Case Report

International Journal of Integrated Health Sciences (IIJHS)
ISSN Print: 2302-1381; ISSN Online: 2338-4506

Crowned Dens Syndrome: A Rare Cause of Sudden Onset Neck Pain

Nauman Ismat Butt,[1] Muhammad Bilal Rasheed,[1] Fahmina Ashfaq,[2] Aniqa Anser[1]

[1]Department of Medicine & Allied, Azra Naheed Medical College, Superior University Lahore, Pakistan
[2]Department of Medicine, Jinnah Hospital, Allama Iqbal Medical College Lahore, Pakistan

Article History

Received: November 05, 2022
Accepted: September 05, 2023
Published: October 30, 2023

DOI: 10.15850/ijihs.v11n2.3083
IJIHS. 2023;11(2):120–124

Correspondence:
Dr. Nauman Ismat Butt,
Department of Medicine & AlliedAzra Naheed Medical College, Superior University Lahore, Pakistan.
Email: nauman_ib@yahoo.com

Abstract

Objective: To report a case of Crowned Dens Syndrome (CDS), which is a rare disorder caused by crystal deposition by calcium pyrophosphate dihydrate in the peridontoid soft tissues surrounding the C1 and C2 vertebrae that presents in elderly with sudden onset neck pain, neck stiffness, fever, and elevated inflammatory markers, with periodontal calcification in a halo or crown configuration on radiography considered diagnostic.

Methods: A 64-year-old diabetic and hypertensive female patient presented with a 6-day history of sudden onset posterior neck pain and stiffness. Movements of the cervical spine were equally limited in all directions, causing marked aggravation of pain. There was no focal neurologic loss. Her inflammatory markers were markedly raised.

Results: Based on radiography, she was diagnosed with Crowned Dens syndrome and started on oral prednisolone, paracetamol, and tizanidine along with topical diclofenac. Oral NSAIDs were contraindicated due to her renal insufficiency.

Conclusion: Crowned Dens Syndrome (CDS) is a rare cause of neck pain. Clinicians should consider this syndrome in their differential diagnosis. Timely diagnosis and treatment of CDS will lead to avoidance of unnecessary investigations and medications in such patients, along with a reduction in the length of stay.

Keywords: Calcium pyrophosphate deposition, crowned dens syndrome, neck pain, neck stiffness

Introduction

Crowned Dens Syndrome (CDS) is a rare disorder, and was first reported in 1985 as a disorder occurring mainly in older individuals with a male-to-female ratio of 3:5. An under-recognized cause of acute neck pain and fever, CDS is a distinctive clinical syndrome linked to Calcium Pyrophosphate Deposition Disease (CPPD). CPPD, occurring mostly in articular cartilage and ligaments, is asymptomatic in 50% patients but can manifest as acute joint inflammation similar to gout, and is therefore also referred as pseudogout. CDS is caused by crystal deposition by calcium pyrophosphate dehydrate in the peridontoid soft tissues surrounding the C1 and C2 vertebrae. This generally presents in elderly with sudden onset neck pain, neck stiffness, fever and elevated inflammatory markers.[1,2] Delirium has also been reported in these elderly patients. CDS is a rare entity and little is known about its epidemiology. It is very likely that many cases of CDS are missed. Its diagnosis relies heavily on appropriate imaging. Diagnosis of CDS is based on presence of periodontal calcification above and lateral to dens of C2 vertebra in a halo or crown configuration on radiography.[3] Aspiration of local fluid collections that demonstrate calcium pyrophosphate crystals in crowned dens syndrome is the most definite means for diagnosis; however, aspirations are rarely performed clinically.[4] Advances in imaging, for example dual-energy computed

tomography that may chemically identify crystal deposition, would certainly help in CDS diagnosis.[5] Nevertheless, until such type of testing becomes easily available, it is essential that CDS be included as a differential diagnosis in clinical evaluation of patients with acute neck pain and raised inflammatory markers especially in elder patients.

In general, CDS needs medical management and anti-inflammatory medications alone is still the mainstay of treatment for this condition.[6] CDS has a good prognosis and the patients generally become asymptomatic within 4–6 weeks after initiation of therapy.[7] Majority of the patients recover without any clinical sequelae. Modification or resorption of the calcific deposits is commonly seen within 4-6 weeks or even earlier. Non-steroidal anti-inflammatory drugs (NSAIDs) are the gold standard of anti-inflammatory treatment in CDS and are given as first line therapy. NSAIDs provide rapid relief of pain generally within a few days after initiation and also reduce the blood inflammatory markers. Oral colchicine may also be prescribed in addition to NSAIDs. In severe disease or disease not responding to NSAIDs, corticosteroids such as prednisolone may be used. Furthermore, in refractory or severe cases where the symptoms persist despite medical therapy, surgical decompression and stabilization may be required, especially when there is evidence of spinal cord compression, myelopathy or cervical stenosis.[8] Herein, the report case of a patient presenting with sudden onset posterior neck pain and stiffness who was diagnosed with CDS after appropriate radiography. Although a rare cause of neck pain, it is important to diagnose CDS timely and clinicians should consider it in their differential diagnosis to avoid unnecessary investigations and medications in such patients.

