Views and Reviews

The "Crowned Dens" Revisited: Imaging Findings in Calcium Crystal Deposition Diseases Around the Odontoid

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ABSTRACT

The so-called "crowned dens" is a peculiar manifestation of calcium crystal deposition diseases, either caused by calcium pyrophosphate dihydrate or caused by calcium hydroxiapatite crystals, characterized by the presence of calcific deposits around the odontoid, often showing a crown-like configuration on imaging. It has protean clinical and radiological pictures, and care should be taken to avoid misinterpretation and diagnostic errors. Although asymptomatic in many patients, this entity may present as a predominantly algic or febrile condition, and in some cases, signs of compression of the spinal cord may be the major complaint. The detection of calcifications in the periodontoid tissues is the key to the diagnosis, erosive osseous changes, and variably calcified soft-tissue masses being occasionally associated. Computed tomography is the most important imaging study to be performed in this setting.

Keywords: Crowned dens syndrome, calcium crystal deposition disease, cervical spine, odontoid process, computed tomography, magnetic resonance imaging.

Acceptance: Received October 6, 2008, and in revised form January 2, 2009. Accepted for publication January 28, 2009.

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The authors have no funding sources to declare.

J Neuroimaging 2010;20:311-323.
DOI: 10.1111/j.1552-6569.2009.00365.x

Introduction

The crystalline arthropathies correspond to a wide group of diseases that have in common the deposition of crystals inside and around the joints and the development of an inflammatory reaction in the affected tissues.[1] Even though the involvement of the peripheral joints is much more common than the axial disease, the latter has been increasingly recognized in the last decades, after the advent of axial imaging (computed tomography [CT] and magnetic resonance imaging [MRI]).[2] Most studies suggest that calcium crystal deposition diseases are the most common crystalline arthropathies affecting the vertebral column, either due to calcium pyrophosphate dihydrate (CPPD) or due to calcium hydroxyapatite (CHA). The ligamenta flava; the longitudinal, supraspinous, and interspinous ligaments; the intervertebral disks; and the sacroiliac and apophyseal joints are among the most frequent sites of crystalline deposition in the spine.[3-7]

Calcium crystal deposition disease in the periodontoid area is a peculiar variety of this entity that has gained increasing attention. Because of the crown-like appearance of the calcifications around the odontoid process on imaging, this manifestation has been called "crowned dens." Although initially regarded as a rare condition, recent works have demonstrated that it is much more common than previously thought.[2,8,9] Given that it is a potentially reversible cause of neurological deficit and may simulate other diseases as well (either from a clinical or from a radiological standpoint), it is important that radiologists are aware of its protean appearance on imaging to avoid misdiagnoses and incorrect management.

Clinical Background

The clinical picture associated with either form of periodontoid deposition of calcium crystals is very similar, reflecting a nonspecific inflammatory reaction set off by the presence of microcrystalline deposits.[10,11] Women are preferentially affected in both types of arthropathy, mostly young or middle-aged in CHA disease and middle-aged or elderly in CPPD deposition.[5,12] The term "crowned dens syndrome" has been coined to describe a quite heterogeneous constellation of signs and symptoms linked to this particular type of crystalline arthropathy, usually of acute onset. It may vary from a mild condition, with cervicalgia and neck stiffness, to more dramatic scenarios, with spiking fevers and meningeal signs. The clinical picture may be recurrent, with bouts lasting days or even weeks, usually subsiding after the use of nonsteroidal anti-inflammatory drugs.[2,11,13,14] This syndrome may simulate a wide array of spinal diseases, including meningitis, gout, rheumatoid arthritis,

