Erosive Inflammatory Pseudotumor of the Odontoid Process in Association With Forestier's Disease (Diffuse Idiopathic Skeletal Hyperostosis)

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Inflammatory pseudotumor, first described in 1954,[1] was initially considered any lesion that simulated a neoplastic condition at clinical, macroscopic, and microscopic levels but that was thought to have an inflammatory and/or reactive pathogenesis. In recent literature, inflammatory pseudotumor is mostly considered a mass lesion characterized microscopically by a proliferation of a spindle-cell component against a heavy inflammatory infiltrate of mixed composition but usually with a predominance of mature lymphocyte and plasma cells.[2]

The World Health Organization accepts the term *inflammatory myofibroblastic tumor,* but, given the heterogeneity of these soft-tissue tumors, other terms, based on original body sites, are still applied.[3] This naming convention reflects the complexity and variable histologic characteristics and behavior of the entity. Pseudotumors are important at least because of their ability to mimic malignant tumors, either clinically or radiologically.[4-6] Inflammatory pseudotumors can develop in many sites but is most commonly reported in the lung, orbit, gastrointestinal tract, and kidney.[4,7] The spinal column is an extremely rare site for pseudotumors; to our knowledge, only 16 cases have been reported,[4,8-20] the last in 2005.[4] Four cases in the periodontoid area were reported.[21] The lesion was associated with diffuse idiopathic skeletal hyperostosis (DISH) in only 1 case, reported by Jun and colleagues.[19] To the authors' knowledge, atlantoaxial dislocation resulting from peri-

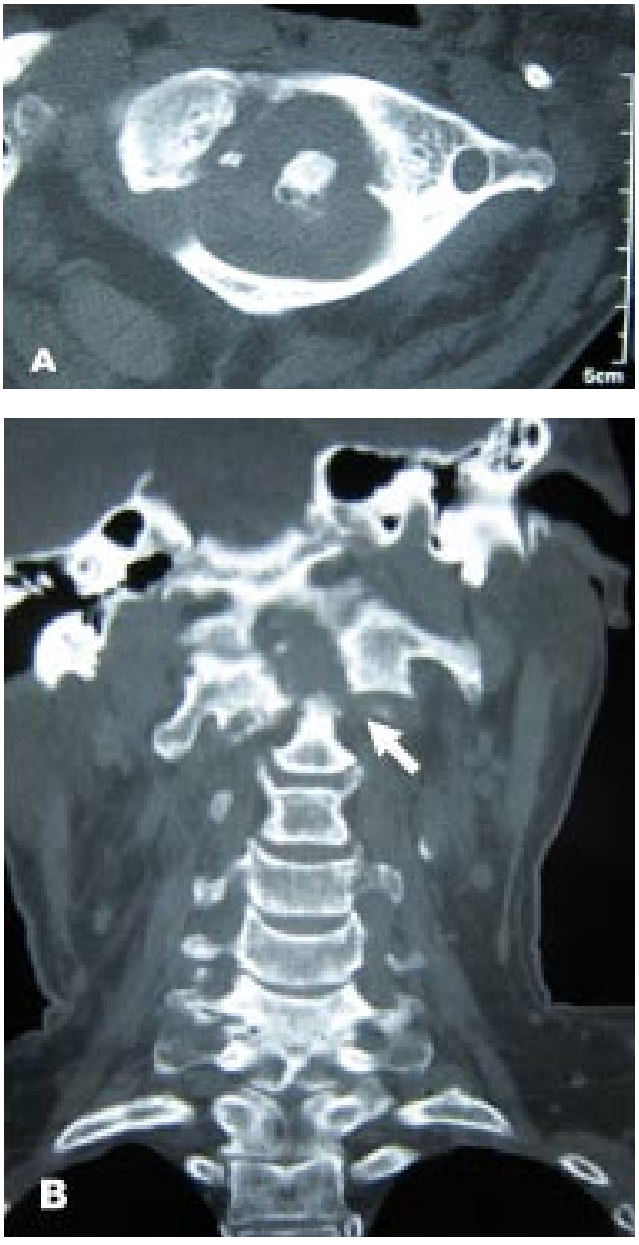

Figure 1. Computed tomography scan of cervical spine: (A) Atlantoaxial subluxation results in spinal canal narrowing; (B) arrow shows erosion of odontoid process and tilting of atlas to right.

