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Case report

Tumoral calcinosis of the cervical spine in a dialysis patient. Case report and review of the literature

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ABSTRACT

The authors present a case of tumoral calcinosis (TC) in a patient with chronic renal insufficiency. The clinical course, imaging features and microscopic findings are detailed. A 60-year-old woman with a 4-year history of hemodialysis presented with a painful mass in the right posterior cervical triangle. The neuroimaging revealed polycystic mass bulging from the C3–C5 facet joints and lamina on the right. The majority of cystic mass was excised and microscopic features of the specimen were consistent with TC. Tumoral calcinosis is a rare disease characterized by calcium salt deposits in periarticular soft tissue, which enlarge to form tumor-like cystic masses containing chalky calcareous material. TC is typically seen around large joints but rarely in the spine. Review of past publications provided six cases of TC involving the spine in dialyzed patients.

1. Introduction

Pathology and clinical manifestation. Tumoral calcinosis (TC) is characterized by calcium salt deposits in periarticular soft-tissue. The lesions enlarge over time and form cystic tumors with fibrotic capsule containing chalky semiliquid material [1–11]. Microscopically, the content is composed of calcium salt deposits (mainly calcium hydroxyapatite), intermixed with epithelioid elements, histyocytes, lymphocytes, macrophages and multinucleated giant cells [1–7,10,12]. The development of tumors is asymptomatic until compression of the surrounding structures occurs and causes local pain, joint motion limitation or neurologic symptoms. In advanced stages the cysts may evacuate through fistulae draining white chalky fluid [8,13]. TC most commonly involves extensor surface of large joints, like hip, elbow, shoulder, foot and wrist [1,5,6,8].

Spinal involvement. Spinal location, which was first recognized by Riemenschneider and Ecker in 1952 [9] is considered to be very rare [3,5,13–33]. In 2011, Kalani et al. reviewed the literature bringing to light 41 individuals with TC of the spine

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reported by that time [19]. Additionally, six cases were reported between 2011 and 2015 [2,15,16,19,21,33] and prior Pakasa and Kalengayi (1997) mentioned four cases diagnosed by means of biopsies of lumbar spine [7]. The largest clinical series of 21 patients with spinal TC was published in 2001 by Durant et al., who concluded that histology of paraspinal lesions was identical to that of TC seen elsewhere in the body [3]. We report cervical spine TC in a patient undergoing long-term hemodialysis. To the best of the authors' knowledge only six cases of spinal TC in dialyzed patients were described apart from this given report [11,14,15,18,22,27]. The aim of our paper is to accumulate the knowledge regarding this rare disease of the spine.

2. Case report

History and examination. A 60-year-old woman was admitted in April 2014 for treatment of cervical spine tumor. Since 2009 she has been on hemodialysis for chronic renal failure. She reported a 4-month history of progressive right-sided neck pain radiating to the occiput, right ear, right shoulder and arm associated with numbness of the right hand; she did not recall any trauma. Examination revealed a palpable tender mass in the right posterior cervical triangle while cervical rightward rotation was limited to 45°. Neurologic examination revealed paresis of right deltoid and biceps muscles (3/5), decreased right deep tendon reflexes (biceps, brachioradialis and triceps) as well as hypoesthesia of fingers of the right hand.

Laboratory studies. Laboratory tests showed slightly elevated serum phosphorus of 4.98 mg/dl (normal: 2.50–4.50), whereas white blood cell count, serum calcium, parathyroid hormone and C-reactive protein were all within normal limits.