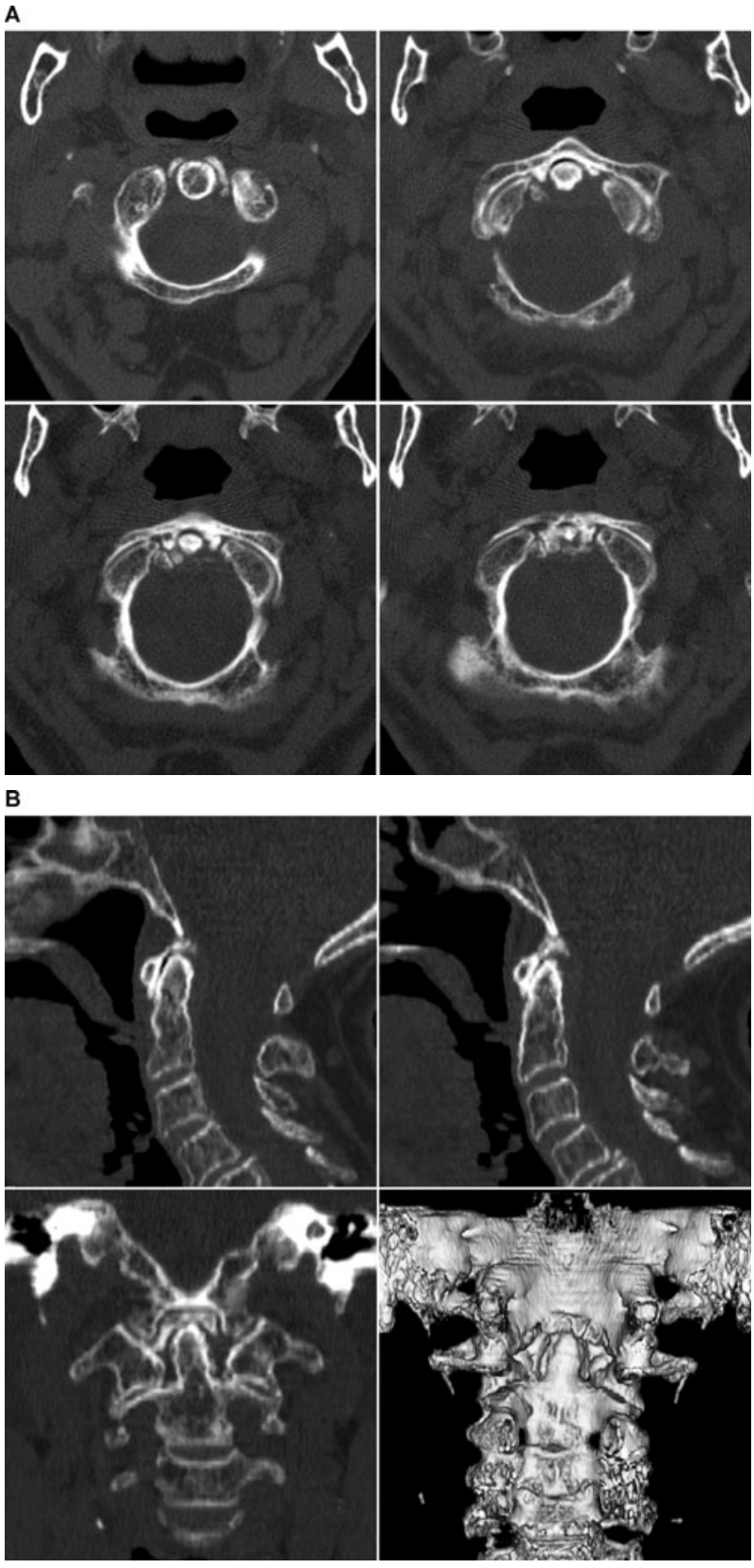


Fig 1. Axial CT scans at C1-C2 level (A) in an asymptomatic 75-year-old woman showing calcified deposits lateral and superior to the odontoid process and degenerative changes of the atlanto-axial joint characterized by gas within the joint space, subchondral sclerosis, and marginal osteophytes. Reformatted images in the sagittal (B, upper row) and coronal (B, lower left image) planes disclose even more clearly the crown-like periodontoid calcifications and the atlanto-axial degenerative changes, also shown in a volume-rendered 3-dimensional reconstruction (B, lower right image). A reformatted sagittal image with inverted window (C) reveals a noncalcified retro-odontoid soft tissue component.

Fig 1. Continued.

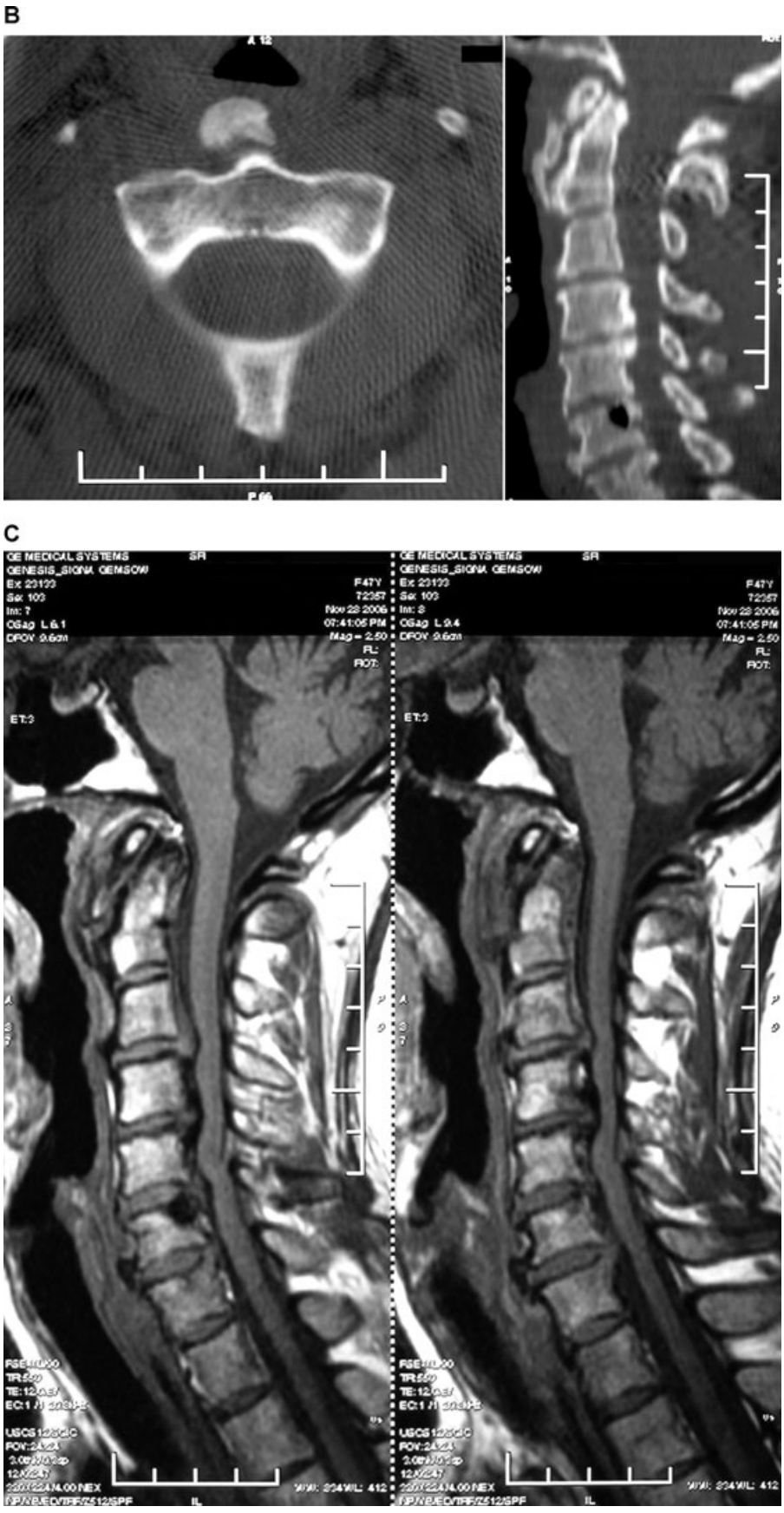
osteomyelitis, spondylosis, ankylosing spondylitis, polymyalgia rheumatica/temporal arteritis, and vertebral tumors.[3,7] In addition, these patients may exhibit signs of cervical spinal cord compression, either due to direct compression exerted by the crystalline deposit itself or due to a displaced fracture of the odontoid process.[4,9] However, the periodontoid deposition of calcium crystals is often a silent process, being incidentally discovered during examinations of a different anatomic site.[13,15] Some imaging findings may help to discriminate asymptomatic from painful calcifications, mostly at MRI (see below).

Imaging Evaluation

A wide variety of tissues may be affected by the crystalline arthropathies in the craniovertebral junction, including cartilage, synovium, bursae, tendons, and the joint capsule.[7,9] The imaging methods per se do not establish with absolute certainty the type of crystal involved in a given patient, as both CHA and CPPD may deposit around the odontoid and produce the classical "crowned dens" appearance[2,10] (Fig 1). Nevertheless, some findings may suggest a specific etiology in many cases. Massive prevertebral calcifications anterior to C2, along the longi colli muscles, dense and homogeneous, are indicative of CHA disease[6,16] (Fig 2). On the other hand, CPPD crystals tend to deposit in the transverse/cruciform and alar ligaments, being the resulting calcifications of delicate appearance, linear or stippled, and commonly (although not inevitably) associated with chondrocalcinosis of other corporal sites[1,15,17] (Fig 3). However, both patterns may be found in the same patient, and it was already proven that CPPD and CHA crystals may coexist.[16,18,19] Given that this condition is frequent in elderly individuals, degenerative disease of the spine is a commonly associated finding, mostly in the atlanto-axial joint[1,6,10] (Figs 4 and 5).