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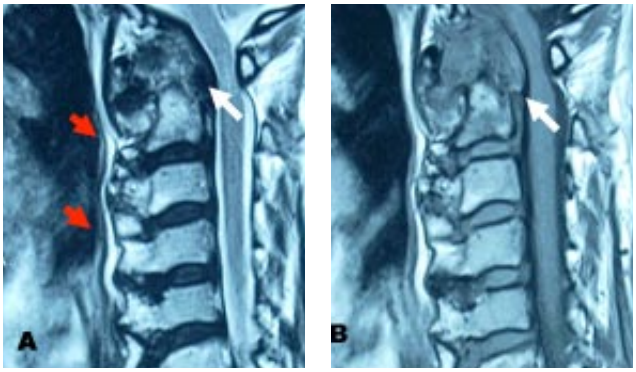

Figure 2. T_1-weighted (A) and T_2-weighted (B) sagittal magnetic resonance imaging scans of cervical region show soft-tissue mass over odontoid process (light arrows) causing cord compression at this level. The mass is predominantly isointense and hypointense on T_1- and T_2-weighted scans, respectively. Red arrows in (A) show bridging osteophytes and wavy ossification of anterior longitudinal ligament—suggesting diffuse idiopathic skeletal hyperostosis.

odontoid pseudotumor in association with Forestier's disease has not been previously reported.

DISH was first described in 1950 by Forestier and Rotés-Querol[21] as *senile ankylosing hyperostosis of the spine,* but soon it was discovered that this disorder is not limited to "senile" age groups and is not limited to the "spine," ie, there are many cases of the disorder among younger age groups and/or those with extraspinal manifestation. However, this condition, now also known as *Forestier's disease,* has a marked predilection for the axial skeleton, particularly the thoracic and lumbar spine. Although DISH is considered a benign rheumatologic disorder, it can be associated with a variety of complications ranging from pain and stiffness to different neurologic disturbances and even dysphagia.[19,22]

Case Report

A man in his mid-60s was admitted for torticollis associated with progressive bilateral upper and lower limb weakness of 4 months' duration. On admission, he was walking with support and had lost urinary control. There were no constitutional symptoms (eg, fever, loss of appetite, malaise).

The patient's medical history consisted of ischemic heart disease, hypertension, and type 2 diabetes mellitus.

Physical examination revealed marked spastic tetraparesis with muscle power of grade 2 or 3 in all limb muscles (worse on right side). Lower limbs showed marked wasting of the thigh and leg muscles. Both touch and pain sensation were decreased in a patchy pattern from the levels C4 to T4. Myelopathy signs, including hyperreflexia, clonuses, Babinski and Hoffmann signs, and spasticity, were present on both sides. Related laboratory findings showed a mild to moderate increase in erythrocyte sedimentation rate (90 mm/h) and C-reactive protein (2.03 mg/L).

The Mantoux test was not reactive, and screening tests for the autoimmune disorders (eg, Rheumatoid Factor and Antinuclear Antibody) were negative. There was no clinical evidence of rheumatoid arthritis as well. Tumor markers did not show any abnormalities.

