rotation disorder of 2 weeks' duration. She was undergoing treatment for periodontal disease at her local dental clinic. She had a history of chronic heart failure, paroxysmal atrial fibrillation, hypertension, and type 2 diabetes, and she had no underlying conditions or musculoskeletal abnormalities associated with neck pain. She developed fever and neck pain on the day she received treatment for teeth 6 and 7 and periodontitis. Treatment continued for approximately 1 h and required her to extend her neck and keep her mouth open.

Fever was controlled with acetaminophen, but neck pain accompanied by rotational restriction persisted. She consulted her cardiologist about the neck pain and fever during a routine checkup. Laboratory testing showed elevation of C-reactive protein (CRP) levels but no other significant abnormalities. Her cardiac conditions were well controlled, and transthoracic echocardiography for infective endocarditis showed no

Summary

- CDS is a form of cervical pseudogout that can be triggered by localized mechanical stress.
- It should be considered a differential diagnosis for fever and neck pain after dental treatment.

intracardiac vegetations. Other medical examinations could not determine the source of CRP elevation. Her cardiologist recommended that she visit her dentist to rule out odontogenic infection, but there were no pathological changes in the oral cavity that were likely to cause severe systemic inflammation. Eventually, she was referred to our outpatient clinic for further evaluation and treatment.

On admission, her temperature while taking acetaminophen was 35.7°C, blood pressure was 134/43 mmHg, heart rate was 81/min, respiratory rate was 18/min, and oxygen saturation was 98% in room air. She was alert and oriented. Visual examination showed no abnormalities in the oral cavity or neck and no neurological abnormalities or signs of meningeal irritation. However, the patient reported posterior neck pain, particularly during lateral movements, and localized tenderness in the upper cervical spine.

2.2 | Methods (Differential Diagnosis, Investigations, and Treatment)

NSAID treatment was started for the neck pain; however, the therapeutic response was partial. Laboratory testing showed elevations in CRP levels (10.94 mg/dL), erythrocyte sedimentation rates (ESR; 122 mm/h), and white blood cell (WBC) count (9190/μL) (Table 1). Subsequent transthoracic echocardiography showed no evidence of vegetation, and blood culture results were negative, suggesting a low likelihood of infective endocarditis. Moreover, no significant abnormalities were observed in the immune and endocrine systems (Table 1). Furthermore, a rheumatologist evaluation indicated a low likelihood of rheumatic diseases such as giant cell arteritis, rheumatoid arthritis, and polymyalgia rheumatica. Noteworthy, a cervical X-ray showed no abnormalities; however, a plane cervical CT scan revealed calcification surrounding the odontoid process, which was consistent with CDS (Figures 1 and 2). The diagnosis of CDS requires the exclusion of various differential diagnoses. Gastrointestinal endoscopy and gallium scintigraphy were performed, but the results did not explain the elevated inflammatory response or her symptoms. Ultimately, accumulating evidence from those examinations, we resulted in the final diagnosis of CDS.

2.3 | Outcome and Follow-Up

Although colchicine treatment was initiated in addition to NSAIDs, it was not effective enough We started 20 mg per day of oral prednisolone, which resulted in the improvement of her symptoms with a decrease in inflammatory markers. Then, prednisolone could be tapered according to her symptoms and terminated after 1 month.

3 | Discussion

An elderly woman presented with fever and neck pain of 2 weeks' duration after dental treatment, and CDS was ultimately diagnosed. There are few reports of CDS induced by dental

TABLE 1 | Laboratory test results at admission.