Odontoid erosions associated with calcified deposits may also be found in a subset of these patients[9] (Figs 6 and 7). As the disease advances, extensive bone destruction due to erosions and subchondral cysts may occur, eventually leading to insufficiency fractures.[9,15] Spinal cord compression is a dangerous complication, occurring more often in elderly patients, and may be due to fractures of the odontoid process, atlanto-axial instability, or mass effect exerted by massive crystalline deposits in the retro-odontoid area (in the transverse ligament of the atlas)[3,6,8,10,18] (Fig 8). In spite of the microcrystalline nature of these deposits, their composition–and, by consequence, the calcium concentration inside them–is variable and so is their appearance at imaging. As a result, frankly

Fig 2. Radiograph of the cervical spine (A) of a 47-year-old woman complaining of severe neck pain revealing advanced spondylosis and an elongated calcification in the prevertebral soft tissues anterior to C2, hardly seen because of overlapping with the styloid processes and the vertical mandibular rami. An axial CT scan (B, left) and a sagittal reformatted image (B, right) clearly show massive prevertebral calcifications along the longi colli muscles and degenerative disease of the cervical spine, with acquired stenosis of the spinal canal. At MRI—sagittal T1-WI (C) and T2-WI (D)—the calcifications are much less clear at MRI when compared with CT images, appearing as a prevertebral mass of heterogeneous signal intensity in all sequences. Although the diagnosis of crystalline arthropathy cannot be firmed based upon MRI images alone, the spinal cord compression due to the associated spondylosis is much better characterized by this method.



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calcified nodules can be found in one end of the spectrum, with masses of intermediate density/signal intensity in the opposite end, faintly calcified or not calcified at all[9] (Figs 1 and 8). Sometimes, these nodular calcifications may simulate odontoid fractures, particularly in a setting of cervical trauma[20] (Fig 9).

Although often requested as part of the workup of these patients because of its low cost and wide availability, radiographic investigation of the periodontoid crystalline arthropathy is limited in the detection of periodontoid calcifications, mostly due to the subtle appearance of the deposits and the inherent superposition of osseous structures in this region[9,10,19] (Fig 2). However, radiographs of the wrists and knees may reveal peripheral chondrocalcinosis, thus helping to establish the diagnosis of calcium crystal deposition disease.[13,21]

CT is the imaging method of choice in this setting by displaying in an unequivocal way the periodontoid calcifications, either the massive deposits due to CHA deposition (Fig 2) or the delicate ligamentous calcifications of the CPPD crystals (Fig 3).[7,13,16] The commonly associated erosions and bone cysts are also clearly shown[22] (Fig 6). Nevertheless, as stated above, the absence of calcifications does not exclude the diagnosis of crystalline arthropathy, particularly during the intercritical periods, given that the calcium deposits may be entirely reabsorbed.[2,10,12,13,17] If there is a strong suspicion, the examination should be repeated, preferably during a painful crisis. Reformatted images are useful to display the abnormal findings in multiple planes, the crown-like pattern of the calcifications being more evident in the sagittal and coronal planes (Figs 1–9). Concomitant calcifications in other sites, such as the disks, can also be demonstrated (Fig 10). Volume-rendered displays, although not indispensable to reach the diagnosis, are a sophisticated tool to aid image interpretation, and many surgeons find them more helpful in operative planning than single-plane images alone when surgery is contemplated (Figs 1, 3, 7, 11 and 12).

