Systemic sclerosis-associated compressive cervical calcinosis: intersection of degenerative spine instability, lesional spinal cord compression, and traumatic spinal cord injury. Illustrative case

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BACKGROUND Tumoral calcinosis, mass-like calcium deposition into the soft tissues, is an uncommon manifestation of the systemic sclerosis subtype of scleroderma. When this process affects the spinal epidural space, it can cause canal narrowing and place the spinal cord at significant risk of injury.

OBSERVATIONS Here a 62-year-old female with systemic sclerosis and no previous evidence of spinal cord compromise who developed acute spinal cord injury and quadriparesis after a mechanical fall is described. She was found to have a large dorsal epidural calcified mass compressing her cervical spinal cord. She underwent medical management for acute spinal cord compression as well as surgical management for acute spinal cord injury and degenerative spine disease. Her case illustrates a rare etiology of simultaneous degenerative spine instability and lesional spinal cord compression with acute spinal cord injury.

LESSONS Tumor calcinosis leading to acute spinal cord injury in the setting of systemic sclerosis is an uncommon but critical entity to recognize in patients with scleroderma and may require the physician to use a combination of medical and surgical management strategies from each of these categories of spine pathology.

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KEYWORDS scleroderma; spinal cord injury; systemic sclerosis; trauma; calcinosis

Abbreviations

AIS = American Spinal Cord Injury Association Impairment Scale; ASIA = American Spinal Cord Injury Association; CT = computed tomography; MRI = magnetic resonance imaging; SCI = spinal cord injury.

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tumoral calcinosis. This abnormal calcified tissue acts as a mass adjacent to the spinal cord and destabilizes the bony spine, thereby setting up the perfect scenario for spinal cord injury. This presentation represents a multifactorial cause of SCI with the compressive features of malignancy, spinal instability of degenerative spondylosis, and acute management concerns of acute traumatic spinal cord injury.

Tumoral calcinosis was first described in the literature in 1898 to describe a familial, hereditary condition of mass lesions that affect the periarticular space. However, it was not until 1943 that this entity was given the name tumoral calcinosis. Since then, the term has been used to describe not only the familial pattern of presentation, but also the similar deposition of calcified material with associated inflammation and fibrosis in the tissues in other diseases as well. Here, we describe a rare case of tumoral calcinosis in the cervical spine resulting in an acute spinal cord injury. Although the literature has described other cases of calcinosis in patients with systemic sclerosis, none have presented such an aggressive case of calcinosis deposition in the epidural space, with near obliteration of the spinal canal coupled with degenerative instability and associated paraspinous calcinosis changes. Features of both malignant tumoral spinal cord compression and acute traumatic spinal cord injury must be addressed and balanced against each other in management.

Illustrative Case

A 62-year-old female with limited systemic sclerosis diagnosed 9 years prior to presentation had symptoms that were controlled on oral prednisone. She had no previous symptoms of myelopathy, although she did have baseline restricted movement in bilateral wrists, hands, and fingers from her scleroderma. She suffered a mechanical ground level fall with head strike, neck hyperextension, and loss of consciousness. After the fall, she experienced immediate quadriplegia and she was transferred by ambulance to the nearest hospital, not our facility. Upon presentation, it was documented that she had 0/5 movement in her upper and lower extremities as well as absent sensation below the neck.

Computed tomography (CT) scanning of the cervical spine revealed spondylolisthesis of C3 on C4. In addition, there was evidence of disc degeneration C4–5 and C5–6 and loss of cervical lordosis. Calcified mass-like material was seen in the dorsal epidural space from C3 to C5 causing significant canal stenosis. Calcified mass-like material was observed in the bilateral parafacet joint and paraspinous regions C2–3 through C7–T1 levels as well (Fig. 1). Magnetic resonance imaging (MRI) of the cervical spine revealed severe spinal canal stenosis C3–6 (Fig. 2).

The patient was treated with intravenous steroids and monitored. She began to show some improvement in motor and sensory function and 4 days later was transferred to our facility for acute inpatient rehabilitation. At that time, clinically, she had an incomplete SCI at the C3 level, and an American Spinal Cord Injury Association (ASIA) Impairment Scale (AIS) score of D. On physical examination, her strength in bilateral biceps and triceps was 4/5. Finger flexors, wrist extensors, and finger abductors were unable to be tested because of underlying contractures from scleroderma. Iliopsoas strength bilaterally was 4/5. The rest of her lower extremity muscles on the right were 5/5 strength. On the left, quadriceps, extensor hallucis longus, and gastrocnemius muscles were 4/5 strength while the left tibialis anterior was 3/5 strength. 