Case

A 64-years old female patient was presented with 6-day history of sudden onset posterior neck pain and stiffness of moderate to severe intensity causing difficulty in neck movements. There was no history of trauma. She was a known diabetic and hypertensive for last 15 years, taking oral medicines with good compliance and good control. On examination, the patient was vitally stable with a temperature of 100°F (37.7°C). She was apprehensive due to pain but was co-operative and reported tenderness on palpation of back of neck. There were no deformity, mass or skin lesions on examination of neck and back. Movements of the cervical spine were equally limited in all directions, with no radiation of pain. However, neck movements caused marked aggravation of pain. Signs of meningeal irritation (Kerning's and Brudzinski's) were negative. There was no sensory loss; normal muscle tone, reflexes and power in both upper and lower limbs. Rest of the examination was unremarkable.

The result of the X rays of the cervical spine demonstrated calcification of para-

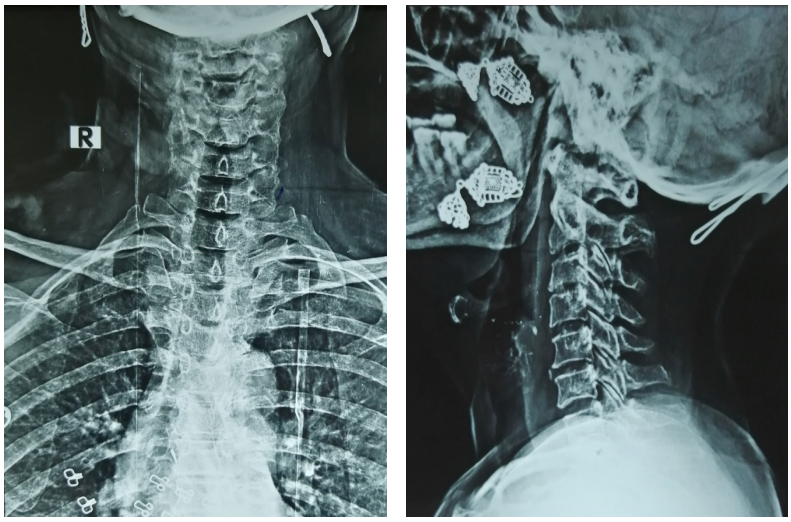

Fig. 1 X-ray Cervical Spine (AP and Lateral Views)

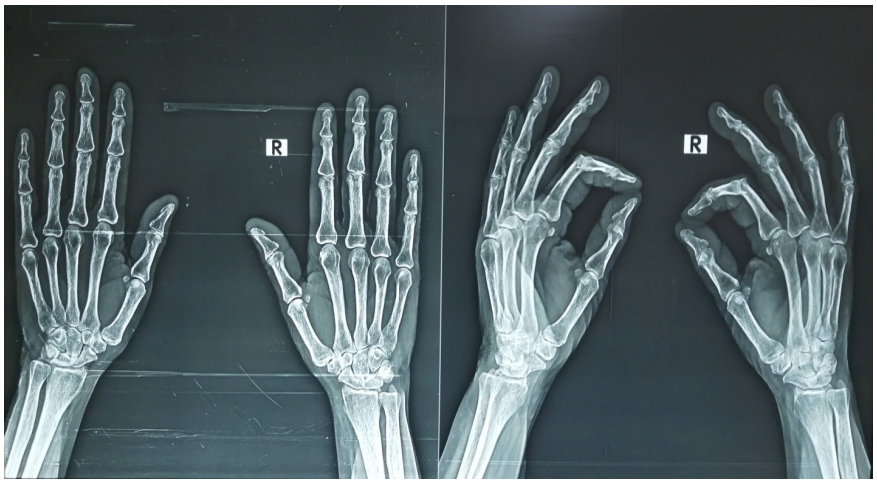

Fig. 2 X-ray Hands (AP and Oblique views)

vertebral soft tissue at C1 and C2 vertebrae, mild osteophyte formation and straightening of cervical spine as shown in Fig. 1. On further investigation, her ESR (98 mm/hour) and CRP (86 mg/dl) were markedly raised. She had moderate renal insufficiency with a creatinine clearance of 45 ml/min. Her CBC, liver profile and urinalysis were normal. HbA1c was 6.6%. Blood and urine cultures were negative. X rays of the hands demonstrated minimal joint space narrowing at 3rd and 4th PIP joints bilaterally, subtle osteophyte formation at right 3rd PIP joint and generalized reduction in bone mineral density as shown in Fig. 2.