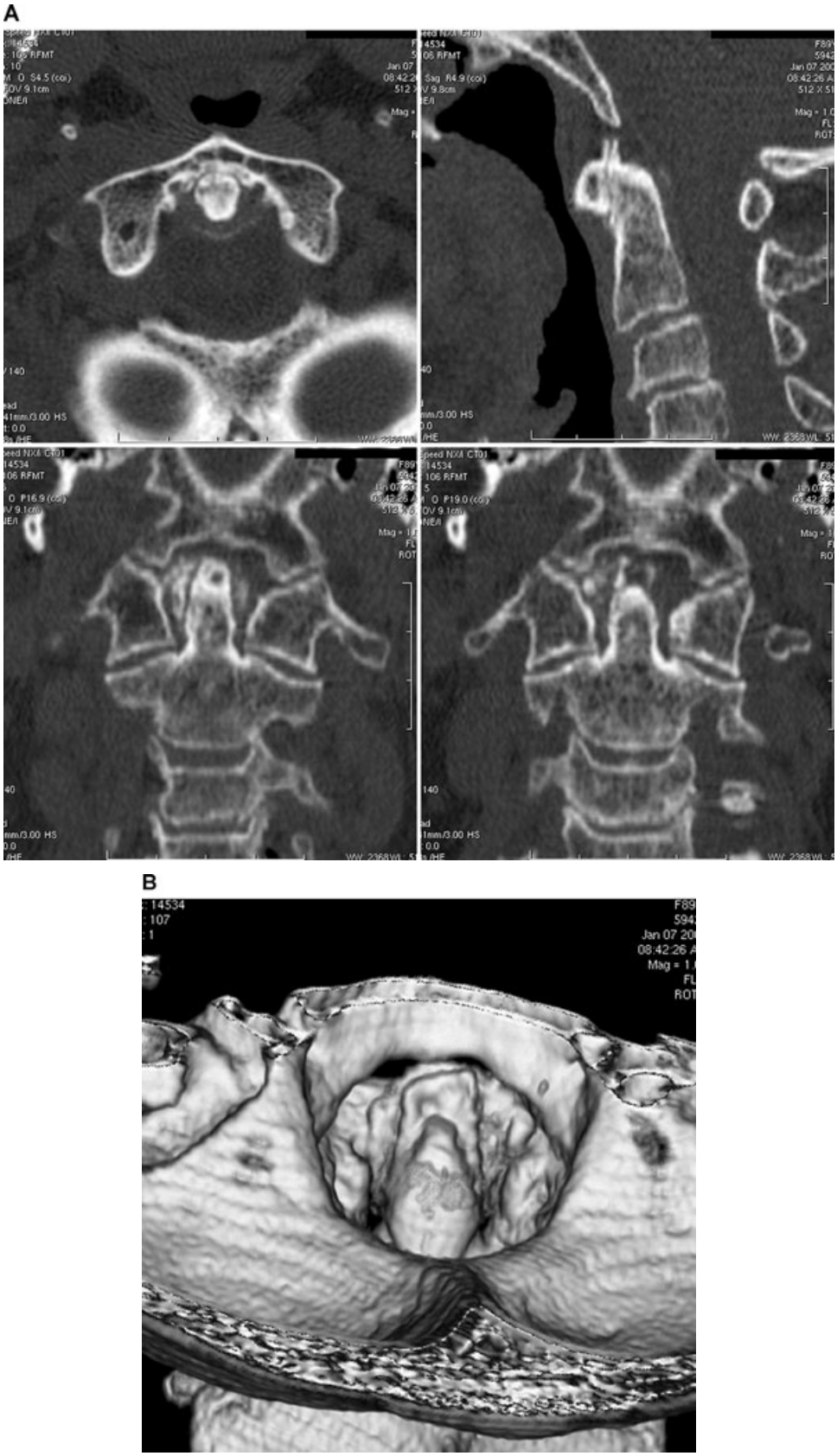


Fig 3. CT of the cranio-cervical junction of an asymptomatic 89-year-old woman referred for preoperative evaluation of dental implants. The axial and reformatted images (A) disclose faint linear calcifications along the cruciform ligament (upper row) and crown-like calcified deposits around the odontoid process (lower row), associated with severe atlanto-axial degeneration. The volume-rendered image (B) displays these findings in a more graphical way.

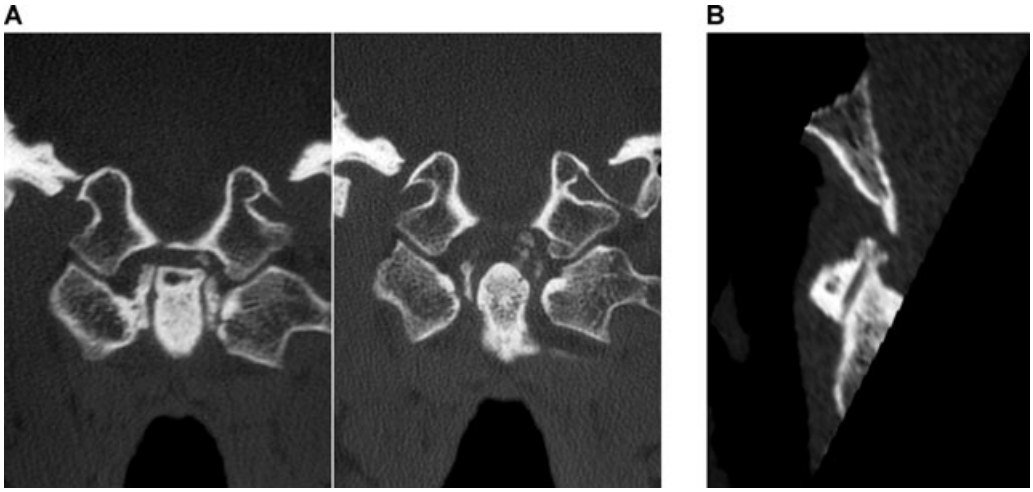


Fig 4. CT scans of the temporal bones of an asymptomatic 73-year-old woman (follow-up of right mastoidectomy for treatment of cholesteatoma). Direct coronal CT scans (A) and a reformatted sagittal image (B) show the typical "crowned" appearance of the odontoid process and severe degenerative changes of the median atlanto-axial joint.

