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Intractable Neck Pain in a Patient With Newly Diagnosed AML: An Underrecognized Cause of a Treatable Syndrome

To the Editor:

Crowned dens syndrome (CDS) is a variant of pseudogout involving calcium pyrophosphate dihydrate (CPPD) deposition in the atlantoaxial junction. Imaging shows characteristic crown-like calcifications surrounding the dens.[1] Patients present with severe neck pain, rigidity, and high fever. Clinical presentations can be variable, including acute- and chronic-relapsing types, and can mimic many other disease processes. Awareness of this rare, likely underrecognized, and treatable syndrome is crucial because palliative care providers care for patients at high risk for this disease. We present a patient with severe, relapsing neck pain due to CDS that was complicated by a new diagnosis of acute myeloid leukemia (AML) at presentation.

Case Description

A 75-year-old man with a history of prostate cancer, gout, Type 2 diabetes mellitus, and chronic low back pain presented to the emergency department with fever and severe neck pain. He had come to the emergency department several times in the past month for severe neck pain, which improved with nonsteroidal anti-inflammatory drugs (NSAIDs). Blood testing showed pancytopenia, and a subsequent bone marrow biopsy provided confirmation for a diagnosis of AML with 60% blasts. Palliative care providers were consulted for pain management on Day 10 of hospitalization for severe, uncontrolled, cancer-related pain.

On examination, the patient appeared uncomfortable. He had neck rigidity, right shoulder pain with pain radiating to his arm, and neck erythema and warmth. His erythrocyte sedimentation rate was high (120 mm/hour), which initially raised concern for leukemic meningitis. Hydromorphone patient-controlled analgesia was started. His right ankle swelled, and ankle arthrocentesis showed CPPD crystals. After he was given corticosteroids, his neck pain improved, and the possibility of CPPD deposition in the neck became a concern. Review of previous computed tomographic scans of the cervical spine showed calcification of the transverse ligament, consistent with CDS (Fig. 1). He began treatment with a prolonged 30-day course of prednisone with a slow taper, which resulted in complete resolution of his neck pain. He then proceeded with induction chemotherapy for AML without recurrence of CDS symptoms.

Comment

CDS is a rare and likely underrecognized disorder that can cause considerable morbidity and prolonged hospitalization. The incidence is uncertain, but CDS has been reported to occur in 1.9% of outpatients with neck pain and in 35% to 45% of patients with articular chondrocalcinosis.[1] CDS is more common in elderly patients (mean age, 71.4 years) and is equally common in male and female patients.[1] The pathophysiology of CDS involves CPPD or calcium hydroxyapatite crystal deposition in the ligaments surrounding the dens. These crystals induce inflammation, causing production of matrix metalloproteinases and prostaglandins.[2]

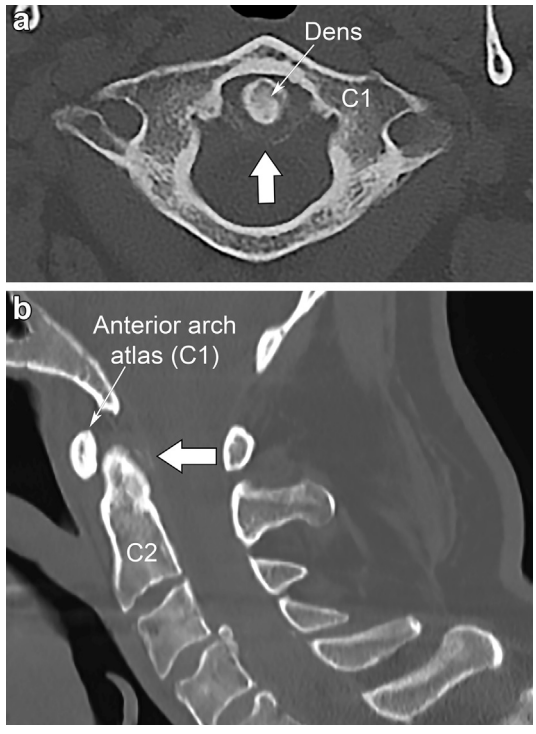

Fig. 1. Computed tomography of the cervical spine. a) Axial view shows linear calcification (arrow) in the transverse ligament of the atlas consistent with crowned dens syndrome. b) Sagittal view shows calcification surrounding the dens (arrow).

Risk factors for CDS include age greater than 60 years, acute illness, recent surgery, local trauma, and prior history of pseudogout.[2,3] In our patient, we suspected that AML was incidentally diagnosed because of severe neck pain from CDS prompting emergency department evaluation. However, AML was likely a risk factor for CDS. It is not currently known whether leukemia is involved in the pathophysiology of CDS. To our knowledge, this is the first reported case of CDS in a leukemia patient, although CDS is likely underdiagnosed given the complexities of clinical presentation.

