Idiopathic tumoral calcinosis–like lesion in the lower cervical spine causing acute central cord syndrome: case report

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A 57-year-old male presented with recurrent falls, bilateral lower-limb paresthesia, and severe neck pain. Imaging revealed a mass compressing his spinal cord. He was admitted for further workup for spinal cord compression. Within 24 hours of admission, he developed upper-extremity weakness while maintaining lower-extremity function. He underwent urgent decompression of his spinal cord. During exposure, a white, creamy odorless substance was noted. This same substance was found under pressure within the spinal canal. The mass was grossly removed, and the patient's weakness improved postoperatively. Based on the clinical picture, intraoperative presentation, and final histological examination, idiopathic tumoral calcinosis-like lesion was considered as the most appropriate diagnosis.

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KEY WORDS tumoral calcinosis; TC-like lesion; calcinosis; oncology

TUMORAL calcinosis (TC) is a rare hereditary, non-neoplastic condition that commonly presents with painless, focal, and/or multifocal periarticular soft tissue calcification.¹⁵ Given the rarity of TC, much of its pathophysiology is still obscured.¹⁵ Nonetheless, it is well documented in the literature that TC presents as a soft-tissue lesion with well-demarcated lobulated calcifications commonly located around the extensor surface of large joints.¹⁵

However, a number of cases present with TC-like lesions but do not fit the rest of the TC clinical picture.⁹ Unfortunately, these lesions have been inaccurately described as TC lesions, causing confusion and uncertainty among many clinicians. A TC-like lesion of the cervical spine in adults is extremely rare, and only 10 cases have been reported in the literature.^{1,2,6,7,10–12,16,18–20} We describe the unique presentation of a TC-like lesion localized in the cervical spine causing progressive acute central cord syndrome.

Case Report

History and Examination

A 57-year-old male presented to the emergency department with recurrent falls. Three days earlier, he had fallen while walking on the street. There was no loss of consciousness or head trauma, and no serious injury was sustained. Over the next 2 days while at home, he fell twice while trying to stand up from a sitting position. At that time, he was also experiencing bilateral paresthesia from the waist, down the legs as well as noticeable bilateral leg weakness that caused him to fall when he stood up. He also complained of weakness in the upper extremities more on the right side than the left and localized pain at the back of the neck. His symptoms had gradually progressed over the last 3 days, and on presentation they were constant.

His medical history included seizures, hepatitis C, cirrhosis, idiopathic thrombocytopenic purpura, anxiety, essential hypertension, and total knee replacement due to

ABBREVIATIONS MRC = Medical Research Council; TC = tumoral calcinosis.

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degenerative joint disease. He had undergone an arthrodesis of C1–2 with associated decompression of the greater occipital nerve 5 years earlier. The patient, an ex-smoker, also had a history of alcohol abuse. There was no significant family history to note.

On initial physical examination, the patient exhibited a broad-based gait and an inability to walk steadily. Romberg test was negative, and both the muscle power of the upper and lower extremities were Medical Research Council (MRC) Grade 5 out of 5. The rest of the physical examination was unremarkable.

Within 24 hours of admission, the patient's condition deteriorated, and he developed upper-extremity weakness greater on the right side while maintaining lower-extremity function. He was unable to extend his right fingers (0/5), his wrist flexion was 2/5, wrist extension was 4/5, triceps was 4/5, right grip was 2/5, and shoulder abduction and elbow flexion were 4/5.

Initial studies including blood tests, electrocardiography, and head CT were all unremarkable. Magnetic resonance imaging of the cervical spine showed a posterior epidural mass extending from C-6 to T-1 and causing severe spinal canal stenosis and cord compression (Fig. 1). Computed tomography showed calcification within the mass. The patient was admitted for further investigations. Given the patient's history, physical examination, laboratory test results, and imaging studies, the differential diagnosis included primary malignancy (for example, osteosarcoma), metastasis, TC, dystrophic calcinosis, and old calcified hematoma. Because of the patient's progressive weakness, he underwent emergency posterior decompression of his cervical spine.