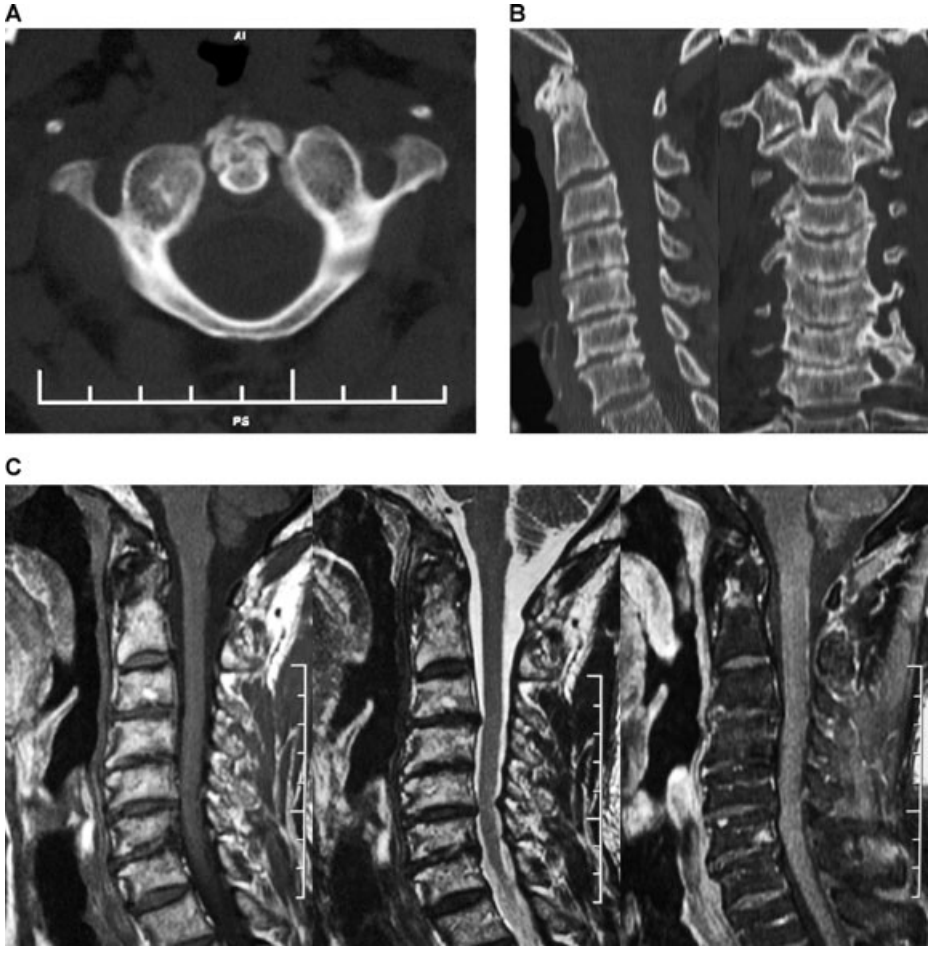
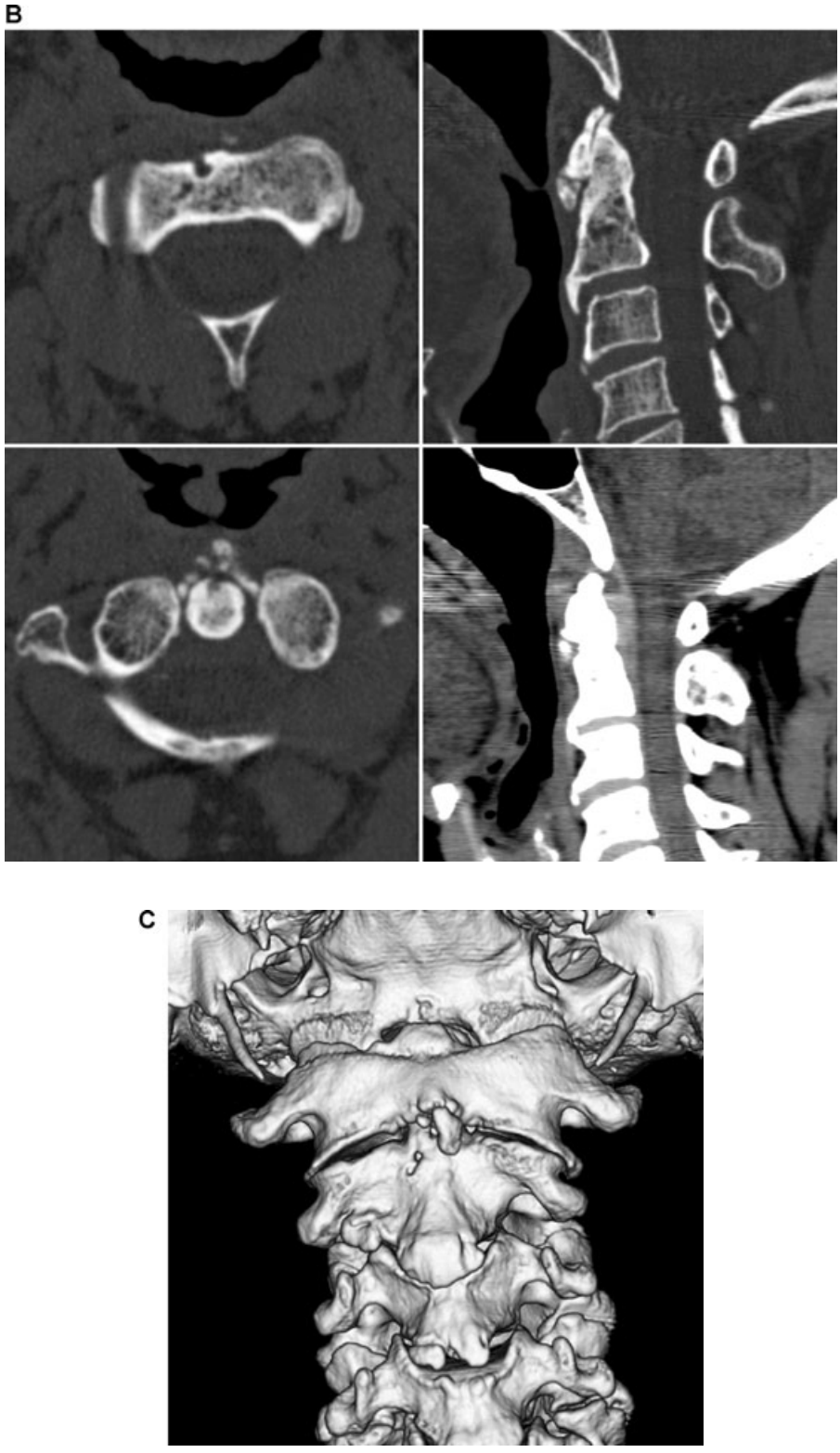


Fig 5. Axial CT section at the level of the atlanto-axial articulation (A) show degenerative articular disease, odontoid cysts and erosions, and periodontoid calcifications. Reformatted images (B), besides showing in a more elegant way the same findings of the axial slices, reveal advanced cervical spondylosis; notice the crown-like calcifications in the soft tissues around the dens and in the periodontoid ligaments. (C) MR sections of the same patient—sagittal T1-WI (left), T2-WI (center), and post-gadolinium T1-WI with fat suppression (right) —show that although the calcifications and the bone erosions are clearly more evident on CT scans, MR demonstrates post-gadolinium enhancement (seen on the right image), indicative of inflammatory activity.

Fig 6. Cervical MRI of a 76-year-old woman referred for evaluation of chronic neck pain showing material of heterogeneous signal intensity around the odontoid process, isointense in T1-WI and hypointense in T2-WI, with erosive changes of the anterior cortex and degenerative disease of the cervical spine (A). In a complementary CT, axial scans (B, left images), multiplanar reformatted images (B, right images), and a volume-rendered 3-dimensional reconstruction (C) confirm the degenerative changes and the bone erosions, revealing that the heterogeneous material described at MRI corresponded to amorphous calcifications laterally to the odontoid peg and to a nodular calcified deposit in the prevertebral region. The retro-odontoid component, although sharing similar signal features, was not calcified at CT.

MRI, in its turn, although less sensible in the detection of the calcifications, is the best imaging method to study poorly calcified or noncalcified masses because of its inherently high contrast and spatial resolutions, thus being very useful to depict subchondral erosions and bone cysts (Figs 2, 5, 6 and 8). Moreover, MRI is the method of choice to evaluate findings like bone marrow edema pattern, edema of the surrounding soft tissues and spinal cord compression—either due to mass effect exerted by the masses or due to odontoid fractures[22] (Fig 8). It is recommended to acquire T1-weighted images (T1-WI) and T2-weighted images (T2-WI, including fat-suppressed sequences) in the three spatial planes.[10] Although the finding of a mass with low signal intensity in T1-WI and T2-WI should suggest massive calcification (Fig 6), intermediate signal intensity seems to be more frequently seen in the deposits of the retro-odontoid area; heterogeneous signal intensity and even hyperintense signal intensity can be found in T2-WI[5,9,10,22] (Figs 2 and 8). In questionable cases, a complementary CT can demonstrate the calcifications beyond any doubt. Even though post-gadolinium enhancement is not a distinctive feature of the microcrystalline masses, it can be found and is indicative of inflammatory activity[9,22]; the bone marrow and the surrounding soft tissues may also exhibit some enhancement[10] (Figs 5 and 8). Although imaging itself is not able to establish whether the periodontoid calcifications are the source of pain in a given symptomatic patient, the concomitance of bone erosions, bone marrow edema pattern, soft tissue edematous changes, and/or post-gadolinium enhancement lend support to this possibility, as long as these are accepted markers of inflammation and disease activity in arthritides elsewhere.[1]