Imaging findings. Cervical computed tomography (CT) scans revealed soft tissue partially calcified masses related to the right facet joints and right laminae from C3 to C5, partially sclerotic C4 vertebral body as well as spondylotic changes

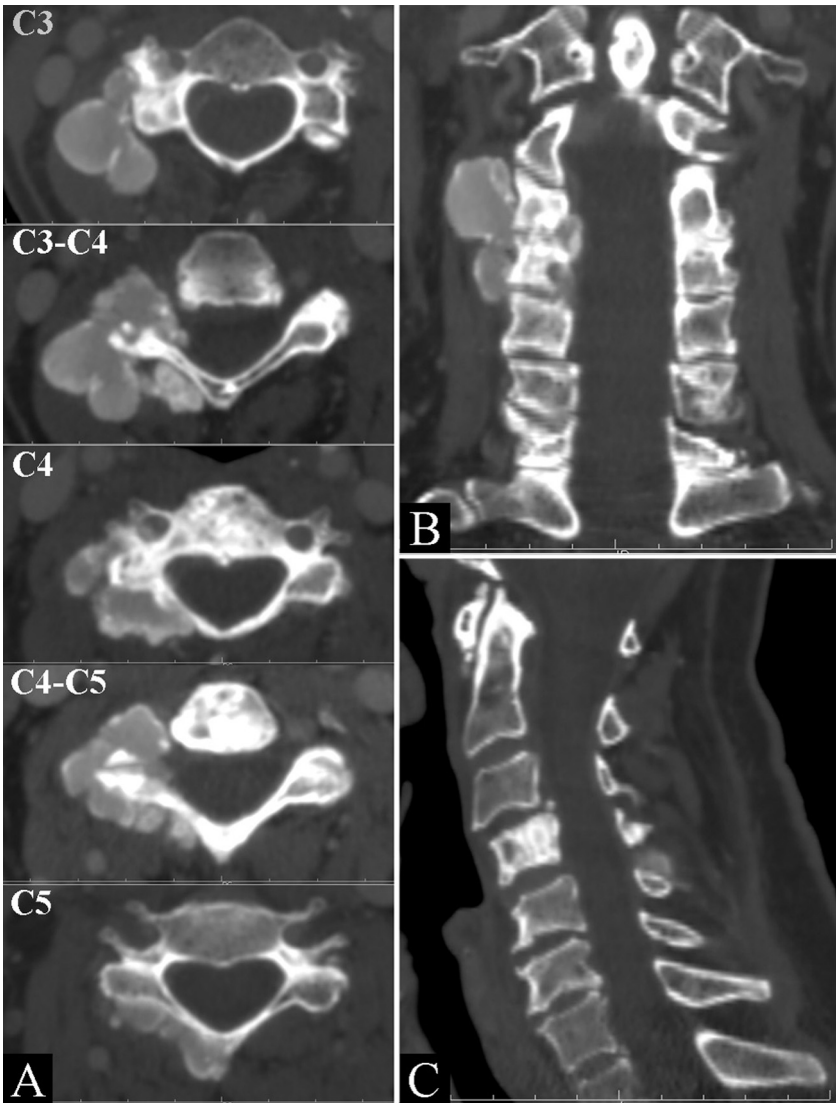

Fig. 1 – Axial CT scans (A) and coronal reconstruction (B) show lobulated partially calcified masses with sclerotic rim involving right facet joints and laminae of C3–C5 vertebrae. The masses narrow down the neural and transverse foramina also penetrate into the spinal canal at C3/C4 level. The sagittal reconstruction (C) show degenerative discs disease and anterior dislocation of C3; the C4 vertebral body appears sclerotic.

