Navigation-assisted resection of tumoral calcinosis of the lumbosacral spine: illustrative case

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BACKGROUND Tumoral calcinosis is an uncommon disease resulting from dystrophic calcium phosphate crystal deposition, with only 7% of cases involving the spine, and it may diagnostically mimic neoplasms.

OBSERVATIONS In this case, a 54-year-old woman with history of systemic scleroderma presented with 10 months of progressive left lumbosacral pain. Imaging revealed an expansile, 4 × 7-cm, well-circumscribed mass in the lumbosacral spine with L5–S1 neuroforaminal compression. Because intractable pain and computed tomography (CT)-guided needle biopsy did not entirely rule out malignancy, operative management was pursued. The patient underwent L4–S2 laminectomies, left L5–S1 facetectomy, L5 and S1 pediculectomies, and en bloc resection, performed under stereotactic CT-guided intraoperative navigation. Subsequently, instrumented fusion was performed with L4 and L5 pedicle screws and S2 alar-iliac screws. Pathological examination was consistent with tumoral calcinosis, with multiple nodules of amorphous basophilic granular calcified material lined by histiocytes. There was no evidence of recurrence or neurological deficits at 5-month follow-up.

LESSONS Because spinal tumoral calcinosis may mimic neoplasms on imaging or gross intraoperative appearance, awareness of this clinical entity is essential for any spine surgeon. A review of all case reports of lumbosacral tumoral calcinosis (n = 14 from 1952 to 2016) was additionally performed. The case featured in this report presents the first known case of navigation-assisted resection of lumbosacral tumoral calcinosis.

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KEYWORDS spine; calcinosis; tumoral calcinosis; spine surgery; interoperative navigation; scleroderma

Tumoral calcinosis is an uncommon disease presenting as a tumor-like calcified mass in periarticular soft tissue that forms from dystrophic calcium phosphate crystal deposition. It predominantly occurs in the hips, elbows, and shoulders, with only 7% of cases occurring in the spine. Spinal tumoral calcinosis can present with pain or neurological deficits due to compressive radiculomyelopathy or may be discovered simply as a growing or painful paravertebral mass. These lesions can mimic neoplasms on imaging or even gross intraoperative appearance, presenting a diagnostic challenge. Although the lesion itself is benign, its resultant symptoms or an uncertainty of diagnosis can lead to operative management. Awareness of this clinical entity is thereby essential for any spine surgeon. Here, we present the case of a 54-year-old woman with lumbosacral tumoral calcinosis causing significant nerve root compression, managed via navigation-assisted en bloc resection and posterior lumbosacral fusion.

Illustrative Case

History

A 54-year-old woman with a history of systemic scleroderma presented with 10 months of progressive and intractable left lumbosacral pain with intermittent radiation down the left leg. The pain worsened with prolonged sitting or standing and improve with ambulation and exercise. Her motor and sensory examinations were normal. Given failure of conservative management, magnetic resonance imaging and computed tomography (CT) were obtained, demonstrating a large, expansile, 4 × 7-cm, well-circumscribed mass in the lumbosacral spine (Fig. 1). The mass was cauliflower-
like in architecture, appearing heterogenous with both calcified and soft tissue areas. It appeared to originate from the L5–S1 facet joint and S1 pedicle with extension through the left L5–S1 neural foramen, compressing the exiting L5 and traversing S1 nerve roots. It extended epidurally into the spinal canal and dorsally through the paraspinal soft tissue up to the fascia in an overall eggplant-like shape. A metastatic workup was pursued, with a CT of the chest, abdomen, and pelvis revealing no other masses. A CT from 4 years prior containing the lumbosacral region did not demonstrate this mass. Given the imaging characteristics and rapid growth, at this point a primary bone tumor, such as chordosarcoma, or soft tissue tumor, such as synovial sarcoma, was suspected. A CT-guided needle biopsy was performed per our institution’s spinal oncology protocol. The biopsy demonstrated no evidence of malignancy but revealed granulation tissue, inflammation, and calcified amorphous material, with no ultimate definitive diagnosis.

At this point, the case was presented at a multidisciplinary complex spine forum. Upon rereview of the CT-guided biopsy specimen, discussion with a sarcoma pathologist concluded that although no definite calcium pyrophosphate crystals were seen on biopsy, the lesion was likely tumoral calcinosis, especially in light of the patient’s history of scleroderma. Because of the patient’s continued intractable pain and the biopsy not entirely ruling out malignancy because of calcifications potentially representing reactive changes found adjacent to tumor, operative management was pursued. The patient consented to surgery, with the goals of foraminal decompression to treat pain and maximal safe resection of the mass to rule out malignancy.

**Operation**

The patient was taken to the operating room, intubated under general anesthesia, and positioned prone with Mayfield pin fixation. Circumferential excision of the mass was performed with the aid of stereotactic CT-guided intraoperative navigation (Fig. 2) and neuro-monitoring, including electromyography, motor evoked potentials, somatosensory evoked potentials, and sphincter signals.

