Tumoral Calcinosis Presenting as Neck Pain and Mass Lesion of the Cervical Spine

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In this article, we present the case of a woman in her early 50s who presented with neck pain. Imaging studies showed an expansile, lobulated, calcified mass in the posterior elements of C2-C3. An open biopsy was performed, and the removed tissue demonstrated tumoral calcinosis. We discuss tumoral calcinosis, including its etiology, radiographic features, and differential diagnoses.

Case Report

A woman in her early 50s presented to her family care physician with a 4-month history of right-side neck pain that was worse at night. No history of neck trauma was recalled. She had been diagnosed with scleroderma 2 years earlier and secondarily had developed interstitial lung disease and chronic renal failure, for which she was undergoing hemodialysis. She was referred to an orthopedic surgeon, who ordered plain cervical radiographs. These showed a lobulated calcified mass involving the posterior elements of the cervical spine. As a neoplastic process could not be ruled out, and the patient was being considered for a kidney transplant, a definitive diagnosis was imperative. She was therefore referred to an orthopedic oncologist for further evaluation. Physical examination revealed limited neck range of motion and tenderness over the right paraspinal musculature and right suboccipital area but no palpable mass. Neurologic examination findings were normal. In the lower gluteal area were bilateral, firm, plaque-like subcutaneous nodules. Laboratory findings were normal except for blood urea nitrogen of 37.0 mg/dL, creatinine of 9.3 mg/dL, serum calcium of 8.1 mg/dL, and phosphorus of 6.7 mg/dL.

Cervical spine plain radiographs (Figure 1), computed tomography (CT) scans (Figure 2), whole-body bone scan (Figure 3), and magnetic resonance imaging (MRI) scans

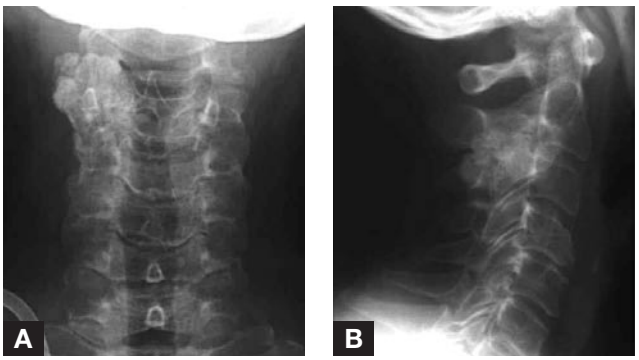

Figure 1. Plain anteroposterior (A) and lateral (B) radiographs of cervical spine show lobulated calcified mass at right C2–C3 facet joint.

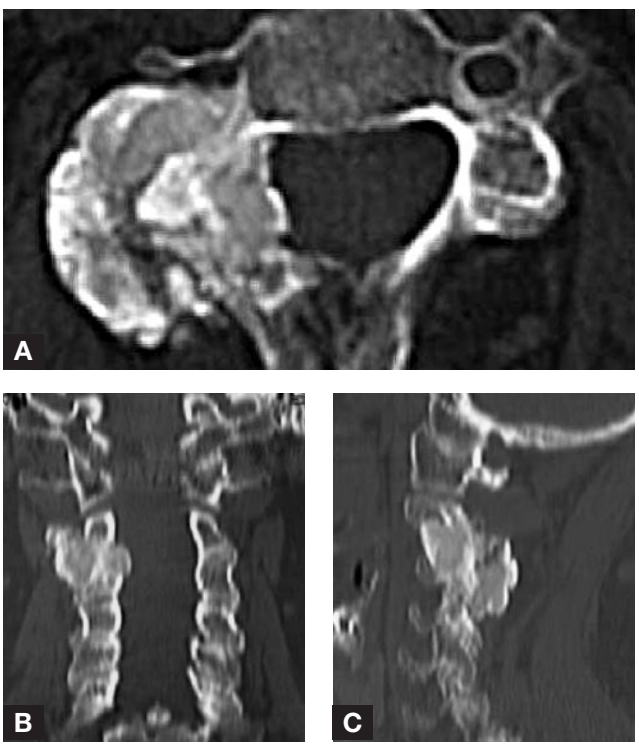

Figure 2. Axial (A), coronal (B), and sagittal (C) computed tomography scans show large calcified mass (with sclerotic rim and slight intraspinal extension) at right C2–C3 facet joint, pedicle, and lamina.