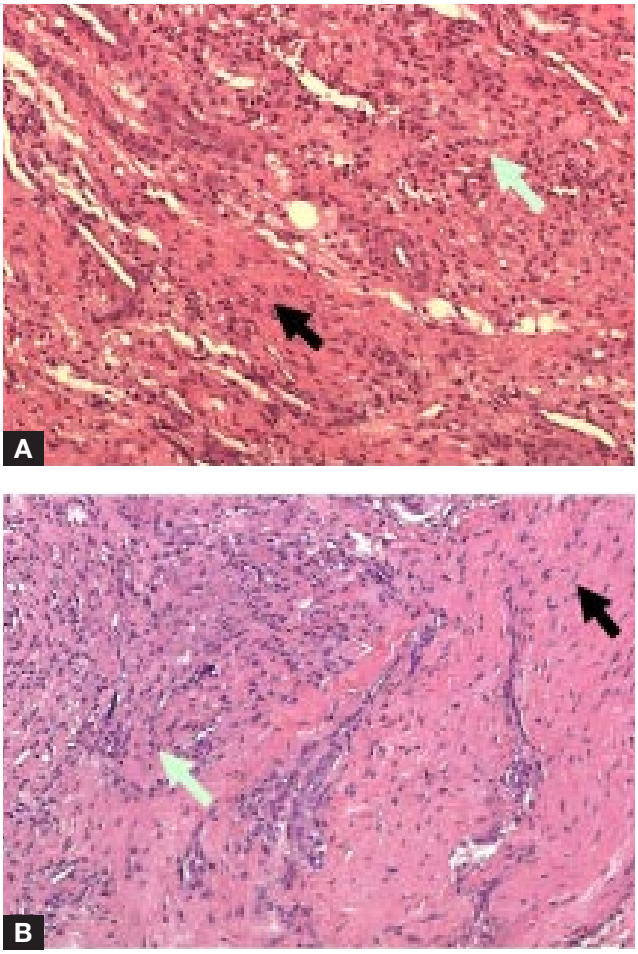

Figure 3. Polymorphic infiltrate composed of fibroblasts (black arrows) and inflammatory cells (light arrows) suggests inflammatory pseudotumor (hematoxylin-eosin, original magnification ×100).

Cervical computed tomography showed erosion of the dens of the axis and widening of the anterior and right lateral C1–C2 space with resultant narrowing of the spinal canal and the left lateral C1–C2 space (Figure 1).

Magnetic resonance imaging (MRI) showed a heterogeneously enhancing soft-tissue mass causing erosion of the dens and the inner surface of the lateral masses of the C1 vertebrae. The soft-tissue mass was predominantly isointense on T_1-weighted imaging and hypointense on T_2-weighted imaging. There was significant enhancement after gadolinium administration (Figure 2). MRI also showed patches of ossification of the posterior longitudinal ligament at the C2 to C7 levels and anterior marginal osteophytes and bridging osteophytes (syndesmophytes) along the cervical vertebral column, in keeping with DISH. Asymmetry of the semispinalis capitis muscles resulted from atrophy of the left muscle in concordance with the torticollis. A pelvic radiograph showed no signs of ankylosing spondylitis.

Given the patient's rapid neurologic deterioration, he was scheduled for posterior stabilization and autogenous bone graft (Ransford method), C1 arch decompression and foram-

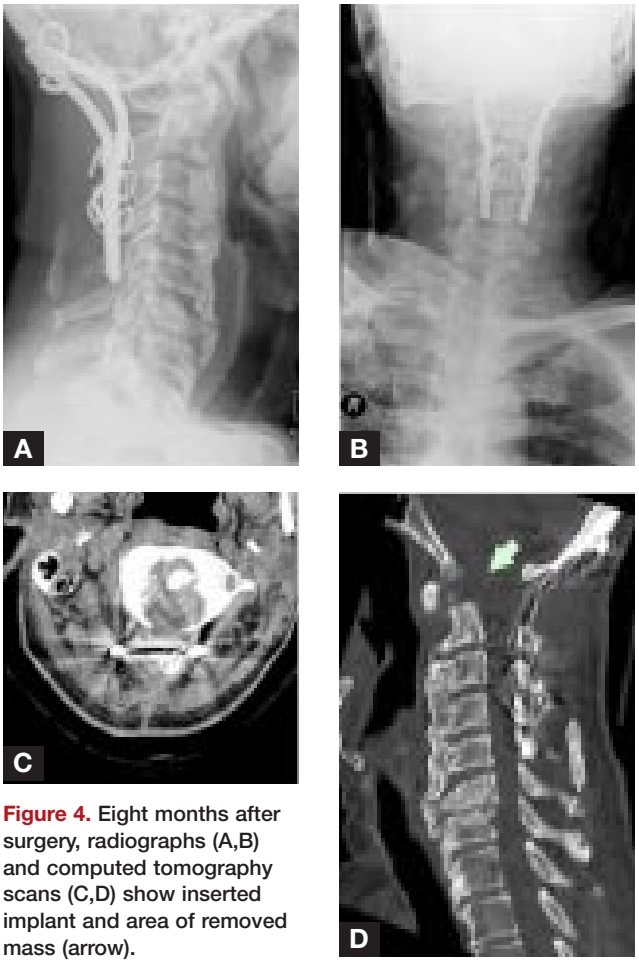

Figure 4. Eight months after surgery, radiographs (A,B) and computed tomography scans (C,D) show inserted implant and area of removed mass (arrow).

inotomy (foramen magnum), and transoral anterior decompression. Intraoperative resection revealed a soft-tissue mass that had significantly eroded the tip of the dens process. This mass was sent for histopathologic investigation. Culture and sensitivity, acid-fast bacilli, polymerase chain reaction, and fungal examinations indicated no abnormalities.