The lesion was widely dissected up to the L3–4 facet, down to the sacroiliac joint and posterior iliac crest, laterally out to the dorsal transverse processes, and medially to the spinous processes. To access the intraspinal elements of the mass and decompress the neural foramen and cauda equina, L4–S2 laminectomies, left L5–S1 facetectomy, and L5–S1 pediculectomies were performed. A large lobular calcified mass was found to be compressing the exiting L5 nerve root at the L5–S1 foramen, consistent with the patient’s symptoms and imaging. The lesion was ultimately resected in two separate pieces: one dorsal to the L5 pedicle and the other below the aspect filling the L5–S1 foramen. Both L5 and S1 nerve roots were able to be completely visualized at the completion of resection. The postresection, preinstrumentation intraoperative CT scan is shown in Fig. 1D.

Given the extensive bony resection of biomechanical spinal elements, subsequent stabilization was required and achieved with instrumented fusion, including L4 and L5 pedicle screws and S2 alar-iliac screws. Given the radical soft tissue dissection involved in the case, closure was aided by the plastic surgery team.

Pathological examination of the surgical specimen was consistent with tumoral calcinosis without evidence of malignancy. The mass was composed of multiple nodules of amorphous basophilic granular calcified material lined by histiocytes and multinucleated giant cells (Fig. 3).

**Postoperative Follow-Up**

Postoperatively, the patient recovered well, with complete resolution of radiculopathy but some persistent axial back pain relieved by multimodal analgesics. She was discharged home with no neurological deficits. She presented again 2 weeks after surgery with positional headaches and high-volume, clear-colored output from her surgical drain, but a cerebrospinal fluid leak was ruled out by imaging, and symptoms resolved promptly following observation and drain removal. There was no evidence of recurrence or neurological deficits at 5-month follow-up (Fig. 4).

**Discussion**

**Observations**

While “tumoral calcinosis” was first termed by Inclan et al. in 1943,1 descriptions of this clinical entity date back to 1899.8 Only an estimated 7% of cases occur in the spine.1,2 Most spinal tumoral
Calcinosis involves the lumbar spine, but lesions in the cervical, thoracic, and sacral regions have also been described. In contrast to the commonly painless and asymptomatic presentation of its extraspinal counterparts, spinal tumoral calcinosis often presents with pain or neurological deficits.

Tumoral calcinosis can arise in patients with underlying genetic or metabolic disorders that result in hyperphosphatemia, uremia, scleroderma, rheumatoid arthritis, and seronegative spondyloarthropathy. Idiopathic cases have been reported as well as those arising secondary to surgery or trauma at the site of involvement. In the spine, posited mechanisms of calcium phosphate crystal deposition in the spine include origin from the articular capsule, disc or facet joint degeneration, and hemorrhage with foamy histiocytic aggregation. In the present case, the lesion appeared to be centered in the bony spine, within the L5–S2 neural foramen, possibly arising from the L5–S1 facet joint. The case discussed was also associated with scleroderma. Soft tissue calcinosis is estimated in 9% to 27% of patients with scleroderma disorders. Tumoral calcinosis in scleroderma may occur via osteolysis or chronic inflammation in the setting of tissue hypoxia.

As seen in Fig. 3, the characteristic pathology of tumoral calcinosis entails amorphous calcium hydroxyapatite crystals surrounded by a reactive process in response to this material, involving foreign body giant cells and a histiocytic reaction, occasionally with intermixed detritic bone. Earlier lesions may contain a large amount of eosinophilic material, but as they continue to calcify over time, the crystals can condense and the overall appearance may become more basophilic, with some cases containing psammomatous elements.

As in the present case, tumoral calcinosis can present a diagnostic challenge due to mimicking neoplasms on preoperative imaging or even gross intraoperative appearance. Moreover, given that similar appearing calcification can also be found adjacent to bony or soft tissue tumors, a CT-guided biopsy without evidence of malignancy may not sufficiently rule out the possibility of unsampled malignancy adjacent to these findings. The suspected tumoral calcinosis found in the patient’s initial biopsy could have represented reactive changes adjacent to an unsampled malignant tumor. Given the patient’s imaging findings and rapid lesion growth raising concern for malignancy, a more radical resection was undertaken.

Nonoperative modalities of management have included observation, due to reported cases of spontaneous resolution, or addressing an underlying medical condition promoting soft tissue calcification. Nevertheless, surgery is frequently indicated for spinal tumoral calcinosis due to progressive neurological symptoms. Such procedures aim for resection, decompression of neurovascular structures with laminectomy or facetectomy, and promotion of stability by fusion or arthrodesis. Case series imply that incomplete resection may be associated with recurrence risk, with recurrence rates for tumoral calcinosis up to 75% following subtotal resection. While the risk of recurrence is less understood for spinal tumoral calcinosis specifically, in one review of 41 cases, Kalani et al. estimated successful gross total resection in 92.7% of patients and no reported episodes of recurrence.