Diagnosis of CDS is based on clinical symptoms along with characteristic findings on imaging. Clinical presentation most commonly includes the triad of neck pain (in 100% of patients), neck rigidity (98%), and fever (80.4%).[1–6] Other clinical symptoms include radiating pain (to the shoulder, temporalis muscle, or jaw), vomiting, myelopathy, and pharyngalgia.[1] Laboratory workup shows elevated inflammatory markers (in 70%–80% of patients), including increased leukocyte count, erythrocyte sedimentation rate, and C-reactive protein level.[1,2] The key physical examination finding is pain on rotation of the neck.[1] Symptom relapse, especially after a short course of corticosteroids or NSAIDs, is also suggestive of CDS. The differential diagnosis includes mechanical neck pain, polymyalgia rheumatica, giant cell arteritis, meningitis, diskitis, inflammatory arthritis, infection, leukemic meningitis, and malignant leptomeningeal disease.[1–3]

Calcifications along the transverse ligament of the atlas, and along other ligaments surrounding the odontoid process, are suggestive of CDS.[3] The preferred imaging modality is computed tomography, which provides better imaging of the calcifications compared with radiography and magnetic resonance imaging.[7] Radiographic densities are usually normal findings in patients with CDS.[8] The use of positron emission tomography can also be considered for assistance in narrowing the differential diagnosis.[2,3,9] In acute attacks, the crystals may be less prominent because of resorption, so subsequent imaging may be necessary.[3] To distinguish between patients with benign calcifications and patients with CDS, clinicians must consider symptoms and clinical presentation.

The first-line treatment is a 2- to 3-week course of NSAIDs.[1] Corticosteroids may be used for severe or recurrent disease, or if the patient has contraindications for NSAID therapy. Acute treatment with colchicine can be considered for certain patients, but the evidence is limited.[2] With treatment, symptoms usually resolve in days to weeks; however, to prevent relapse, treatment should continue for 2 to 3 weeks after the symptoms resolve. Most patients show considerable improvement within days after initiation of NSAID therapy, and symptoms gradually resolve over several weeks.[3] In patients with recurrent disease, colchicine has been used as a preventive medication.[3] Our patient was treated with prolonged course of corticosteroids for four weeks based on CDS severity and exacerbation risk. He was at high risk for future exacerbations due to the severity of CDS at clinical presentation and ongoing acute illness from AML. Corticosteroids were also preferred given our patient's advanced age, pancytopenia from AML, and other comorbidities that increased his risk for NSAID-related adverse events. Our patient started chemotherapy during CDS treatment, and his symptoms completely resolved without CDS recurrence or complications.

CDS complications include deposition of bulky, mass-like CPPD crystals or bone erosion from an intense inflammatory response, which can lead to myelopathy and fracture of the atlas.[1,10] This may cause neurologic compromise, and patients may require surgical decompression.[10] These complications may be more likely with untreated CDS; therefore, early intervention with NSAIDs or corticosteroids may decrease the chance of complications.[1]

CDS is a diagnostic challenge, and it can cause considerable morbidity and prolonged hospitalizations for patients. A team-based approach to diagnosis and treatment is key to patient care. With health care provider awareness, CDS can be recognized and treated early, and patients can avoid unnecessary testing and invasive procedures.

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Preferences for Help With a Sleep Problem Before Starting Cancer Treatment

To the Editor

Being diagnosed with cancer can be overwhelming and is often associated with significant disruptions. Before beginning cancer treatment, many patients experience poor sleep. Disrupted sleep has meaningful clinical implications beyond "just" not feeling rested. Patients who sleep poorly prior may experience worse pain, fatigue, and depression during treatment.[1] Furthermore, sleep often gets worse as treatment progresses,[2] and chronic sleep problems are associated with major negative health consequences.[3,4] Unfortunately, patients are unlikely to seek treatment for sleep problems despite robust evidence supporting the effectiveness of interventions. These interventions are often disregarded in the oncology setting by patients and providers who consider them to be an expected problem after a cancer diagnosis.[5]

Cancer patients about to begin treatment are actively engaged with the medical system due to ongoing treatment and follow-up.[6] This provides an opportunity to screen for sleep disorders and provide affected patients with evidence-based treatments. Unfortunately, only a handful of reports have explored cancer patients' information preferences for learning about sleep treatment. It is critical to investigate help-seeking among cancer patients to improve the clinical management of sleep disorders at oncology centers. We sought to address this key gap by surveying a large cohort of newly diagnosed cancer patients across a range of diagnoses to understand their treatment interest and preferences as it relates to sleep.

Methods

As part of standard clinical care, outpatients at a comprehensive cancer center were asked to complete an electronic biopsychosocial distress screen[7] before they initiated cancer treatment at a routine medical appointment between 2009 and 2016. Patients were aged ≥18 years and completed the screening questionnaire in English, Spanish, or Chinese. This study was approved by the cancer center's Institutional Review Board.

Sample and Measures

The study sample comprised 5671 participants who had been diagnosed with one of eight cancers (hematological malignancy, gastrointestinal cancer, head/neck cancer, prostate cancer, gynecologic cancer, lung cancer, urinary cancer, or prostate cancer). Most patients were female (67.5%), were white (76.9%), had completed some college (71.7%), and had an annual household income of at least $40,000/year (56.0%).

The "*You, Your Family, and the City of Hope are a Team*" distress screen was developed following a literature review and input from patients, families, nurses, and physicians.[8] The distress screen first asks respondents to report on their level of bother specific to a list of 53 commonly experienced physical, psychological, and social problems. The question is presented as "How much of a problem is this for you: ___" and patients are provided five response options, ranging from "not a problem" to "very severe problem." Second, patients are asked "How can we best work with you on this problem?" and must select one of four options: 1) "provide written information"; 2) "talk with a member of the team"; 3) "written information and talk with a member of the team"; 4) "nothing needed at this time." In this study, we examined patient responses to the question about "sleeping." In addition, demographic characteristics were obtained from the survey and the patient's electronic medical record.