FIG. 1. Preoperative sagittal (A and B) and axial (C and D) CT scans. The dorsal epidural tumoral calcinosis mass extends across the C3–5 levels causing central canal stenosis (A and D). There are also multilevel calcified lesions involving the facet joints and parafacet regions bilaterally (B and C). In addition, diffuse degenerative changes and spondylolisthesis C3 on C4 (A) can be appreciated.

FIG. 2. Preoperative sagittal (A and B) and axial (C and D) magnetic resonance (MR) images showing severe spinal canal stenosis and spinal cord compression due to dorsal epidural calcified mass lesion. T2-weighted (A, C, and D) and T1-weighted (B) MR images are featured.
Her physical examination and imaging were consistent with acute central cord syndrome secondary to severe spinal canal stenosis from likely tumoral calcinosis. In addition, she had severe degenerative spinal instability as a result of the calcinosis. She was taken to the operating room for cervical decompression with C3–5 laminectomies and removal of dorsal epidural tumoral calcinosis, as well as spinal stabilization with C3–7 posterior spine fusion and lateral mass instrumentation (Fig. 3). The calcinosis deposits encountered intraoperatively in the C3–4 through C6–7 parafacet and paraspinous regions were soft and had the semisolid consistency of toothpaste. These were debulked by suction and careful dissection of the tissue away from the surrounding normal structures. Next, the lateral mass screws were placed from the C3–7 levels. Then, laminectomies were performed using Leksell rongeurs and a high-speed matchstick drill. The dorsal epidural calcium mass encountered upon removal of the laminae was solid and flaky. It appeared to have replaced the ligamentum flavum and was severely indenting the thecal sac. It was mildly adherent to the dural surface. Once this was removed, the spinal cord was visually inspected and had reexpanded dorsally.

Postoperatively, the patient returned to the acute rehabilitation service where her strength and sensation continued to improve. Six months after the operation, she had regained full strength in all muscle groups in upper and lower extremities—sparing her baseline scleroderma-related contractures and stiffness in bilateral wrists, hands, and fingers—and had return of normal sensation.

**Discussion**

**Observations**

SCI from tumoral calcinosis of systemic sclerosis presents a unique disease state in which one must consider aspects of compressive spine malignancy, spinal degenerative disease, and acute traumatic spinal cord injury to determine the most clinically efficacious intervention(s). For the patient presented here the calcinosis acted as a mass lesion, similar to malignancy. Spinal cord compression from tumor is often treated with steroid administration for spinal cord edema. However, consensus for treatment of traumatic central cord syndrome patients, often due to underlying degenerative spinal canal narrowing, favors blood pressure elevation to support perfusion of the spinal cord. In addition, for those patients with spinal cord injury and ongoing compression, similar to patients with acute traumatic SCI, rapid surgical decompression is indicated. And finally, because of the spondylolisthesis, spine instrumentation and fusion is indicated. Our patient completed a course of steroids after the dorsal epidural mass lesion was identified at the presenting hospital and later, after transfer to our facility, underwent decompression and spine fusion.

**Lessons**

Tumoral calcinosis of the spine is a nonspecific but uncommon finding in systemic sclerosis or scleroderma, CREST syndrome (calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia), primary and secondary hyperphosphatemia syndromes, renal failure leading to uremia, rheumatoid arthritis, and other idiopathic etiologies. The calcinosis of systemic sclerosis is due to calcium hydroxyapatite deposits in connective tissues. This effectively converts connective tissues of the spine (including ligamentum flavum, facet capsule, muscle) into calcified structures. When these calcifications form mass lesions, they are termed tumoral calcinosis. Unlike other causes of spinal degenerative disease and SCI including disc herniations and ossified posterior longitudinal ligament, systemic sclerosis affects predominantly the dorsal structures of the spine. This leads to accelerated degenerative instability and spondylolisthesis of the spine at the levels affected by calcium deposition.

Depending on the morphology and location of the lesions, patients may present with a variety of symptoms ranging from pain to spinal cord dysfunction. Spinal cord compression in scleroderma-associated calcinosis is rare. Motegi et al. report that of 159 patients with systemic scleroderma, 27 (17%) had spinal calcinosis. Of those with spinal calcinosis, 7 of 27 (25.9%) had spinal cord compression from calcinosis, which represents 4.4% of patients with systemic scleroderma who have spinal cord compression.