Operation

He was placed prone, and a posterior cervical thoracic incision was made, exposing the posterior elements. The fascia was opened parallel to the skin incision, and a thick, milky odorless substance was encountered. Gram stain was negative. Histological examination of the encountered substance showed fragments of a fibrous-walled pseudocyst containing abundant, amorphous, finely granular calcification consistent with TC of the spine (Fig. 2). A hemilaminectomy on the left side was performed using a high-speed diamond-tipped matchstick bur; the laminae of C-6, C-7, and T-1 were burred away. During the burring, more of the thick milky substance was encountered and was evacuated. In addition, a pasty white substance along the epidural space was found and also removed. Further dissection toward the dura mater was performed. A pseudomembrane around the dura was encountered and removed using Stevens scissors. Intraoperative neuromonitoring improved with regard to lower-extremity function on the right side. Following decompression of the spinal cord at C-6, C-7, and T-1, the wound was irrigated and closed in layers over a drain.

Postoperative Course

The patient's postoperative course was uneventful. He underwent physical therapy to improve his muscular strength. One month after surgery, muscular strength in his extremities returned to normal (MRC Grade 5/5). Neurological sensations in both upper and lower extremities were intact. There was still residual tenderness along the upper thoracic spine and lower third cervical spine with prominent spinous processes. One-month postoperative

FIG. 1. Preoperative axial (A) and sagittal (B) cervical spine CT scans showing a calcified soft-tissue lesion in the posterior and posterolateral spinal canal, extending from C-6 to T-1 and narrowing the spinal canal and compressing the spinal cord. Sagittal T2-weighted MR image (C) showing a posterior epidural mass extending from C-6 to T-1 and causing severe spinal canal stenosis and cord compression. Axial (D) and sagittal (E) CT scans obtained 1 year after resection of the calcified tissue lesion, showing posterior decompression.

FIG. 2. Fragments of a fibrous-walled pseudocyst containing abundant, amorphous, finely granular calcification, resembling TC histological features. H & E, original magnification $\times 40$ (left) and $\times 200$ (right). Figure is available in color online only.

CT of the spine showed a residual calcinosis at C5–6, which was aspirated under local anesthesia. The patient made a full recovery after 6 months from the date of surgery, and 1-year postoperative CT scanning (Fig. 1D–E) showed resolution of the calcified mass.

The patient underwent further laboratory testing to determine the cause of his spinal calcinosis lesion (Table 1). His calcium and phosphate levels were within the normal range. His renal function was normal. The parathyroid hormone, 1,25-dihydroxyvitamin D, and fibroblast growth factor were also normal. Connective tissue diseases autoantibodies, including anti-nuclear antibody, anti-double stranded DNA, anti-RO, anti-LA, anti-SM, and anti-RNP, were all negative. Considering the clinical presentation, imaging results, histological studies, and laboratory findings, idiopathic TC-like lesion was considered the most appropriate diagnosis.

Discussion

The term “tumoral calcinosis” was introduced in 1943 by Inclan et al., who described 3 patients with large juxta-articular lobular calcified masses without visceral or skin calcifications.⁴ They differentiated TC from the dystrophic and metabolic calcifications by characterizing its metabolic features. Since then, many studies have reported similar lesions that met the description by Inclan et al.^{3,8} In the mid-1960s, literature reviews established that TC mainly presents in the 1st and 2nd decades of life, commonly affecting males of African descent. Patients frequently have a positive family history and multiple TCs affecting multiple joints, with a high recurrence rate after excision. Despite this clear characterization of the disease, its name has been broadly and imprecisely used to describe many clinical and/or pathological features similar to those of TC.

In 1996, Smack et al. reviewed 122 TC cases and proposed a new classification system that divided TCs into 2 types: primary and secondary.¹⁷ Two subtypes of the primary type exist, a hyperphosphatemic and a normophosphatemic primary TC. The secondary TC is associated with underlying pathology such as chronic renal failure.

Later, Olsen and Chew found that this classification lacks practicality from the clinician’s point of view¹⁵ since radiologists are the first to report the character of this lesion and since serum phosphate levels are rarely measured at the time of imaging. In addition, a normal serum phosphate level should raise suspicion that there is probably an underlying connective tissue disease causing a dystrophic lesion. Therefore, Olsen and Chew proposed using the term “tumoral calcinosis” strictly in reference to a disease caused by a hereditary metabolic dysfunction of phosphate regulation associated with massive periarticular calcinosis.