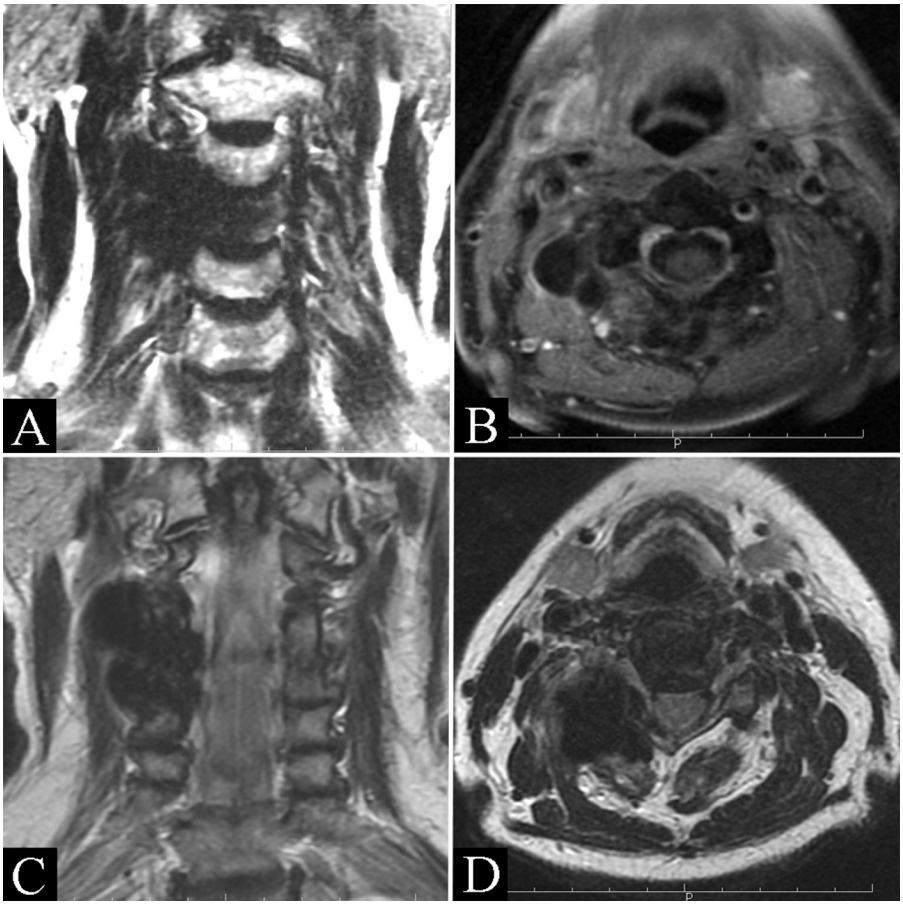

Fig. 2 – T1-weighted MR images with fat suppression after Gadolinium administration (A, B) and T2-weighted MR images (C, D) show a mass involving the right C3–C5 facet joints and the paraspinal soft tissue with slight penetration into the spinal canal. The lesion is hypointense in both T1- and T2-weighted images.

(Fig. 1). Magnetic resonance (MR) examination showed that the masses were hypointense in both T1 and T2 images; best seen on coronal views (Fig. 2). After Gadolinium administration, there was no contrast enhancement. Given the history and imaging studies, our colleague radiologist (B.S.) diagnosed uremic periarticular calcification. Surgical treatment was considered for this lesion.

Operation. The procedure was realized under general anesthesia. The patient was placed in a supine position with her head turned to the left. A longitudinal skin incision was made along the posterior border of the sternocleidomastoid muscle. This muscle was retracted medially and blunt dissection denudated a cystic tumor firmly attached to the right C3/C4 and C4/C5 facet joints. Some branches of cervical plexus running in the proximity of tumor capsule were identified and protected. The cyst contained white chalky fluid, which expressed from the incised capsule (Fig. 3A). As the fluid resembled pus, it was taken for the cultures and aspirated from the capsule. The tumoral mass was fully excised and the area of its implantation to the spine was curetted. Wound healing was uneventful.

Pathological findings. Microscopic examination found multiple calcifications surrounded by connective tissue with multinucleated giant cells and macrophages (Fig. 3B,C). The staining for amyloid and the cultures turned out negative. Histologic findings were in line with the diagnosis of TC.

Postoperative course. After surgery, the patient experienced complete pain relief and regression of numbness. The range of cervical motion slightly improved, muscular force of the right upper extremity increased to 4/5. She continued hemodialysis program complemented by low-kalium diet, no phosphate binders were prescribed. In June 2014, a follow-up CT scan confirmed absence of excised cyst, however, the right C3/C4 and C4/C5 facet joints were involved by soft tissue mass with minor paraspinal extension suggesting incomplete resection (Fig. 4A). As the lesion was asymptomatic, its surgical removal was not advised. In January 2015, the patient presented after a 3-month history, with nuchal pain. The CT scan revealed absence of previously documented pathologic mass, which apparently had regressed spontaneously (Fig. 4B). The facet joints from C3 to C5 on the right side were destroyed, additionally further progression of cervical kyphosis was observed (Fig. 4C). Given this finding, anterior cervical spine surgery was suggested to the patient. The procedure included anterior approach to C3–C6 spinal segment, discectomies and interbody fusions with placement of PEEK cages. The surgery