Variable		Reference range
White cell count ($10^3/\mu L$)	9.19	3.30–8.60
Neutrophils (%)	78.8	40.0–70.0
Monocytes (%)	8.8	2.0–10.0
Eosinophils (%)	2.2	0.0–8.5
Basophils (%)	0.5	0.0–2.5
Lymphocytes (%)	9.7	16.5–49.5
Hemoglobin (g/dL)	10.3	11.6–14.8
Hematocrit (%)	32.5	35.1–44.4
Platelet count ($10^3/\mu L$)	257	158–348
C-reactive protein (mg/dL)	10.94	0.00–0.15
Erythrocyte sedimentation rate (mm/h)	122	3–15
Rheumatoid factor (IU/mL)	20.5	0.0–15.0
Anticyclic citrullinated peptide antibody (U/mL)	0.8	0.0–4.5
Matrix metalloproteinase-3 (ng/mL)	110.2	17.3–59.7
Antinuclear antibody	< 40	< 40
Proteinase 3-antineutrophil cytoplasmic antibody (IU/mL)	< 0.60	< 2.00
Myeloperoxidase-antineutrophil cytoplasmic antibody (IU/ML)	< 0.51	< 3.50
Sodium (mmoL/L)	136	138–145
Potassium (mmoL/L)	4.0	3.6–4.8
Chloride (mmoL/L)	103	101–108
Calcium (mg/dL)	8.7	8.8–10.1
Phosphate (mg/dL)	3.5	2.7–4.6
Aspartate aminotransferase (U/L)	19	13–30
Alanine aminotransferase (U/L)	13	7–23
Lactate dehydrogenase (U/L)	170	124–222
Alkaline phosphatase (U/L)	92	38–113
Uric acid (mg/dL)	5.7	2.6–5.5
Albumin (g/dL)	3.1	4.1–5.1
Creatinine (mg/dL)	0.69	0.46–0.79
Creatine kinase (U/L)	61	41–153
Hemoglobin A1c (%)	7.6	4.9–6.0

treatment, and fever after odontotherapy is a typical presentation of odontogenic infective endocarditis. Thus, the diagnosis of CDS was delayed. The present case highlights both the potential for dental treatment to trigger CDS and the importance of making a diagnosis on the basis of the patient's medical history and physical findings, without being misled by the unusual course.

CDS is a subtype of CPPD in the cervical spine [1] and is characterized by calcification deposits around the odontoid process [2] that are detectable on a cervical plane CT scan but not by radiography. Patients with CDS frequently present with fever and neck pain and often exhibit localized tenderness over the atlantoaxial joint and posterior neck pain that worsens with rotation [5]. An acute attack of CPPD, including CDS, can occur in people with a history of acute illness, joint trauma, or joint surgery [1]. In patients with such a history, a cervical CT scan can be helpful for detecting the cause of fever and neck pain. Although the pathophysiology of CDS is not completely understood, it is believed that calcium pyrophosphate crystals induce an innate immune response, leading to increased production and secretion of IL-1β [6]. This, in turn, activates the production of other inflammatory cytokines, triggering the recruitment of neutrophils from the blood into the joint cavity [6]. Thus, it is likely that systemic physical stress elicits a systemic inflammatory response that causes crystal-induced inflammation, leading to CDS.

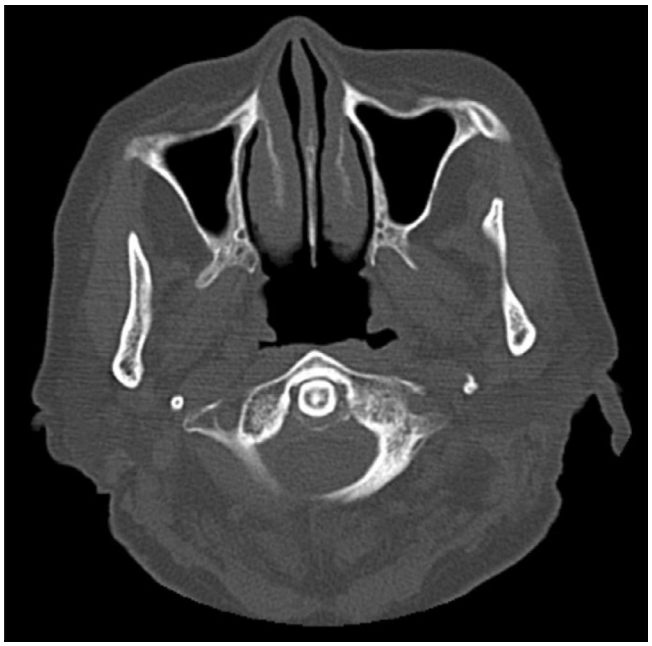

FIGURE 1 | Plain CT scan showing calcification surrounding the odontoid process. An axial CT scan of the cervical spine shows calcification on the posterior aspect of the odontoid process.