The X rays of the knees demonstrated moderate joint space narrowing of medial compartment more of left knee and early osteophyte formation as shown in Fig. 3. The differential diagnosis included cervical spondylitis, vertebral fracture, metastatic vertebral tumor, meningitis, polymyalgia rheumatic and giant cell arteritis which were ruled out after clinical examination and investigations. Based on clinical and radiographic findings, she was diagnosed as Crowned Dens syndrome and started on oral prednisolone 10mg per day, colchicine 1mg per day, paracetamol 2gms per day and tizanidine 8mg per day along with topical diclofenac gel application. Oral NSAIDs were contraindicated due to her renal insufficiency. The patient improved and was discharged 7 days after admission with advice regarding regular follow up and tapering of oral corticosteroids.

Discussion

Crown Dens Syndrome or CDS is a rare cause

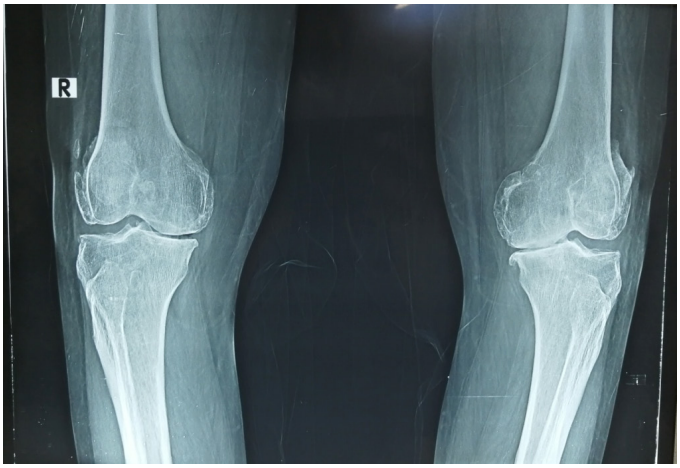

Fig. 3 X-ray Knees (AP view)

of neck pain, stiffness, fever and elevated inflammatory markers. The differential diagnosis of neck pain, stiffness and fever is quite broad and includes cervical spondylitis, vertebral fracture, metastatic vertebral tumor, meningitis, polymyalgia rheumatic and giant cell arteritis.[9] There is often a delay in diagnosis of CDS or it may be misdiagnosed entirely leading to extensive investigations, inappropriate medications and prolonged hospital stay.[9,10] CDS is not uncommon but is often misdiagnosed or ignored due to a lack of knowledge about the disease. Statistics suggest that CDS is a major cause of acute neck pain causing limited mobility accounting for up to 2% cases of acute neck pain.[11] However with adequate clinical finding and appropriate investigations the differential diagnosis may be ruled out and a diagnosis of CDS can be made timely. The current diagnostic criteria for CDS include a history of acute neck pain and limited cervical activity in addition to raised inflammatory markers such as ESR, CRP and white blood cells. Radiographic finding of CDS is presence of periodontal calcification above and lateral to dens of C2 vertebra in a halo or crown conFig.uration.[3] The limitation of X-rays is that anatomical structures surrounding odontoid process are not distinguished separately. CT scan is considered investigation of choice to identify calcification of the periodontal ligaments especially the transverse ligament of atlas cruciform ligament, apical ligament and alar ligament.[12] MRI scan does not help to diagnose calcification but is superior to see inflammatory response and spinal cord compression.[13] The definitive diagnosis relies on histological demonstration of calcium pyrophosphate crystals on biopsy.[14] However the current patient was needle phobic and refused aspiration biopsy. The incidence of CDS is associated significantly with sex and age being more prevalent in elderly females.[11,15] Oka *et al*.[16] reported that patients with CDS had a mean age 71.4 years with 60% female preponderance. Trauma, genetic factors and the co-presence of rheumatologic diseases may affect development of CDS.[11] However, there has been no proven relationship of CDS with diabetes or hypertension. Calcium Pyrophosphate crystal deposition in peripheral joints can lead to chondrocalcinosis and early osteoarthritis of these joints.[17] Performing radiographs of peripherals joints (knee, hands, and wrist) in absence of specific symptoms may demonstrate involvement of joints other than atlantoaxial joint in CDS patients. In the current patient, there was no radiographic evidence of chondrocalcinosis in joints of hands, wrists and knees. This is similar to the case reported by Lee *et al*.[18] CDS has a good prognosis and the patients generally improve with medical management encompassing anti-inflammatory drugs (NSAIDs, corticosteroids, colchicine).[19,20,] The current patient had renal insufficiency due to which NSAIDs were contraindicated. She was started on oral prednisolone 10mg per day and colchicine 1mg per day after which she reported improvement in her symptoms. Follow up was planned with an aim to taper and stop colchicine in 2 weeks and corticosteroids in 4 weeks.