Spinal calcinosis in patients with scleroderma remains an uncommon, although potentially neurologically devastating entity. Therefore, we conducted a review of the literature of cervical spine calcinosis in patients with scleroderma/systemic sclerosis over the last 20 years (since 2000). We performed a PubMed search of the published literature and also reviewed the reference lists of articles returned from this search. We limited our search to patients with a confirmed diagnosis of scleroderma or systemic sclerosis; cases in which individual patients, their symptoms, imaging, and treatment courses, could be identified and reviewed; and English language articles. This returned 19 cases (Table 1). Fifteen of 19 (78.9%) affected patients were women, suggesting a female predominance. Three patients were treated nonoperatively and none of these patients had spinal canal stenosis due to the lesions. Their lesions were confined to the paraspinous regions and/or the facet joints. Only nine cases in the literature (47.4%) reported spinal canal compromise due to the calcinosis, and one case resulted in neural foraminal narrowing.

Sixteen patients were treated surgically. The majority (n = 13) of patients were treated using a posterior surgical approach, either fusion alone or decompression and fusion depending on whether or not the neural elements were compromised. One patient was treated with an isolated anterior approach and two were treated with a combined anterior and posterior approach. Of those who did undergo surgical intervention, 10 patients (84.2%) required instrumentation for spine stability whereas six had decompressions alone or isolated resection of the calcified lesions. Cases reviewed did not discuss whether patients underwent medical interventions of either blood pressure support or of steroid administration during hospitalization.