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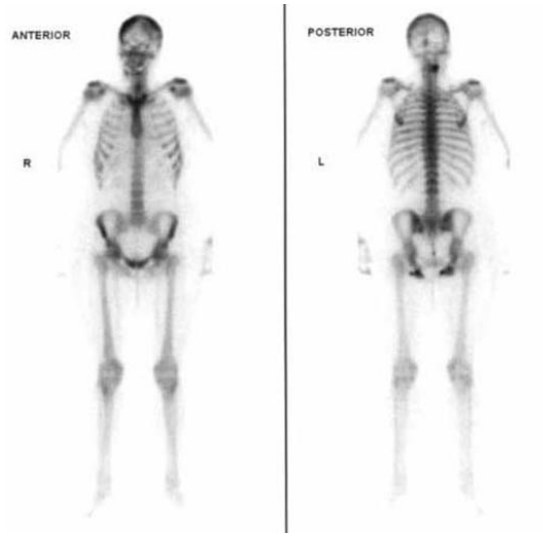

Figure 3. Whole-body bone scan shows increased uptake to right of midline in midcervical region and symmetric, increased uptake within inferior aspects of gluteal region.

(Figure 4) were obtained. The radiographs showed a lobulated, calcified lesion of the cervical spine at C2–C3; the CT scans showed an expansile calcified mass at the right-side posterior elements of C2–C3, with lateral encroachment into the spinal canal; the bone scan showed increased activity in the midcervical spine on the right, plus symmetric, increased uptake in the gluteal region; and the MRI scans showed edema within the arch of the right C2 and a mass involving the posterior elements of C2 and C3, near the facet joint.

Given the history, physical examination findings, laboratory test results, and imaging studies, the differential diagnosis includes osteoblastoma, osteoid osteoma, tumoral calcinosis, exuberant callus, and osteochondroma.

"The differential diagnosis included osteoblastoma, osteoid osteoma, tumoral calcinosis, exuberant callus, and osteochondroma."

An orthopedic spine surgeon helped to biopsy the mass. During the procedure, a fair amount of gelatinous, chalky material exuded and effectively evacuated the majority of the lobulated mass. The removed chalky tissue was sent for histologic examination (Figure 5). Histologic sections revealed fibrous tissue with amorphous, calcified masses. No tumor cells were seen (Figure 4). The diagnosis of tumoral calcinosis of the cervical spine secondary to scleroderma was made. At most recent follow-up (10 months), the patient reported no neck pain, stiffness, or radicular symptoms. Plain

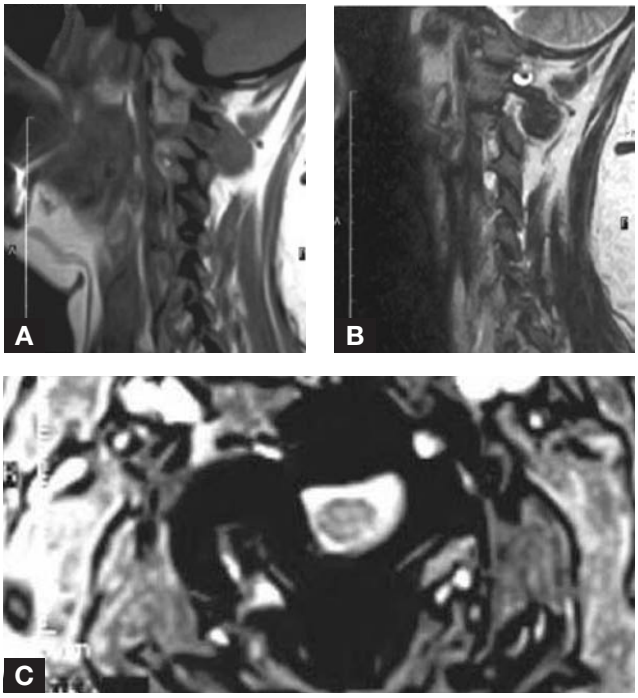

Figure 4. Magnetic resonance imaging (MRI) scans of cervical spine: (A) T_1-weighted sagittal scan shows uniformly low signal intensity lesion encroaching on right C2–C3 facet joint. (B) T_2-weighted sagittal scan shows lesion maintaining low signal intensity. (C) T_2-weighted axial scan shows involvement of right C2–C3 facet joint of low signal intensity lesion. There are no significant impingement into the spinal canal and no abnormal signal in the spinal cord.

radiographs showed good maintenance of alignment (Figure 6) and no recurrence of the mass.