Gross histopathologic examination showed a solid, white-brown mass almost 1.5 cm in diameter. A section revealed areas of proliferating spindle-shaped fibroblasts, fibrocollagenous tissue, and inflammatory cell infiltrates composed mainly of lymphocytes and plasma cells. There was a focal proliferation of small vessels but no evidence of neoplasia, the feature characteristic of inflammatory pseudotumor (Figure 3). Six months after surgery, the patient showed significant sensorimotor improvement and was ambulating with a cane.

By 16-month follow-up, the patient's neurologic disturbances had shown some steady further improvement. The patient was walking without aid and performing his activities almost comfortably. There were no clinical or radiologic signs of tumor recurrence (Figure 4).

Discussion

In this article, we have described a rare coincidence, pseudotumor secondary to Forestier's disease in the cervical region that resulted in the first ever reported case of C1–C2 dislocation.

As for surgical management of the dislocation, some experts believe that the Ransford method is not optimal. In fact, other procedures, such as translaminar facetal screw (Magerl) fixation and newer methods (eg, Harms technique) providing more rigid fixation over the atlantoaxial joint, can also be considered as other valuable alternatives. In addition, there are arguments regarding the need to obtain flexion-extension radiographs to ensure reducibility of the C1–C2 dislocation. In our patient's case, we did not do preoperative flexion-extension radiography. The chosen procedure and investigations were based on the previous experiences and skills of the corresponding surgeons (three spine surgeons and one neurosurgeon) at the time of admission.

Although pseudotumors have been found in every organ, they seldom occur in the vertebral column. Our patient's case shows that, when this disorder occurs in the high cervical region (ie, the tip of the odontoid process), it can be associated with anatomical disarrangements that may eventually jeopardize atlantoaxial integrity and subsequent C1–C2 subluxation. It is interesting that the retro-odontoid lesion reported by Jun and colleagues[19] was also associated with neurologic complication secondary to spinal cord compression. The other common, but probably much more important, feature of these two cases was association of pseudotumor with DISH in "high cervical vertebra," the place where DISH does not normally occur.

DISH is a common rheumatologic problem that occurs most often in the thoracic and lumbar spine. The cervical spine, the site of the disorder in our patient's case, is least often affected.[23] Myelopathy associated with Forestier's disease is usually reported secondary to protrusion of the hyperostotic formation into the spinal canal or ossification of the posterior longitudinal ligament or yellow ligament[24]; therefore, C1–C2 joint disruption is not a well-known complication of Forestier's disease. Some authors believe that DISH is not necessarily a disease and may even have some protective role against degenerative changes (ie, lumbar osteoarthritis).[22,25]

Considering the high incidence of DISH in elderly men (≤27%[26]), particularly those with a diabetic background, this finding was probably a primary factor in the emergence of our patient's coexisting condition—inflammatory

"We emphasize the probable role of DISH in the cervical region as an etiologic factor for the emergence of pseudotumors in the peri-odontoid region and subsequent dislocation of the atlantoaxial joint."

pseudotumor in the atlantoaxial joint. Although the precise mechanism of this association is still not clear, we believe that the microbiomechanical effects that DISH can exert in the cervical region may play a role in this situation. Indeed, periodontoid pseudotumors may result from repeated injuries to the C1–C2 joint secondary to the abnormal forces transferred from the rigid cervical vertebra to the single mobile portion of the cervical axis still available: C1–C2. This conclusion is in concordance with what was first suggested by Jun and colleagues,[19] as DISH can be a causative agent for either fracture or pseudotumor of the odontoid process. This hypothesis needs to be proved through further investigations into pseudotumor pathophysiology and through biomechanical studies of Forestier's disease.

Authors' Disclosure Statement and Acknowledgments

The authors report no actual or potential conflict of interest in relation to this article.

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Our patient gave us informed consent to publish this report.

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This paper will be judged for the Resident Writer's Award.