In conclusion, Crowned Dens Syndrome (CDS) is a rare cause of neck pain and should be considered by clinicians in their differential diagnosis. Timely diagnosis and treatment of CDS will lead to avoidance of unnecessary investigations and medications in such patients along with a reduction in length of stay.

References

1. Deroux A. Une cervicalgie fébrile [A febrile neck pain]. Rev Med Interne. 2023;44(7):381–2. doi:10.1016/j.revmed.2023.03.003
2. Sakai D, Ono R, Ichibayashi R. Physical findings of crowned dens syndrome. Clin Case Rep. 2023;11(1):e6852. doi:10.1002/ccr3.6852
3. Awisat A, Rosner I, Rimar D, Rozenbaum M, Boulman N, Kaly L, et al. Crowned dens syndrome, yet another rheumatic disease imposter. Clin Rheumatol. 2020;39(2):571–4. doi:10.1007/s10067-019-04822-9
4. Heck A, Nolan N, Rojas-Moreno C. Crowned dens syndrome: calcium pyrophosphate deposition disease masquerading as osteomyelitis. J Rheumatol. 2018;45(10):1422–3. doi:10.3899/jrheum.170921
5. Zimmer V. Crowned dens syndrome. Clin Case Rep. 2020;8(10):2088–9. doi:10.1002/ccr3.3058
6. Bamgboje AO, Mohandas N. Crowned Dens Syndrome Masquerading as Meningitis. Cureus. 2021;13(1):e12678. doi:10.7759/cureus.12678
7. Yamada T, Saitoh T, Hozumi H, Ishikawa Y, Noguchi H, Shimada Y. Crowned dens syndrome. Acute Med Surg. 2015;2(4):273.

doi:10.1002/ams2.98
8. Oka A, Okazaki K, Takeno A, Kumanomido S, Kusunoki R, Sato S, et al. Crowned dens syndrome: report of three cases and a review of the literature. J Emerg Med. 2015;49(1):e9–e13. doi:10.1016/j.jemermed.2015.02.005
9. Michel J, Bedel JB, Pinson C, Joly LM. Crowned dens syndrome: an uncommon mimic of meningitis in the emergency department. Am J Emerg Med. 2023;63:180.e1–180.e3. doi:10.1016/j.ajem.2022.09.045
10. Igami E, Fukae J, Kanazawa K, Fuse A, Nakajima A, Eguchi H, Hattori N, et al. Two rare diseases, acute calcific retropharyngeal tendinitis, and crowned dens syndrome, mimicking meningitis: A case report. Front Neurol. 2022;13:946222. doi:10.3389/fneur.2022.946222
11. Goto S, Umehara J, Aizawa T, Kokubun S. Crowned dens syndrome. J Bone Joint Surg Am. 2007;89(12):2732-6. doi:10.2106/JBJS.F.01322
12. Ross JS, Moore KR. Diagnostic imaging: spine. US: Elsevier Health Sciences; 2015.
13. Zeng L, Liao H, Xie M, Ren F, Zhang Y, Wang Q. Images in neuroscience: crowned dens syndrome. Neurol India. 2022;70(3):1329–30. doi:10.4103/0028-3886.349617
14. Rosenthal AK. Calcium pyrophosphate deposition and crowned dens syndrome. Cleve Clin J Med. 2021;88(4):206–7. doi:10.3949/ccjm.88a.21008
15. Sano M, Yamashita S, Aiba T. The prevalence of calcification around odontoid process and the incidence of crowned dens syndrome in the neurosurgical ward: A single institution's analysis. Mod Rheumatol. 2018;28(1):182–7. doi: 10.1080/14397595.2017.1316461.
16. Oka A, Okazaki K, Takeno A, Kumanomido S, Kusunoki R, Sato S, et al. Crowned dens syndrome: report of three cases and a review of the literature. J Emerg Med. 2015;49(1):e9–e13. doi: 10.1016/j.jemermed.2015.02.005.
17. Stack J, McCarthy G. Calcium pyrophosphate deposition (CPPD) disease-treatment options. Best Pract Res Clin Rheumatol. 2021;35(4):101720. doi:10.1016/j.berh.2021.101720
18. Lee GS, Kim RS, Park HK, Chang JC. Crowned dens syndrome: a case report and review of the literature. Korean J Spine. 2014;11(1):15–7. doi:10.14245/kjs.2014.11.1.15
19. Koyfman A, Yaffe D. Crowned dens syndrome. A case report. Neuroradiol J. 2014;27(4):495–7. doi:10.15274/NRJ-2014-10056
20. Abdelkefi I, Saidenberg-Kermanac'h N, Boissier MC, Semerano L, Sigaux J. Crowned dens syndrome. Joint Bone Spine. 2022;89(1):105288. doi:10.1016/j.jbspin.2021.105288