We have obtained the patient's informed, written consent to publish her case report.

Discussion and Treatment

The differential diagnosis for symptomatic, calcific spine masses that on radiographs appear to expand the bone comprises osteoblastoma, osteoid osteoma, exuberant callus, and osteochondroma. In a patient with a history of scleroderma, a disease associated with soft-tissue calcifications, tumoral calcinosis should be included in this list. Although there is some inconsistency as to the pathophysiologic mechanism or mechanisms involved in tumoral calcinosis, there is general agreement as to the appearance of the lesion on imaging studies. Tumoral calcinosis is a tumor-like deposition of dense, calcified masses, often with a lobular appearance, in the tissues around large joints, though it may be associated with smaller masses in the spine and other joints. These masses may not be easily visualized on plain radiographs, but on MRI they are distinguished by their hypointensity in both T_1- and T_2-weighted sequences. The physical examination findings of firm, subcutaneous plaques in the gluteal region are consistent with the systemic nature of the underlying disease in this patient and are not explained by the other differentials. Last, the gross finding of chalky material and the histologic examination

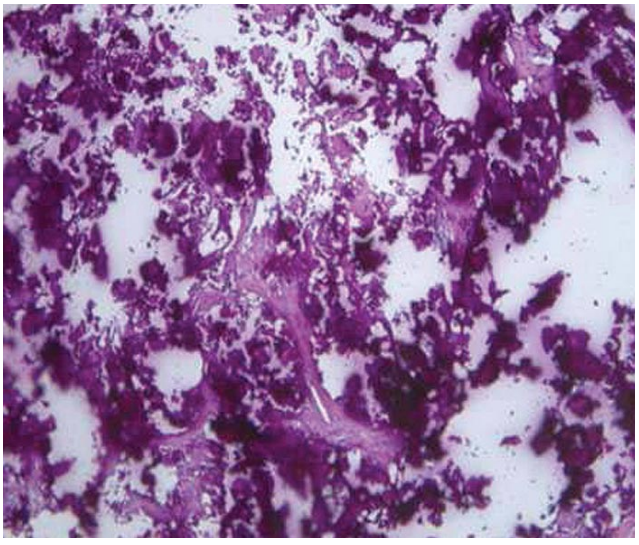

Figure 5. Fibrous tissue with amorphous calcification and no visible tumor cells (hematoxylin-eosin, original magnification ×100).

finding of amorphous, calcified masses in the absence of neoplastic cells confirmed the diagnosis.

Osteoid osteomas and osteoblastomas are benign bone lesions that typically present with pain that is worse at night. Osteoblastomas are larger than osteoid osteomas (usually ≥2 cm) and in aggressive cases may show extensive lytic destruction of bone and cortical breakthrough.[1,2] Osteoblastomas in the spine have a predilection for the posterior elements. In the cervical region, they may present with torticollis and restriction of neck motion.[3] Bone scan may reveal increased uptake, and CT scans show a reactive sclerotic bone surrounding a central nidus.[4] However, no central nidus was visible in our patient's imaging studies.

Solitary osteochondroma usually presents as an asymptomatic bony mass. It commonly arises from the cortical surface of long bones. It may also involve the cervical spine, typically the posterior elements (more than 70 cases have been reported in the orthopedic literature since 1907[5]). Described as sessile, polypoid, and cauliflower-like,[6] some lesions may extend from the vertebral surface into the neural foramina, causing neural impingement. In our patient's case, CT scans did not show a confluence of the mass and the medullary portion of the host bone.

