**Uremic tumoral calcinosis in the cervical spine: case report**

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Tumoral calcinosis is an uncommon condition characterized by the calcification of periarticular soft tissue. In uremic patients, the disease is secondary to metabolic disturbances in predisposed patients. The authors report the case of a 73-year-old woman who presented with a new painful cervical mass while undergoing continuous ambulatory peritoneal dialysis for long-standing end-stage renal disease (ESRD). A CT scan of the neck showed a lobulated, calcified mass in the left paraspinal soft tissue at C2–3. This mass affected the facet joint and also extended into the neural foramen but did not cause any neurological compromise. Due to the patient’s significant medical comorbidities, resection was deferred and the patient was followed in the clinic. Subsequent repeat imaging has shown a significant decrease in the size of the mass. In the context of ESRD, a diagnosis of uremic tumoral calcinosis (UTC) was made. The authors conducted a search of the PubMed and EMBASE databases and identified 7 previously reported cases of UTC of the cervical spine. They present a summary of these cases and discuss the etiology, diagnosis, and management of the condition. Although the metabolic disturbances seen in patients undergoing dialysis can lead to tumoral calcinosis, most reported cases involve large joints such as the shoulder or the hip; however, the spine can also be affected and should be considered in the differential diagnosis of patients with uremia as it can mimic aggressive bone-forming neoplasms.

UTC often presents as painful tumor-like masses around both large weight-bearing joints and smaller overused joints. The bony hard masses are most commonly extra-articular and rarely erode bone, however, they may extrude through the skin, creating an infection risk. While the etiology of UTC remains poorly understood, a consistent prerequisite is an elevated serum calcium-phosphate product (Ca × P)^4^.

Currently, treating UTC in patients undergoing peritoneal dialysis remains challenging. Aggressive medical management alone does not always control tumoral calcinosis, and several studies have suggested adding intermittent hemodialysis as part of the treatment strategy to better...

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Cervical tumoral calcinosis

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manage electrolyte abnormalities. Other medical interventions include dietary phosphorus restriction, oral phosphate chelation, utilizing low-calcium dialysate solutions, and calcimimetics. Surgical interventions including local mass resection and parathyroidectomy have also been suggested, but neither strategy is curative in all patients. To date, the only curative option remains kidney transplant.

Most cases involve the shoulder, hip, and metatarsophalangeal joints; spinal cases are, overall, quite rare. We report on a 73-year-old woman who was undergoing peritoneal dialysis and presented to the neurosurgical service with a neoplastic-appearing UTC lesion of the cervical spine.

Case Report

This 73-year-old woman had hypertension, obstructive sleep apnea, hypothyroidism, and a renal transplant for ESRD secondary to polycystic kidney disease. One year prior to presentation her transplant failed and she had to resume dialysis. She subsequently had a myocardial infarction and developed congestive heart failure. She underwent intermittent hemodialysis for 4 months and then transitioned to peritoneal dialysis. Two months after beginning peritoneal dialysis (referred to as the presentation date hereafter), the patient presented with a growing painful mass in the left posterolateral aspect of her neck.

An ultrasound examination performed 1 month after presentation (Fig. 1) demonstrated a 2.1 × 1.4 × 2.6-cm hypoechoic, avascular lesion located 1 cm deep to the skin within the posterior triangle of the neck. A follow-up CT scan performed 3 months after presentation (Fig. 2A and D) revealed an enlarged 2.9 × 2.3 × 2.2-cm calcified, multilobular, and peripherally corticated mass in the paraspinal soft tissues adjacent to the left C2–3 facet joint. There was also extension into the left C2–3 intervertebral foramen. A bone scan (Fig. 3) showed increased uptake in the left paraspinal mass. The differential diagnosis included bone-forming neoplasms such as an osteochondroma, osteosarcoma, or chondrosarcoma. Blood tests at presentation showed various serum chemistry abnormalities, including elevated levels of urea (23.0 mmol/L, reference range 3.0–7.0 mmol/L), phosphate (2.67 mmol/L, reference range 0.80–1.35 mmol/L), and calcium-phosphate product (6.35 mmol²/L², reference range 1.68–3.51 mmol²/L²).

Throughout, the patient denied radicular pain and was neurologically intact on detailed examination. Resection of this growing lesion, for diagnosis and potential tumor
control, was deferred until her medical status could be optimized, given her significant comorbidities. In the interim, hemodialysis was initiated, and a follow-up CT scan was performed 10 months after presentation (Fig. 2B and E). This CT scan showed an unexpected decrease in the size of the lesion (to $2.4 \times 1.7 \times 1.8$ cm). Repeat blood tests confirmed ongoing uremia ($14.6$ mmol/L) but showed normalization of the calcium-phosphate product ($3.41$ mmol$^2$/L$^2$). Given this finding, the decision was made to continue monitoring the patient’s clinical condition. Her neck pain decreased over time. At the time of this writing, the patient continues to receive hemodialysis, her neck pain has resolved, and she has had no other symptoms related to the lesion. A CT scan performed at 17 months after presentation demonstrated continued decrease in the size of the lesion (Fig. 2C and F).