FIG. 2. Intraoperative photographs of tumor (A, gray outline) with lumbosacral spine anatomical onlay (B) with a green pointer corresponding to the intraoperative neuronavigation seen in panel D. Screenshots (C and D) demonstrate the use of intraoperative neuronavigation. (Z. L. Gokaslan and P. Z. Sullivan retain copyright.)
Following a review of the literature, we report on the characteristics of 14 case reports of lumbosacral tumoral calcinosis from 1952 to 2016 (Table 1). 42.9% (6/14) cases were idiopathic and the most commonly known cause was scleroderma disease (35.7%). Eleven cases (78.6%) received surgical management, with fusion reported in only 2 cases (14.3%). 13,14 No cases of recurrence were documented. The present case is only the third reported case to describe the use of fusion to promote biomechanical stability after lumbosacral tumoral calcinosis resection. Moreover, CT-guided navigation of the present case is a salient example of the use of intraoperative navigation-assisted resection of lumbosacral tumoral calcinosis.

Lessons
Spinal tumoral calcinosis is a rare disease that can masquerade as a malignancy and may require surgical treatment because of neurological symptoms and the need for a definitive diagnosis. Although cases may be idiopathic, potential etiologies include hyperphosphatemia, uremia, scleroderma, rheumatoid arthritis, and seronegative spondyloarthropathy. An illustrative case of lumbosacral tumoral calcinosis associated with scleroderma and causing radiculopathy is presented, in which stereotactic-guided surgery led to safe gross total resection and symptomatic resolution. This report presents the first known case of navigation-assisted resection of lumbosacral tumoral calcinosis.

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## TABLE 1. Summary of case reports of lumbosacral spinal tumoral calcinosis

| Authors & Year | Age, Sex | Location | Etiology | Presenting Symptoms | Resection of Mass | Treatment | Recurrence |
|---------------|----------|----------|----------|--------------------|-------------------|-----------|------------|
| Blay et al., 2001 | 44, F | L5 | Inherited metabolic disorder | Low back pain | No | Conservative treatment w/ analgesics | NA |
| Cho et al., 2007 | 37, F | L3-4 | Idiopathic | Tender paravertebral mass | Yes | En bloc resection | None (at 12 mos) |
| Durant & Farge-Bancel, 2011 | 62, M | L4–S1 | Sclerodermal disease | Incidentally found | No | Not reported | NA |
| Ebot & Nottmeier, 2019 | 51, F | L4–5 | Previous op at site | Tender sacroiliac joint, pain w/ weight bearing | Yes | L4–5 HL & L4–5 transforminal lumbar interbody fusion | None (at 3 mos) |
| Emon et al., 2011 | 70, F | L5–S1 | Seronegative spondyloarthropathy | S1 hypoesthesia, hypoactive Achilles reflex, positive straight-leg raise | Yes | L5–S1 HL | NR |
| Iglesias et al., 2002 | 55, M | L5–S1 | Idiopathic | Numbness & weakness of L leg, hypoactive Achilles reflex | Yes | L5–S1 laminectomy | NR |
| Liberato et al., 2016 | 47, F | L5–S1 | Sclerodermal disease | Low back pain, L5 radiculopathy | No | Conservative treatment w/ analgesics & steroids | NA |
| Riemenschneider & Ecker, 1952 | 59, F | L5 | Idiopathic | Low back pain, hypoalgesia of posterior thighs | Yes | L5 HL | None (at 5 mos) |
| Sharma et al., 2006 | 55, M | L3 | Idiopathic | Low back & rt leg pain, areflexia in bilat LE | Yes | L3 laminectomy | NR |
| Shibuya et al., 2006 | 49, F | L3–4 | Sclerodermal disease | Low back pain, bilat LE weakness, gait disturbance, L3–4 spondylolisthesis | Yes | En bloc resection & posterolateral fusion (levels not reported) | None (at 22 mos) |
| Vaicys et al., 1999 | 49, M | L3 | Idiopathic | Growing paravertebral mass | Yes | Not reported | NR |
| Ward et al., 1997 | 53, M | L3–S1 | Sclerodermal disease | Lt leg pain & weakness | Yes | L4–5 laminotomy & discectomy, L5 HL, & S1 laminotomy | NR |
| Watanabe et al., 2000 | 55, M | L4–5 | Idiopathic | Gait disturbance | Yes | L5 laminectomy & duraplasty | None (at 3 yrs) |
| Weerakoon et al., 2014 | 60, F | L4–5 | Sclerodermal disease | Low back & lt leg pain | Yes | En bloc resection | None (at 12 mos) |

**HL** = hemilaminectomy; **lami** = laminectomy; **LE** = lower extremity; **NA** = not applicable; **NR** = not reported.

Summary of 14 earlier case reports of lumbosacral spinal tumoral calcinosis published from 1952 to 2016. There were no operative case series found on literature review. The search strategy encompassed a search of (“spine” OR “spinal” OR “vertebral”) AND (“tumoral calcinosis” OR “tumor calcinosis”) on PubMed on December 31, 2021. All articles discussing lumbosacral tumoral calcinosis were included for summary. The references list for all included articles was also manually reviewed for additional articles that warranted inclusion.
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Disclosures
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