Presence of an exuberant callus at the site of an undetected previous fracture at C2–C3 has been reported[7] with symptoms of occipital neuralgia. Although our patient denied any history of trauma, trauma was considered a possibility given the similarity in radiographic appearance and the symptomatology at initial presentation. CT scan usually shows a fracture line, but this was not seen in our patient's case.

Various types of soft-tissue calcifications have been reported.[8,9] Boulman and colleagues[10] noted that soft-tissue calcifications "may represent a nonspecific local response or be a manifestation of a complex underlying disease" and classified them into 5 categories: metastatic, tumoral, dystrophic, calciphylaxis, and idiopathic. According to Boulman and colleagues:

Metastatic calcification is associated with an abnormality in calcium metabolism and occurs as firm, subcutaneous nodules near large joints. These nodules usually involve normal tissues and are associated with conditions such as hyperparathyroidism, milk-alkali syndrome, and hypervitaminosis D.

Tumoral calcification is a rare familial disorder associated with increased phosphorus and normal to decreased calcium levels. It tends to occur as large, subcutaneous calcium deposits near joints or pressure areas.

Dystrophic calcification, which includes calcinosis, occurs in the presence of normal metabolism as subcutaneous nodules, plaques, or extensive deposits in tissues "damaged" by trauma, infection, lupus, scleroderma, or dermatomyositis.

Calciphylaxis is commonly found in patients with chronic renal failure with abnormalities in the serum calcium-phosphate levels. Small-vessel vasculopathy with intimal fibrosis and thrombosis leads to tissue ischemia and necrosis.

Idiopathic calcification occurs in the presence of normal calcium metabolism as asymptomatic, subcutaneous nodules in otherwise healthy individuals.

Scleroderma, or progressive systemic sclerosis, is associated with calcinosis, or soft-tissue calcifications, in 9% to 27% of cases,[11,12] though usually of the limited cutaneous type. Several cases of calcinosis affecting the spine in patients with scleroderma have been reported. Schweitzer and colleagues[13] reported on 5 patients with lobulated calcifications of the cervical spine—4 of whom

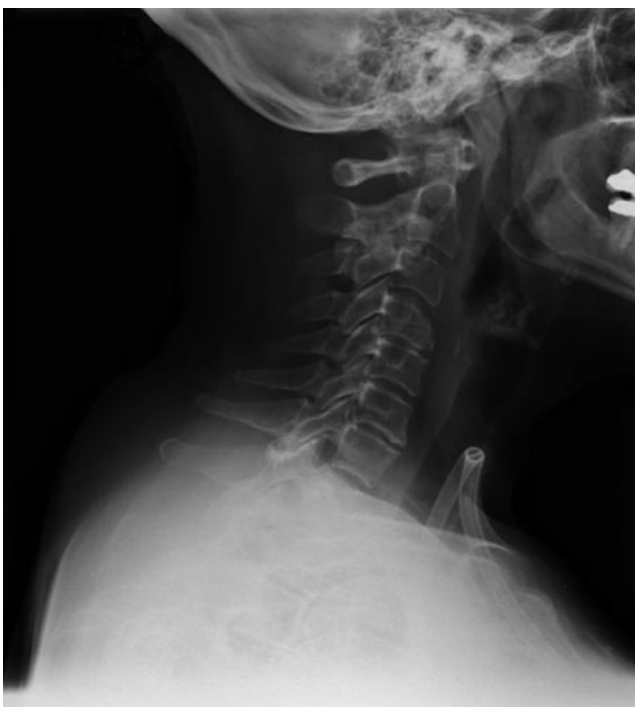

Figure 6. Plain lateral radiograph of cervical spine at most recent follow-up shows good alignment maintained.