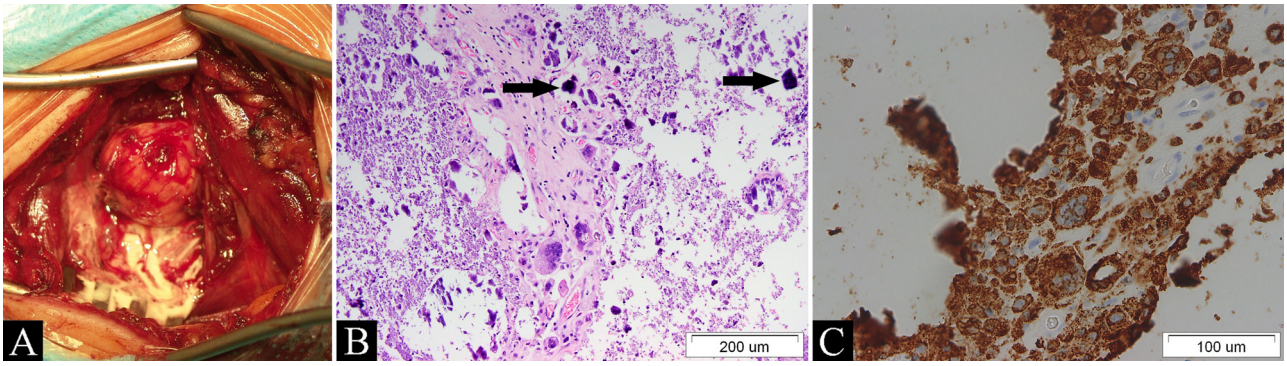

Fig. 3 – (A) Intraoperative photograph demonstrating the cystic tumor and exudation of a white chalky liquid from the incised capsule. (B) Calcifications (arrows) and multinucleated giant cells in the cyst wall (staining with hematoxylin and eosin, microphotographs x10). C: macrophages in the cyst wall (immunohistochemical staining for CD68 antigen, microphotographs ×20).

was completed by anterior cervical plating to maintain spinal lordosis and stability. The convalescence was uneventful with relief of nuchal pain; the patient is on the kidney transplant waiting list. For the purpose of this research paper we have obtained the patient's written consent to publish her case.

3. Discussion

Etiology, pathogenesis and classification. The first case reports on TC were published independently by two French authors: Giard and Duret in 1889 and 1890, respectively [1,4,6,12]. Inclan et al., in 1943, reported on three affected patients and first used the term *tumoral calcinosis*, which became widely acknowledged [34]. In 1966, Smack et al. distinguished three types of TC: (1) primary normophosphatemic (predominantly seen in young individuals living in the tropics), (2) primary hyperphosphatemic (with genetic predisposition, frequent in individuals of African descent) and (3) secondary, most commonly observed in dialyzed patients; also present in spectrum of rare diseases capable of causing soft tissue calcifications [35]. TC develops in 0.5–3% of chronically dialyzed patients with renal failure as a result of hyperparathyroidism [1,4,6,11,15,18,22,30–32]. The classification proposed by Smack et al. gained wide acceptance [2,4–6,10,16,17,23,26,28,30,33]. However, some authors point out that the term *tumoral calcinosis* signifies primary pathology [1,6,12,26], whereas lesions secondary to pre-existing disease should be named *tumoral calcinosis-like lesions* [18]. Irrespective of the cause, cystic masses in primary and secondary TC show no differences [1,3,4,6,8,10]. Some researchers explain the development of TC in the proximity of joints, based on mechanical conception. They argue that the periarticular forces and repetitive mechanical stimulation may initiate localized disturbance of the calcium metabolism, which in turn provoke calcium phosphate depositions in soft tissue [1,4,6,8,10,14,22,28].