However, a significant number of cases resemble TC histologically and/or radiologically but do not fit the rest

TABLE 1. Summary of the main laboratory tests performed to exclude any potential underlying causes of the cervical TC

Test	Value
Serum electrolytes	
Serum calcium level	9.1 mg/dl
Corrected serum calcium level	3.4 mg/dl
Serum phosphate level	2.7 mg/dl
Renal	
Serum creatinine level	0.75 mg/dl
Estimated glomerular filtration rate (EGFR)	>60 ml/min/1.73 m ²
Endocrinological	
Parathyroid hormone (PTH)	17 pg/ml
Vitamin D (VITD-T25OH)	21 ng/ml
Fibroblast growth factor 23	122 RU/ml*
Rheumatological	
Anti-nuclear antibody (ANA) titre	Positive at 1:640
Anti-double stranded DNA (anti-dsDNA)	Negative
Anti-RO	Negative
Anti-LA	Negative
Anti-SM	Negative
Anti-RNP	Negative

* Reference range ≤ 180 RU/ml.

TABLE 2. Summary of case reports mentioning TC of the cervical spine with neurological symptoms*

Authors & Year	Demographics & Comorbidities	Neurological Symptoms	Imaging†	Surgical Management	Follow-Up
Carlson et al., 2007	Female, 39 yrs; diabetes, renal disease, Raynaud disease, systemic sclerosis, gout in toe	Chronic progressive weakness of bilat upper extremities, 5-day history of progressive quadriparesis & monoplegia	Overgrowth of exophytic dense calcification extending anteriorly & posteromedially from region of rt C4–5 facet joint w/ severe canal narrowing	Ant piecemeal extraction from dura w/ C4–5 corpectomies	2 mos: MRC Grade 3+/5 rt upper extremity, 4+ on lt; good integrity of fusion construct
Jackson et al., 2007	Female, 29 yrs; renal failure, hyperparathyroidism	2-day history of worsening upper-back pain & lt hand tingling	Large lesion of cervicothoracic spine, involving body & rt pedicle of L-4	Decompression & stabilization from C-7 to T-2, w/ pst instrumentation from C-5 to T-4	Neurological symptoms resolved w/in 2 wks, fusion anteriorly w/ no recurrence of calcinosis
Matsukado et al., 2001	Female, 54 yrs; renal disease	2-wk history of progressive radiculomyelopathy, & weakness of upper extremities	Spinal epidural mass from C-2 to C-4 posterolaterally, extending to atlantoaxial joint	Removal of lesion via midline pst approach	Good recovery from spastic gait & weakness of upper extremities
Miyakoshi et al., 2007	Male, 54 yrs; NA	2-wk history of progressive bilat weakness of upper & lower extremities	Extradural pst mass lesion at C3–4 level w/ marked spinal cord compression	Spinal cord decompression (C-2 laminoplasty, C3–4 laminectomy, & resection of mass lesion)	Immediate improvement in strength, no signs of recurrence after 2 yrs
Wong et al., 2013	Female, 77 yrs; rheumatoid arthritis	2-wk history of progressive bilat hand numbness, bilat leg weakness	Anterolisthesis of C-4 on C-5 consistent w/ abnormal calcification affecting pst elements, including facet joints, of C-4 & C-5	Pst decompression followed by ant & pst fusion	4 yrs: stable neurological exam w/o recurrence of TC

ant = anterior; NA = not available; pst = posterior.

* PubMed search conducted on April 2, 2015.

† Magnetic resonance imaging, CT, or bone scan imaging.

of the TC clinical picture, thus posing a pertinent diagnostic dilemma to both pathologists and clinicians.⁹ Unlike conventional TC, these “TC-like” cases usually present in uncommon sites such as the spine or small joints of the hand and tend to be nonrecurrent solitary lesions in patients who present after the 2nd decade of life with no family history of TC. In the current report we described a rather exceptional presentation of a TC-like lesion in terms of its anatomical location, classification, and acute presentation.

Our initial working differential diagnosis, based on the imaging findings along with the clinical picture, consisted of heterotopic ossification versus TC-like lesion. The lack of a T2 hyperintense signal within the markedly flattened cord favored a chronic process, and there was no enhancement associated with this heterotopic ossification. Given the presence of the very similar abnormality around the right C5–6 facet joint well outside the spinal canal, less likely differential considerations included dystrophic calcification within an old epidural hemorrhage or calcified meningioma.