**Literature Review**

A thorough search of the PubMed and EMBASE databases was completed on May 24, 2015, and citations from previous case reports were also reviewed.\textsuperscript{12,19} The search was limited to native and translated English-language articles. A PRISMA flow diagram (Fig. 4) outlines the systematic review.

Thirty-three potentially relevant references were identified on the basis of the MeSH keywords (“tumoral calcinosis” or “dialysis related calcinosis”) and “cervical spine.” An additional 2 cases were added from citation...

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Discussion

Uremic tumoral calcinosis (UTC) is a rare phenomenon affecting 0.5%–3% of patients with renal failure.\textsuperscript{4,19,20} In this disease, calcium-phosphate salts precipitate in the periarticular soft tissues surrounding weight-bearing or overused joints including spinal facet joints. A review of the English literature yielded 7 cases of UTC affecting the cervical spine; in 6 of these cases the patients were undergoing hemodialysis, and in 1 case the patient was undergoing peritoneal dialysis. In this limited sample, disproportionately more female patients were affected (5 female, 2 male).

The etiology of this disease is not fully understood, but systemic factors, genetic factors, and local conditions have been implicated. The most important systemic factor is hyperphosphatemia, which was reported in nearly all patients regardless of serum calcium levels.\textsuperscript{4,19,20} Local conditions that have been implicated in the stimulation of calcium salt deposition include trauma, hematomas, and de novo overproduction of calcifiable matrix protein. Other possible predisposing factors are genetic mutations in the \textit{FGF23} and \textit{GALNT3} genes, leading to a decrease in the level of functional fibroblast growth factors, which is associated with hyperphosphatemic tumoral calcinosis.\textsuperscript{3,8,15}

In the spine, the differential diagnosis of a growing, calcified, paraspinal mass commonly includes sarcomas, primary osteochondral tumors, infection, and myositis ossificans. We stress the importance of including UTC in the differential diagnosis when managing a patient who may have an underlying metabolic disturbance.\textsuperscript{9}

The cornerstone of medical therapy is correcting the calcium-phosphate product of less than 4 mmol/L to minimize the risk of aberrant calcification (Level III evidence).\textsuperscript{7} To achieve this goal, many patients are treated with aggressive phosphate binders, calcimimetics, dietary phosphorus restriction, oral chelation, and increased length and frequency of hemodialysis treatments.\textsuperscript{19}

Surgical approaches include both parathyroidectomy and local resection of masses. Two studies have reported performing parathyroidectomy in patients with severe hyperparathyroidism and hypercalcemia refractory to medical treatment.\textsuperscript{4,19,20} Resection is generally undertaken for lesions causing acute, progressive, or refractory neurological dysfunction. Surgery is also indicated where severe pain and functional limitation are attributable to the lesion. In a recent report of UTC causing atlantoaxial subluxation and limited range of motion due to pain, Chang et al. describe lesion resection and fusion resulting in satisfactory pain relief and functional improvement.\textsuperscript{3} In the subset of patients with tumoral calcinosis and continued metabolic disturbances, a recurrence rate of 33% has been reported.\textsuperscript{10} Other studies have suggested that incomplete resection is associated with higher rates of recurrence.\textsuperscript{11,14,17}

Conclusions

UTC may involve the cervical spine, causing pain and radiologically mimicking an osteoblastic neoplasm. Our patient might have undergone a surgical intervention had her significant medical comorbidities not led to initial conservative management. As her metabolic abnormalities resolved, the calcified mass significantly decreased in size. Consistent with previous reports, control of serum phosphate and calcium-phosphate product levels remains the cornerstone of preventing UTC. The unique presentation in this report highlights the importance of including UTC in the differential diagnosis of bone-forming lesions in patients with ESRD.

<table>
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<tr>
<th>Authors &amp; Year</th>
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<th>Treatment</th>
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<td>Snitchler &amp; Silverman, 2011</td>
<td>55, M</td>
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<td>Not reported</td>
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<td>Sunder et al., 2013</td>
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<td>Increasing neck mass for 1 yr</td>
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<td>Carlson et al., 2007</td>
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<td>Progressive quadripareseis &amp; right UE monoplegia</td>
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<tr>
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<td>Increasing neck mass for 2 mos</td>
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<td>Local resection, phosphate binder, calcimimetic, Resolution of neurol Sx &amp; decreased lesion sizes at 1 yr</td>
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HD = hemodialysis; neurol = neurological; PD = peritoneal dialysis; pt = patient; PTX = parathyroidectomy; sig = significant; Sx = symptoms; UE = upper extremity.
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Disclosures
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Author Contributions
Conception and design: Ginsberg, Ahuja. Drafting the article: Fatehi. Critically revising the article: Ahuja, Wang. Reviewed submitted version of manuscript: all authors. Study supervision: Ginsberg.

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