From an imaging standpoint, the differential diagnosis should include neoplasms (primary or metastatic), infectious conditions, and inflammatory arthropathies (like rheumatoid arthritis) as the most important hypotheses, principally the latter. However, the presence of calcifications in calcium crystal deposition diseases and the intense gadolinium enhancement observed in the arthritides (instead of the subtle or even absent enhancement of the crystalline arthropathies)



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allow to distinguish these conditions in most cases, especially if there is chondrocalcinosis in peripheral joints.[15,16,18,23] The diagnosis of spondylodiscitis–which may be a clinical concern–can be safely ruled out by demonstrating normal intervertebral disks and vertebral bodies.[10] Although conservative measures suffice in most cases, surgery can be contemplated in selected patients (eg, in cases of spinal cord compression).[3,8,17]

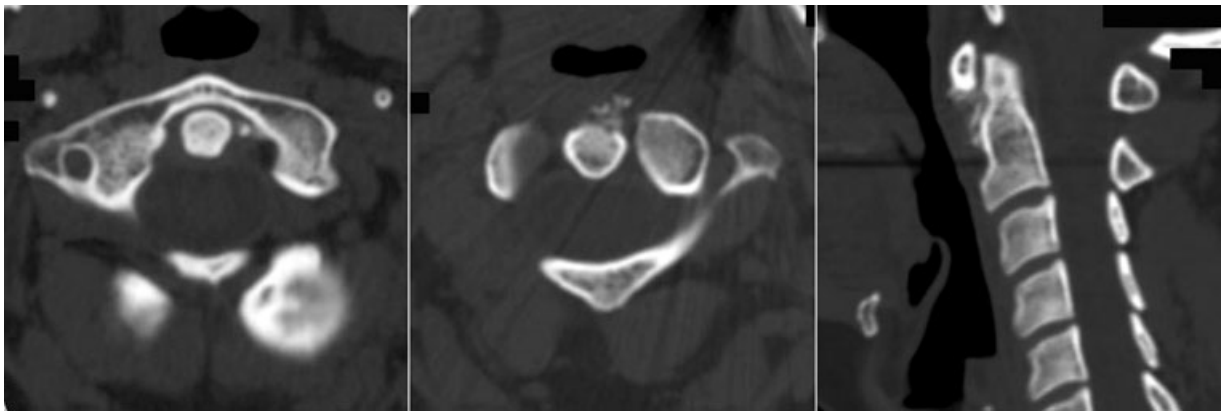

Fig 7. Axial CT slices (left and middle images) show calcified deposits anteriorly and to the left of the dens, with erosion of the anterior cortex of the odontoid peg. Sagittal reformatted image (right) confirms the erosion and the relation of the deposits with the anterior arch of C1.

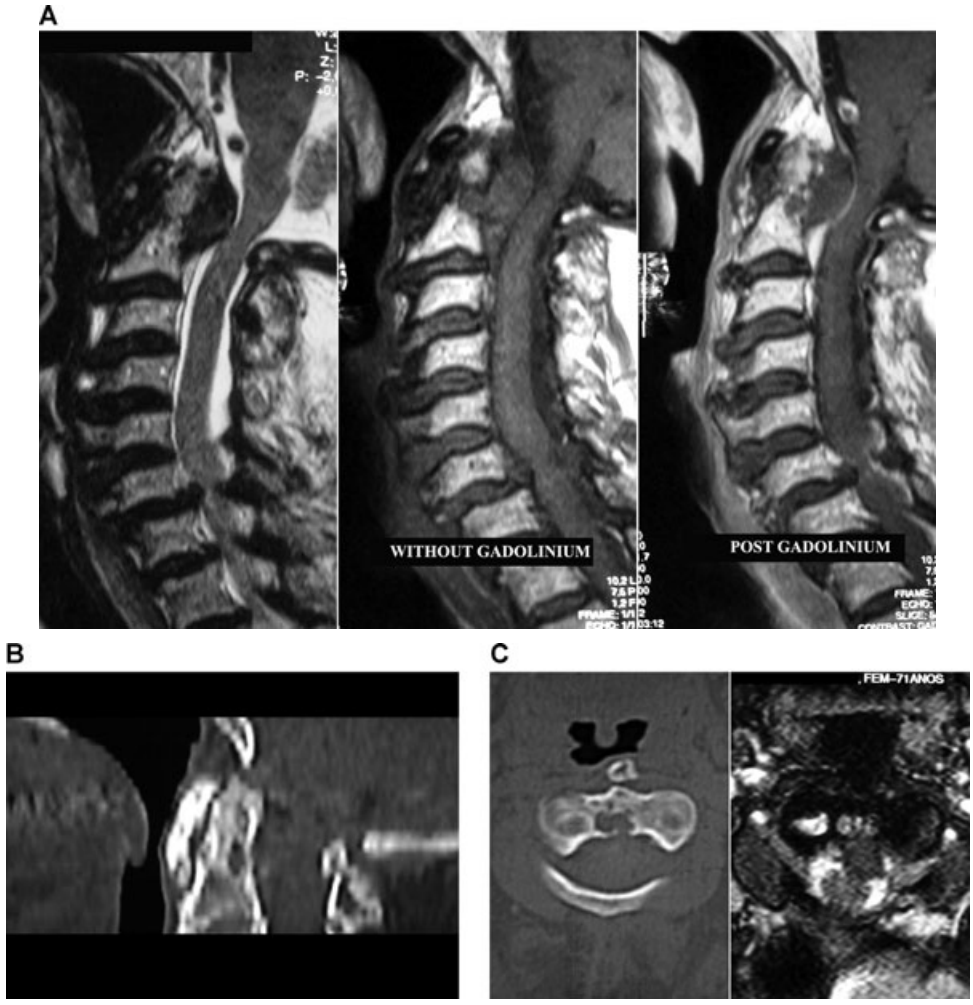

Fig 8. A 71-year-old woman with progressive tetraparesis. Sagittal MRI images disclose a periodontoid mass of heterogeneous signal intensity, predominantly isointense in T2-WI (A, left image) and hypointense in T1-WI (A, middle image), showing mild post-gadolinium enhancement (A, right image) and compressing the spinal cord against the posterior arch of C1. There are also extensive erosive changes of the odontoid process, anterior and posterior, with imminent risk of pathologic fracture. (B) A sagittal reformatted image reveals a massive prevertebral calcification, with only faint calcifications in the retro-odontoid component. (C) Axial slices at similar levels shows that CT (left) is better to demonstrate the calcified nature of the deposits and excellent to depict the erosions, whereas MRI (right) is the undisputed method of choice to describe the spinal cord compression, besides being very good to study the erosive component of the disease. Radiographs of the hands (not shown) revealed chondrocalcinosis of the triangular fibrocartilage complexes and erosive changes of the ulnar styloids, firming the diagnosis of pseudotumoral crystalline arthropathy.