The intraoperative findings along with the histological studies were suggestive of a TC-like lesion. An effort was made to identify a plausible etiology behind our patient's presentation. After taking a detailed medical history, performing a thorough examination, and reviewing the patient's serum biochemical profile, we identified no potential etiology. The more disease-specific investiga-

tions such as vitamin D (VITD-T25OH), anti-nuclear antibody (ANA), anti-double stranded DNA (anti-dsDNA), anti-RO, anti-LA, anti-SM, and anti-RNP antibodies were all negative. Considering the late onset of our patient's TC-like lesion, its anatomical location, the lack of a family history and plausible underlying condition, we considered an idiopathic TC-like lesion as the appropriate diagnosis.

A literature search of PubMed revealed 13 comparable case reports on TC or TC-like lesion localized to the cervical spine.^{1,2,5–7,10–14,16,18–20} Of those, 3 were pediatric cases (Supplementary Table 1) and 10 were adult cases (Tables 2 and 3). Interestingly, one report of TC described a case almost identical to ours in a 54-year-old male with intraspinal TC of the cervical spine who had presented with subacute (2 weeks) progressive weakness.¹² However, our case is unique in that there was an acute (3 days) and severe neurological deterioration.

Conclusions

In summary, we described the case of a patient with acute presentation of an idiopathic TC-like lesion of the lower cervical spine with severe spinal cord compression. The cause of the lesion remained unclear. After surgical removal of the mass and multilevel laminectomies, the patient's neurological course notably improved and no recurrences were observed at the 1-year follow-up. Spinal decompression was necessary to prevent neurological worsening and was deemed successful.

TABLE 3. Summary of case reports mentioning TC of the cervical spine without neurological symptoms*

Authors & Year	Demographics & Comorbidities	Clinical Symptoms	Imaging†	Surgical Management	Follow-Up
Chang et al., 2013	Female, 44 yrs; uremic	Explosive headache, pst neck soreness, limited range of motion	Lesion over right C1–2 region, causing bony destruction w/ atlantoaxial subluxation	C-1 laminectomy w/ eradication of calcific lesion plus internal fixation (occipital-C3–5) & posterolat fusion	1 wk: relief of neck soreness & headache w/ normalization of blood pressure
Lebl & Girardi, 2013	Female, 63 yrs; scleroderma	Severe axial neck pain, limited range of motion	Extensive soft-tissue calcifications adjacent to cervical facet joints	NA	NA
Remy-Leroux et al., 2009‡	Male, 29 yrs; NA	Large cervical mass, multiple episodes of cervical trauma	Tumoral mass extending to cervical muscles & lower cervical spine (C-6, C-7, T-1), accompanied by C-6 osteolysis	Parathyroidectomy	Wks later: regression of TC, renal transplantation w/ no recurrence of tumoral calcification after 6 yrs
Teng et al., 2006	Female, 59 yrs; Raynaud disease, esophageal dysmotility, sclerodactyly, telangiectasis	Chronic pst neck pain, occasional mild radiculopathy, mild bilat weakness	Expansion & sclerosis of rt pedicles & spinous processes at C-4 & C-5 w/ pst heterogeneous mass	Wide resection of lesions, C4–5 laminectomy, facetectomy & partial resection of C-3 & C-6, fixation	1 yr: neck pain resolved, neurovascular status remained intact
Tuy et al., 2008	Female, early 50s; scleroderma, interstitial lung disease, renal failure	Neck pain	Expansile calcified mass at rt side pst elements of C2–3, w/ lat encroachment into spinal canal	Removal of lesion	10 mos: no neck pain, stiffness, or radicular symptoms; no recurrence of mass

* PubMed search conducted on April 2, 2015.

† Magnetic resonance imaging, CT, or bone scan imaging.

‡ Based on abstract information.

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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: Schwab, Al-Sukaini. Acquisition of data: Al-Sukaini, Paulino Pereira, Chebib, Bredella. Analysis and interpretation of data: Schwab, Al-Sukaini, Paulino Pereira, Yu. Draft-

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Supplemental Information

Online-Only Content

Supplemental material is available with the online version of the article.

Supplementary Table 1. <http://thejns.org/doi/suppl/10.3171/2016.6.SPINE151565>.

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