Our patient had no history of systemic stress that was sufficiently severe to cause CDS, which suggests that CDS was triggered by excessive neck extension and prolonged mouth opening during dental treatment. We were unable to determine when the periodontoid calcification developed, but certain postures during odontotherapy may place sufficient stress on the neck to enhance crystal-induced inflammation and trigger CDS. A previous study reported elevation of blood inflammatory markers, such as IL-1β and TNF-α, in patients with chronic nontraumatic neck pain and whiplash-associated disorder [7], which suggests that multiple dental treatments could potentially damage cervical tissues, thereby resulting in an inflammatory response. Although the impact of calcium pyrophosphate crystal on inflammation in pseudogout remains unclear, it is possible that mechanical stress, such as cervical hyperextension and oral irritation during dental treatments, could trigger CDS attacks at the sites of crystal deposition. Therefore, the present case suggests that many cases of dental treatment–induced CDS might have been overlooked.

The present case indicates that a recent history of dental treatment can complicate the diagnosis of CDS. Although CDS is regarded as an uncommon disease worldwide [8–10], it is regarded as a common disease in elderly Japanese, perhaps because CT scans are ordered more frequently in Japan than in other countries [11]. However, diagnosing CDS is challenging, even in Japan, as there are many diseases with similar presentations. CDS is often misdiagnosed as polymyalgia rheumatica, giant cell arteritis, meningitis, and cervical spondylitis [12, 13].

FIGURE 2 | Comparison of a cervical sagittal radiograph and CT scan. Calcification around the odontoid process is not visible on a radiograph.

Moreover, the presence of crown-like calcification, which is not sufficient for a diagnosis of CDS, increases with age and is often asymptomatic in older adults [14]. Thus, a relationship between symptoms and calcification in the cervical spine should not be presumed; instead, other causes must be carefully excluded. As seen in our patient, it was essential not to be misled by the course of fever after dental treatment. Indeed, an early comprehensive evaluation might have alerted us to the possibility of CDS and prompted CT scanning of the cervical region. However, infective endocarditis and the spread of odontoid infection to the neck should have been excluded [15], and there is no doubt that the necessity to rule out these conditions has made the situation more complicated.

NSAIDs [1] and colchicine [13] are effective treatment options for CDS, and systemic glucocorticoids should be considered for nonresponders to those drugs [11]. Because steroids can exacerbate infection and mask underlying diseases, mimics of CDS must be thoroughly excluded before their use.

Our findings indicate that because of the risk of CDS, neck symptoms and physical findings should be thoroughly evaluated in patients with persistent fever after dental treatment. CDS is a type of CPPD and basically follows the diagnostic criteria for CPPD. However, due to the technical difficulty of atlantoaxial joint aspiration, confirming the diagnosis of CDS is challenging. Thus, it is essential for the diagnosis of CDS to suspect the possibility based on the presentation, to detect the presence of crown-like calcification on CT scanning, and to rule out the mimickers. Although the diagnosis of CDS requires complex testing, symptoms dramatically improve with simple treatment. Therefore, CDS should be included in the differential diagnosis when warranted, to facilitate early diagnosis and treatment.

4 | Conclusion

We experienced a case of CDS in an elderly woman who developed fever and neck pain after dental treatment. Diagnosis of CDS is not easy in older adults because crown-like calcifications can be asymptomatic and the differential diagnosis is not straightforward. Moreover, dental treatment–induced CDS may be misdiagnosed because it is not widely known and because other conditions, such as infective endocarditis, may mimic CDS. However, we believe that physicians and dentists should be made aware that dental treatment can trigger CDS, and such knowledge will contribute to early resolution of unexplained fever and neck pain after dental treatment and prevent unnecessary examinations and treatments.

Author Contributions

Satoru Morita: conceptualization, data curation, investigation, methodology, visualization, writing – original draft. **Yuki Otsuka:** conceptualization, investigation, methodology, writing – review and editing. **Yohei Masuda:** conceptualization, investigation, methodology, project administration, supervision, writing – review and editing. **Yoshiaki Soejima:** conceptualization, investigation, methodology, writing – review and editing. **Fumio Otsuka:** conceptualization, methodology, supervision, writing – review and editing.

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Consent

Written informed consent was obtained from the patient to publish this case report in accordance with the journal's patient consent policy.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

The data supporting this article's findings are available from the corresponding author upon reasonable request.

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