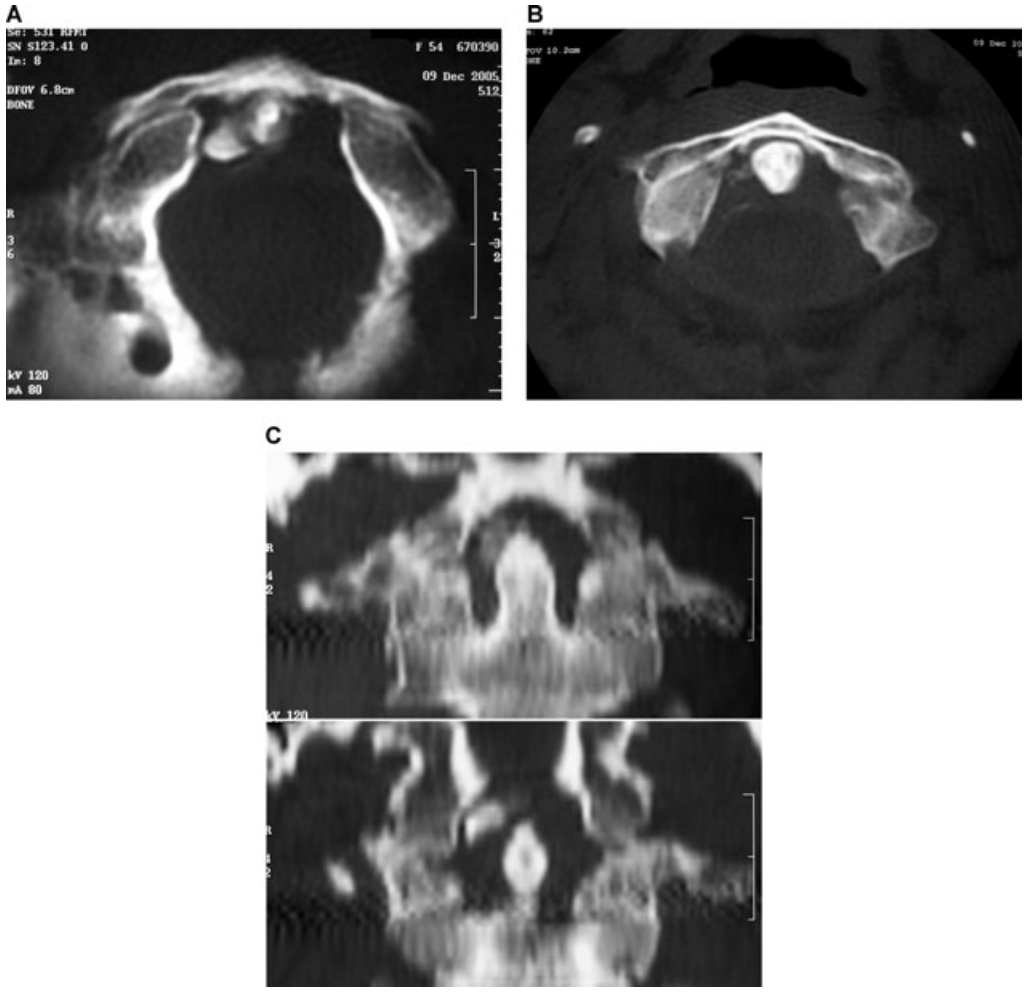

Fig 9. CT of the cervical spine performed for evaluation of cervical trauma due to motor vehicle accident. Although a densely calcified periodontoid nodule displayed in the first axial scan simulated bone avulsion (A), a lower image disclosed amorphous calcifications in this site (B), whose appearance did not suggest bone fragments. Reformatted coronal images (C) disclose an intact odontoid process, with calcifications between it and the right occipital condyle due to crystalline deposition.

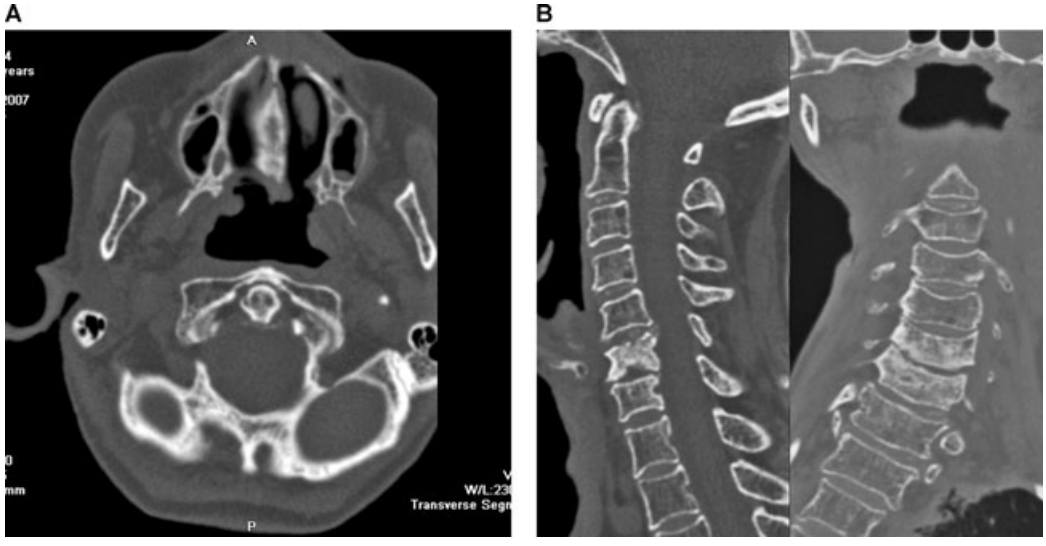

Fig 10. An axial section through the dens shows a subtle calcification of the transverse ligament (A). Reformatted images (B) show additional findings related to the crystal deposition disease: there is extensive disk calcification, mostly in C2-C3, C4-C5, C5-C6, and C6-C7; the vertebral body of C6 is sclerotic, with erosions of its endplates; and cervical scoliosis is also evident.