had scleroderma. Ojemann and colleagues[14] reported a case with diffuse calcifications at the C2–C3 and C7–T1 facet joints in a patient who developed full-blown CREST (calcinosis, Raynaud phenomenon, esophageal motility disorders, sclerodactyly, telangiectasia) syndrome 1 year later. Walden and colleagues[12] reported diffuse paravertebral and intraspinal calcifications in the thoracic spine "mimicking tumoral calcinosis" in a patient with scleroderma. Inclan[15] first used the term *tumoral calcinosis* to describe the tumor-like deposition of dense, nodular, calcareous masses in the tissues about the hips, shoulders, and elbows, sometimes with smaller masses adjacent to the spine and other joints. Smit and Schmaman[16] described 2 cases and stated that tumoral calcinosis does not involve the skin and is not associated with collagen disease. Nevertheless, the tumoral calcinosis diagnosis has been applied in some cases in which these lesions were found in patients with scleroderma. Shibuya and colleagues[17] reported symmetric, calcifying tumor–like lesions at the L3–L4 facet joints in a patient with scleroderma, and they referred to this as a case of tumoral calcinosis. Reporting on one of the largest series of tumoral calcinosis of the spine, Durant and colleagues[18] stated that pathology is caused by dystrophic calcification in the soft tissues; 1 patient in that series had scleroderma. Teng and colleagues[19] described tumoral calcinosis of the cervical spine in a patient with CREST syndrome.[20,21]

Our patient's past medical history was complicated by chronic renal failure requiring hemodialysis. Widespread calcium deposition in soft tissue is commonly seen in renal failure with or without hemodialysis.[22] The etiology has largely been identified as secondary hyperparathyroidism, increased serum phosphate levels, and decreased serum calcium levels. In 2 cases reported by McGregor and colleagues,[23] tumoral calcinosis–like lesions were found in the setting of renal failure and hyperparathyroidism, albeit in the sacrum and the foot. Clearly, *tumoral calcinosis* is often used in the orthopedic literature as a morphologic description of dense, calcific masses about the joints and spine with various proposed etiologies. The classification scheme of Boulman and colleagues,[10] on the other hand, is based on pathophysiologic mechanism. The etiology of our patient's tumoral calcinosis may represent an overlap between dystrophic calcification and calciphylaxis (Boulman and colleagues), which may also account for the plaque-like lesions in the gluteal region.

Tumoral calcinosis of the spine in scleroderma may present as painful, calcific masses that appear to expand the bone. These masses may cause osteolytic destruction of the posterior arches and cause spinal cord compression and instability.[18,24,25] In some cases, they are not readily visible on plain radiographs but can be seen as hypointense lesions on T_1- and T_2-weighted sagittal MRI scans.[18] They may strongly resemble tumors (eg, osteoblastomas) or even infections.[18,19] Qadri and colleagues,[26] highlighting some of the differentiation pitfalls, stated that tumoral calcinosis should remain in the differential diagnosis of any calcified spinal compressive lesion. Because the differential diagnoses of neoplasms and infections often require histologic confirmation, a biopsy is performed.

Destructive cervical spine lesions related to scleroderma are rare and may require complex spine reconstruction in the setting of severe refractory pain, neurologic symptoms, or instability.[25] In these cases, lesion resection followed by spinal fusion with instrumentation has been advocated. Local recurrence in the spine has not been reported.[26] In the 21-patient series reported by Durant and colleagues,[18] 9 patients underwent laminectomies for spinal decompression, 3 underwent corpectomies for compression fractures, and 8 underwent anterior and posterior spinal fusions for spinal instability.

"Qadri and colleagues[26]...stated that tumoral calcinosis should remain in the differential diagnosis of any calcified spinal compressive lesion."

Our patient had no neurologic deficit or spinal instability. Persistent drainage of chalky fluid has been reported after percutaneous biopsy with needles and trocars.[25] Given this finding in our patient's case, and her skin being compromised by scleroderma, we elected to perform an open biopsy with a small incision that was localized with fluoroscopy during surgery. After an adequate specimen was obtained, additional material exuded from the biopsy site and was thus effectively evacuated. A small amount of bone wax was applied over the exposed bony surface, and the fascia was carefully closed under direct visualization. After the biopsy findings established the diagnosis, the patient was given weak opioid analgesics for pain relief. Follow-up radiographs showed no evidence of instability at the involved motion segment.

After completing a course of physical therapy, the patient was asymptomatic. The exact reason for her dramatic pain relief is unclear. We speculate that the large lobulated mass was irritating the posterior cervical musculature. Given that the biopsy findings confirmed a benign pathology, the patient was able to get on the kidney transplant waiting list. She will undergo serial clinical and radiographic follow-up to monitor for disease progression and signs of spinal instability.

Authors' Disclosure Statement and Acknowledgment

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

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