**FIG. 3.** Postoperative anteroposterior (left) and lateral (right) cervical spine radiographs following C3–5 laminectomies, debulking of calcified mass, and C3–7 posterior spine fusion and instrumentation.
<table>
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<tr>
<th>Authors &amp; Year</th>
<th>Sex</th>
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<th>Imaging Findings</th>
<th>Surgical Intervention</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Durant et al., 200114</td>
<td>M</td>
<td>48</td>
<td>Unknown</td>
<td>C3 on C4 subluxation, C3–4 pst element calcinosis, spinal cord compression</td>
<td>Cervical laminectomies (unknown levels), pst spine fusion (unknown levels)</td>
<td>Unknown</td>
</tr>
<tr>
<td>Olsen &amp; Chew, 200415</td>
<td>F</td>
<td>75</td>
<td>Torticollis</td>
<td>Calcinosis of rt C2–4 facet joint, paraspinous region, &amp; transverse process; calcinosis mass extending to anterior cervical muscle space at C2–4 levels compromising the esophagus</td>
<td>“Excision of the mass”</td>
<td>Unknown</td>
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<tr>
<td>Khan et al., 200522</td>
<td>F</td>
<td>64</td>
<td>Neck pain, acute quadripareis</td>
<td>Calci ...</td>
<td>C3–7 laminectomies, C2–7 pst fusion</td>
<td>Resolution of neck pain, improved strength</td>
</tr>
<tr>
<td>*Lima et al., 200623</td>
<td>F</td>
<td>51</td>
<td>Weakness</td>
<td>Calci ...</td>
<td>Cervical &amp; thoracic laminectomies (unknown levels)</td>
<td>Unknown</td>
</tr>
<tr>
<td>*Smucker et al., 200624</td>
<td>M</td>
<td>60</td>
<td>Neck pain, hand dysfunction, gait difficulty</td>
<td>C2–3 subluxation</td>
<td>Halo, C3 corpectomy &amp; anterior fusion, C1–4 pst onlay fusion</td>
<td>Unknown</td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>59</td>
<td>Neck pain</td>
<td>C4–5 destructive lesion involving pst elements</td>
<td>C3–7 pst fusion</td>
<td>Unknown</td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>73</td>
<td>Neck pain, hand dysfunction</td>
<td>Multiple cervical levels of listhesis</td>
<td>C2–T2 pst fusion, anterior C3–T1 fusion</td>
<td>Unknown</td>
</tr>
<tr>
<td>Carlson et al., 200725</td>
<td>F</td>
<td>39</td>
<td>Bilat upper extremity weakness, quadripareis, monoplegia</td>
<td>Rt C4–5 facet joint calcinosis involving spinal canal causing canal narrowing</td>
<td>C4–5 corpectomies</td>
<td>Improved bilat upper extremity strength</td>
</tr>
<tr>
<td>*Tuy et al., 200826</td>
<td>F</td>
<td>50s</td>
<td>Neck pain</td>
<td>Rt C2–3 pst element calcinosis mass w/ encroachment into spinal canal</td>
<td>Removal of lesion</td>
<td>Symptom-free</td>
</tr>
<tr>
<td>Shoji et al., 201127</td>
<td>F</td>
<td>34</td>
<td>Occipital headache</td>
<td>Calcinosis around odontoid process, dislocation odontoid process due to basilar invagination w/ compression of medulla</td>
<td>Halo, occipitocervical pst fusion &amp; instrumentation</td>
<td>Resolution of headaches, symptom-free</td>
</tr>
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<tr>
<th>Authors &amp; Year</th>
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<tbody>
<tr>
<td>Lebl et al., 2013</td>
<td>F</td>
<td>63</td>
<td>Neck pain, limited neck range of motion</td>
<td>Bilat multiple cervical facet joint calcinosis (unknown levels)</td>
<td>None</td>
<td>NA</td>
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<tr>
<td>Al-Khudairi et al., 2015</td>
<td>F</td>
<td>62</td>
<td>Bilat arm &amp; leg weakness &amp; numbness, myelopathy</td>
<td>C3 on C4 anterolisthesis, Lt C3 lateral mass destruction w/ calcinosis, Lt C4 lateral mass destruction, narrowing of C2 foramen transversarium, perched rt C3 on C4 facet</td>
<td>C3–4 laminectomies, reduction of C3–4 listhesis, C2–6 pst spine fusion &amp; instrumentation</td>
<td>Residual paresthesias of rt hand &amp; elbow</td>
</tr>
<tr>
<td>Sambataro et al., 2015</td>
<td>M</td>
<td>62</td>
<td>Neck pain</td>
<td>Lt C4–5, C5–6 facet &amp; lateral mass calcinosis, paraspinous region calcinosis</td>
<td>“Surgical excision of the mass”</td>
<td>Unknown</td>
</tr>
<tr>
<td>Nakamura et al., 2016</td>
<td>F</td>
<td>74</td>
<td>Neck pain, bilat upper extremity numbness</td>
<td>Calcinosis of bilat C1–2 facet joints</td>
<td>“Debulking of tissue,” reconstruction of C1–2 area</td>
<td>Resolution of neck pain &amp; arm numbness, symptom-free</td>
</tr>
<tr>
<td>Nguyen et al., 2016</td>
<td>F</td>
<td>57</td>
<td>Bilat hands, arms &amp; legs numbness &amp; weakness, gait imbalance, falls, myelopathy, incontinence</td>
<td>Calcinosis of pst elements C4/5 Subluxation C4 on C5, spinal cord compression due to dorsal epidural calcified mass C4–5, spinal cord edema C3–6, rt C2–3 facet joint calcinosis, bilat C4–5 facet joint calcinosis</td>
<td>C4–7 laminectomies, C3–T1 pst spine fusion &amp; instrumentation</td>
<td>Improved strength, improved bowel/bladder function</td>
</tr>
<tr>
<td>Tan et al., 2016</td>
<td>F</td>
<td>79</td>
<td>Shortness of breath</td>
<td>C3–5 paraspinous calcinosis masses, no spinal cord compression, L3–5 paraspinous masses</td>
<td>None</td>
<td>NA</td>
</tr>
<tr>
<td>Motegi et al., 2017</td>
<td>F</td>
<td>54</td>
<td>Neck pain, bilat arm weakness &amp; numbness</td>
<td>C4–5 dorsal epidural tumoral calcinosis, bilat C3–4 facet calcinosis</td>
<td>Resection of calcification</td>
<td>Symptom-free</td>
</tr>
<tr>
<td>Logothetis et al., 2018</td>
<td>M</td>
<td>67</td>
<td>Neck pain, palpable neck mass</td>
<td>Rt C3–4 facet joint calcinosis mass, no spinal cord compression</td>
<td>None</td>
<td>NA</td>
</tr>
</tbody>
</table>
Similar to the majority of reported cases associated with systemic sclerosis in the literature, our patient was female and required surgical intervention. She was treated in accordance with malignancy guidelines of surgical decompression and fusion. She required posterior cervical decompression and fusion to address the compressive and destabilizing pathology of the spinal canal and causing severe cord compression. This was one of the most severe cases of such an entity presented in the literature (Table 1). Our case is unique, however, in that this patient presented with an extensive and massive lesion invading the spinal canal and causing severe cord compression. This one of the most severe cases of such an entity presented in the literature. Thereby illustrating that calcinosis of scleroderma can present in the most severe cases of such an entity presented in the literature to date. Thereby illustrating that calcinosis of scleroderma can present complex clinical management questions and may ultimately require surgical intervention in which the surgeon employs practices and principles from all 3 categories of spine pathology.

References


Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Cage, Pham, Lee. Acquisition of data: Cage, Pham, Lee. Analysis and interpretation of data: Cage, Pham. Review of the literature: Cage, Pham, Singh, Lee. Drafting the article: Cage, Pham. Critically revising the article: Cage, Pham, Singh, Lee. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Cage. Statistical analysis: Cage, Pham. Administrative/technical/material support: Pham, Miao. Study supervision: Cage, Pham.

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