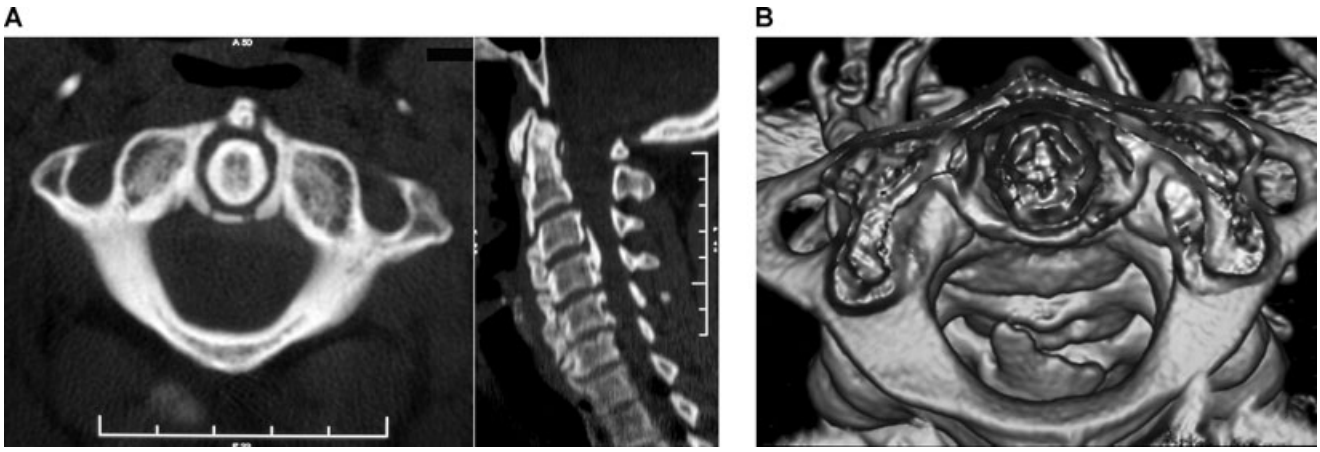

Fig 11. CT of the cervical spine of a 58-year-old woman complaining of dizziness, limited rotation of the head, and paresthesia of the upper limbs. Axial (A, left) and reformatted sagittal (A, right) images show a thick calcification of the transverse ligament associated with voluminous, flowing osteophytes and calcification/ossification of the anterior and posterior longitudinal ligaments. A volume-rendered display (B) shows in an elegant way the calcified transverse ligament and a huge calcification of the longitudinal posterior ligament protruding into the spinal canal at C3-C4 level.

Conclusion

The diagnosis of crystalline arthropathy in the periodontoid region can and should be done based on imaging, with no need for biopsy in most cases. The clinical presentation of this condition can be misleading, and imaging is crucial to a correct management. The demonstration of periodontoid calcifications at CT scans allows a safe diagnosis, MRI being the method of choice to study the neurological complications. In addition,

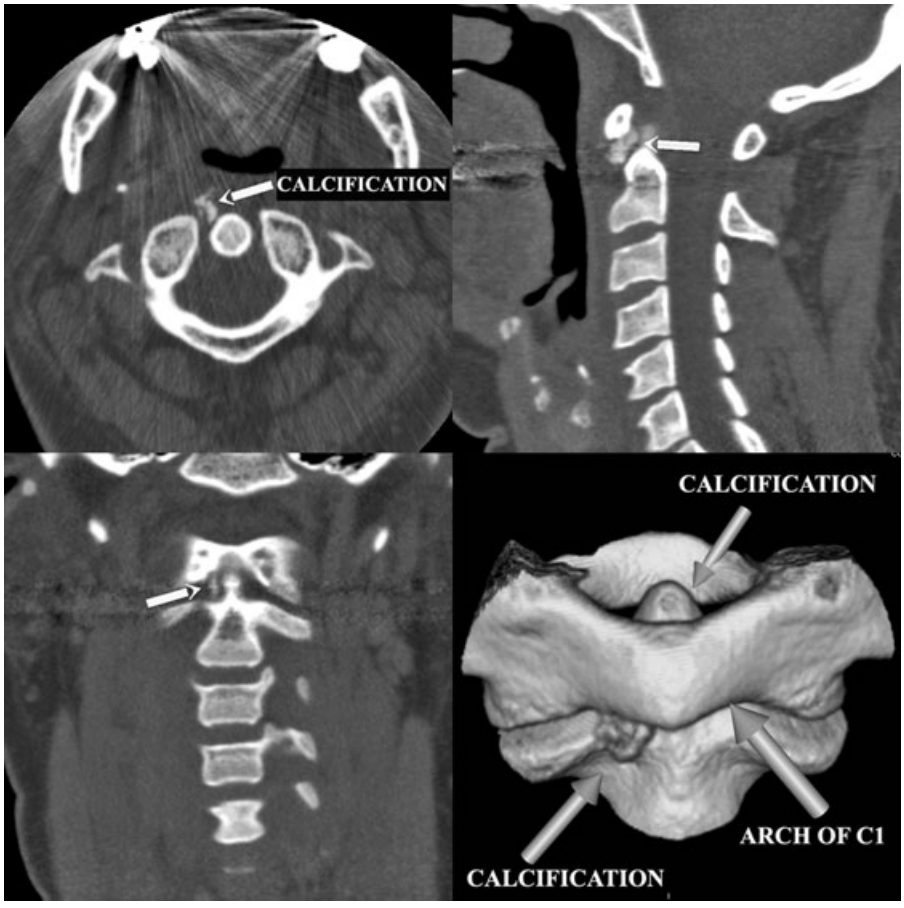

Fig 12. The periodontoid calcifications, displayed in all the three spatial planes in a CT scan of a 39-year-old woman complaining of neck pain and stiffness (arrows), are elegantly demonstrated in a volume-rendered display (lower right image) next to the tip of the dens and adjacent to the anterior atlanto-axial articulation.

the presence of bone erosions, bone marrow edema pattern, soft tissue edema, and/or post-gadolinium enhancement may indicate that periodontoid calcifications are the source of pain in a symptomatic individual.

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