Regarding the spinal TC, the causative role of facet joint degeneration, hypermobility and local inflammation was suggested [3,17,30]. In the majority of cases the calcified masses were observed around posterior vertebral elements [11–14,18,21,22,24,25,27,30,31,33] and in the spinal canal [2,11,14,15,17,19,21,22,24,25,27,28,36]. Rarely found inside the dura mater of the spine [5,23,29,32]. Clinical manifestation of spinal TC entailed: palpable tender paraspinal mass [18,20,26,31], axial pain in affected spinal segment [2,3,11,13,15,18–23,27,30], limited cervical or lumbar range of motion [9,11,13,15,20], tetraparesis [14,22,28,33], paraparesis [2,3,17,30,32,36], lower extremity spasticity [5] and radiculopathy in the distribution of the compressed nerve roots [3,9,13,16,18,29,30,36]. Spinal deformity, such as torticollis or rapidly progressing scoliosis, was the main symptom in case of children [19,24,25].

Imaging. Radiographs showed a paraspinal calcified mass [9,11,13,18,20,24–26,30,31], however, in a fraction of cases calcifications were not apparent on plain films [3,23,32,36]. The CT scans revealed paraspinal partially calcified multi-lobulated masses [11,13–15,18,21,24–27,30,31,33]. Some lobules demonstrated fluid-fluid levels [21,25,26]. The masses most frequently presented low signal intensity on both T1 and T2-weighted MR images [2,3,11,15,16,18–21,23,25,27,29,30]. Mixed signal intensity was also observed [3,5,13,17,22,28,32,36]. Gadolinium enhanced intratumoral septa or tumoral margins in a fraction of cases [5,17,20,23,28,30,32]. In majority of reports, the preoperative differential diagnosis based on radiologic images was uncertain; it included the gamut of pathologies capable of causing calcifications, such as: (1) degenerative/traumatic (calcifications or hypertrophy of the ligamentum flavum, calcification of the filum terminale, calcified hematoma, calcified soft-tissue mass, cartilaginous mass, disk hernia, exuberant callus, synovial cyst, facet fracture) [2,3,5,11,16,17,23,24,29,36]; (2) inflammatory (osteomyelitis, discitis, pannus, infection, abscess, myositis ossificans) [2,3,5,13,18,27]; and (3) primary or metastatic tumors [2,3,5,11,13,18,19,22,24,27,32,36].

Treatment. The literature review shows that the treatment of spinal TC was surgical. It comprised removal of the compressive mass followed by spinal stabilization, if necessary [2,3,5,6,9,13–20,22–33,36]. The final diagnosis of TC was made after microscopic examination of specimens obtained by biopsy or surgical resection [3,5,9,11,13,14,16–28,30–33,36].

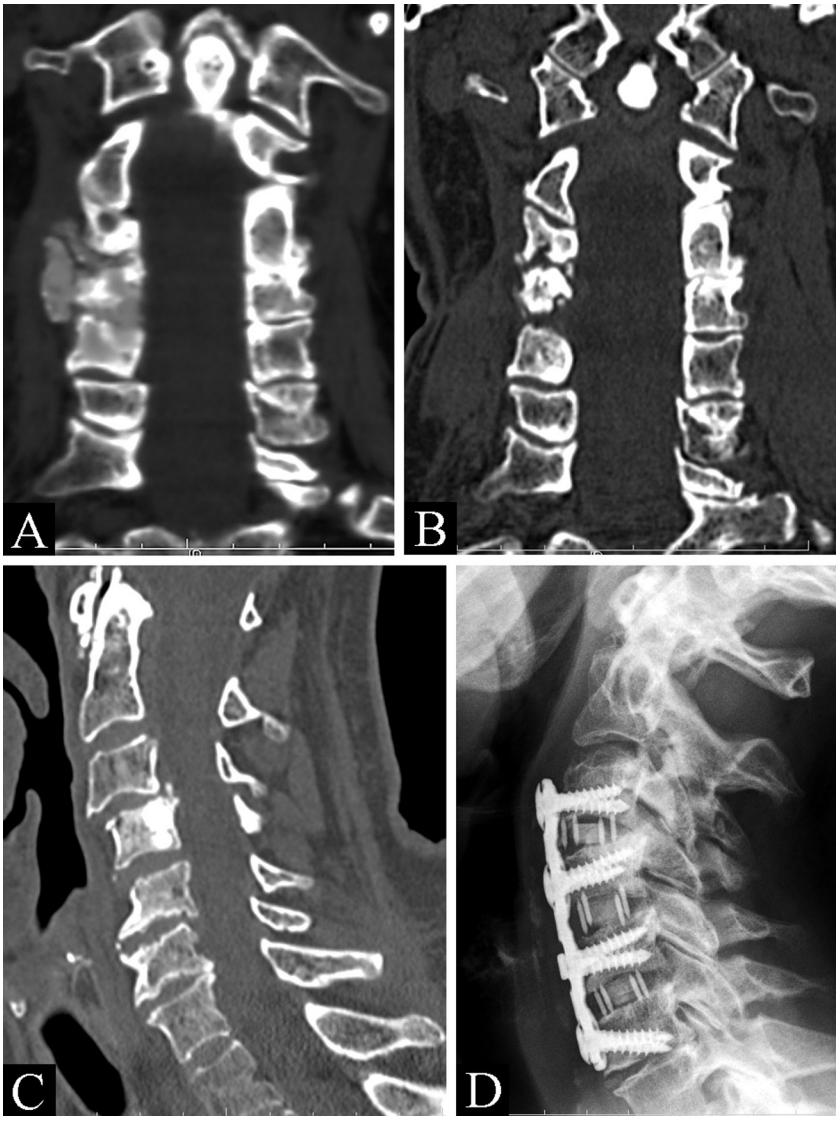

Fig. 4 – (A) Coronal reconstruction of CT images performed two months after the surgery shows no recurrence of the resected cyst; the right C3/C3 and C4/C5 facet joints are involved by amorphous soft tissue mass with a minor paraspinal extension. (B) Coronal reconstruction of CT images performed eight months after the surgery shows bony erosion of the right C3/C4 and C4/C5 facet joints without any neighbouring pathological mass. (C) Sagittal reconstruction of CT images performed eight months after the surgery shows degenerative discs disease, anterior dislocation of C3, sclerotic appearance of the C4 vertebral body and a kyphotic deformity of the cervical spine. (D) Lateral cervical roentgenogram performed three months after the surgery of C3–C6 segment shows reconstitution of cervical lordosis.

Comparison. The described case shows similarities with previously reported six cases of spinal TC in dialyzed patients [11,14,15,18,22,27]. All six patients were females aged 24–59. The pathological cystic masses were situated in the cervical spine near the facet joints invading the intervertebral foramina or narrowed the spinal canal. Pain in the involved spinal segment was a common symptom in all cases [11,14,15,18,22,27]. Laboratory tests showed elevated serum phosphorus in four patients [11,14,15,18] and low calcium in two [14,18]. All patients who underwent surgical treatment were healing afterwards uneventfully. In 5 cases the procedures comprised surgical excision of pathological masses [14,15,18,22,27]. One patient with the cyst involving C2–C3 facet joint was successfully treated by a small incision (open biopsy) during which the chalky gelatinous material was evacuated from the cyst [11]. In all cases except one, the cysts contained a white or yellowish fluid resembling pus [11,15,18,22,27].

Two unique aspects of the presented case need to be highlighted. Firstly, the anatomic features of spinal involvement and paraspinal masses were best seen on CT in comparison with MR images. The CT revealed bone erosions and destruction of facet joints. This finding differs from the majority of reports, which highlighted superiority of MR.

Secondly, the residual paravertebral calcifications documented two months after the surgery healed spontaneously over time. One can hypothesize that the aspiration of calcium salt deposits and the removal of the capsule ceased local irritation, which in turn promoted disappearance of residual masses.

4. Conclusions

To conclude, in the described case the degenerative changes of facet joints concurrent with discs degeneration and spinal instability might have triggered development of tumoral-calcinosis-like lesions. Here the surgical resection of the lesion and curettage led to healing.

Conflict of interest

None declared.

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Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

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