Tumoral calcinosis of the craniovertebral junction as a cause of dysphagia with treatment by transoral decompression: case report

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Tumoral calcinosis is characterized by tumor-like deposition of calcium in periarticular soft tissue. Spinal involvement is rare, and perioperative diagnosis of tumoral calcinosis can be difficult because lesions may be confused with bony neoplasms. Symptoms of tumoral calcinosis result from bony involvement and/or direct compression of surrounding anatomical structures, for which treatment with surgical decompression can be highly successful. The craniovertebral junction is rarely affected by tumoral calcinosis, and patients with this condition may present with distinct symptoms. Herein, to their knowledge the authors present the first case of tumoral calcinosis affecting the craniovertebral junction in a patient who presented with severe dysphagia and required transoral decompression. Recognition of tumoral calcinosis by neurosurgeons is essential for facilitating diagnosis and treatment, and the transoral approach is an effective method for decompression in select patients.

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KEY WORDS bony neoplasm; calcium deposition; craniovertebral junction; dysphagia; transoral decompression; tumoral calcinosis; cervical spine

TUMORAL calcinosis (TC) is a rare clinical syndrome caused by calcium deposition in periarticular soft tissue. Most commonly, TC affects the joints of the upper limbs and the hips; however, it is known to involve the articular surfaces of the cervical, thoracic, and lumbar spine.¹⁰ In cases of spinal involvement, direct compression of neural elements and/or surrounding anatomical structures causes clinical symptoms that can be treated with direct surgical decompression.^{1,3,4,8,15,16,20,23,25–28,30,32,33} Awareness of TC is essential for spine surgeons because surgical specimens can be misdiagnosed if a neoplasm is suspected as the primary diagnosis.⁸

Few authors have reported on the diagnosis and management of TC in the cervical spine,^{1,4,8,15,20,23,25,26,30} likely due to the rarity of this condition and the diagnostic dilemma that it presents for clinicians.^{8,30} Furthermore, patients with TC at the craniovertebral junction (CVJ) may present with unique symptomatology and require a different treatment paradigm. Herein, we report a unique case of TC affecting the CVJ in a patient who presented with severe dysphagia. The lesion was successfully treated by transoral decompression without fusion. To our knowledge, this is the first case of TC affecting the CVJ that has been treated

using the transoral approach. We review the clinical presentation, radiographic findings, and surgical management of TC in the CVJ of this patient.

Case Report

History and Examination

A 60-year-old woman with a history of hypertension presented to our clinic with progressive dysphagia and mild neck pain of 1 year's duration. The patient's symptoms had progressed to the point that she could tolerate only a liquid diet for the past 8 months. She denied any focal motor weakness, sensory symptoms, or gait disturbance. On physical examination, she exhibited full strength in all muscle groups, with no evidence of myelopathy; however, a large protrusion was visible on examination of the posterior pharynx. A preoperative swallow evaluation performed by speech therapy staff revealed oropharyngeal mass effect as the cause of the patient's dysphagia. A CT scan of the cervical spine revealed a calcified mass centered near the left C1–2 facet, causing compression of the pharynx ventrally (Fig. 1A–C). MRI demonstrated a T1 and T2 hypointense mass with minimal retropulsion

ABBREVIATIONS CVJ = craniovertebral junction; TC = tumoral calcinosis.

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FIG. 1. **A and B:** Axial CT scans demonstrating a calcified mass at C-1 (A) and C-2 (B) with compression of the oropharynx (arrows). **C:** Sagittal CT scan demonstrating ventral extension of the mass (arrow). **D and E:** Sagittal (D) and axial (E) T2-weighted MR images demonstrating the hypointense mass (asterisk) with minimal canal compromise or cord compression. **F:** Left shoulder radiograph demonstrating a separate calcified lesion adjacent to the acromion (dashed outline).

(Fig. 1D and E). A radiograph of the left shoulder showed a large calcified mass adjacent to the acromion (Fig. 1F).

Surgical Treatment

In light of the patient's severe, progressive dysphagia, surgical decompression via a transoral approach was offered as a treatment option. The patient was brought to the operating room and intubated under general anesthesia. Her head was placed in a Mayfield head holder, and the O-arm Surgical Imaging System (Medtronic plc) was used to register image guidance (Video 1).

VIDEO 1. Operative video demonstrating the transoral approach for decompression of tumoral calcinosis involving the craniovertebral junction. Narration by the senior author (N.T.). Copyright Barrow Neurological Institute, Phoenix, Arizona. Published with permission. [Click here to view.](#)

The Spetzler-Sonntag transoral retractor (Integra LifeSciences Corp.) was then placed (Fig. 2A), and the operative field was prepared. A posterior pharyngeal mass was readily identifiable. A midline incision was made, and white “chalky” fluid was easily evacuated (Fig. 2B). The resection was continued deep and laterally, with the aid of image guidance to decompress the region while sparing the anterior arch of C-1 and the left C1–2 facet (Fig. 2C–F). Pathological examination of an intraoperative frozen specimen indicated a benign process with masses of amorphous deposits of calcium hydroxyapatite surrounded by a histiocytic and giant cell reaction. After adequate decompression of the lesion, the oropharynx was closed in a single layer, and a feeding tube was passed beyond the oropharynx under direct visualization.

Postoperative Course

Upon awakening, the patient was neurologically stable. She remained intubated for 48 hours, as is routine for

transoral approaches at our institution, and she was subsequently extubated without difficulty. She was administered broad-spectrum antibiotics for 4 days to protect against a potential infection due to oral flora. The feeding tube was left in place over the 4 days to facilitate surgical wound healing and provide nutrition. The patient's diet was subsequently advanced under the guidance of speech therapy staff, and she was tolerating a soft diet on discharge from the hospital. The final pathology results revealed dystrophic calcification, consistent with TC. Postoperative dynamic radiographs demonstrated stability of the CVJ. The patient had no complications on discharge from the hospital. At 12 months after the operation, dynamic radiographs demonstrated stability of the CVJ, and CT of the cervical spine showed no recurrence of the lesion. At the 14-month clinical follow-up, the dysphagia had significantly improved, and the patient was tolerating a solid diet.

Discussion

Tumoral Calcinosis

The term “tumoral calcinosis” was first used in 1943 by Inclan et al.,¹⁴ but the condition was first described by Duret in 1899.⁹ It is a pathological condition defined by the presence of calcium hydroxyapatite crystals in periarticular soft tissue. Most patients have multiple lesions that remain asymptomatic until the persistent growth of the lesions leads to local mass effect, causing pain or compression of neurovascular structures.^{10,30} TC can be separated into 3 subtypes—familial, sporadic, and secondary to chronic renal disease²⁶—and 23% of cases are associated with a primary metabolic disorder such as hyperphosphatemia.^{5,31}

CT and MRI are complementary radiological studies for lesions in patients with TC. A CT study typically demonstrates an irregular, lobulated, isodense-to-hyperdense

FIG. 2. **A:** Preoperative positioning with the Spetzler-Sonntag retractor in place. **B:** Initial evacuation of the lesion upon incision of the oropharynx. **C:** Resection cavity with the anterior aspect of C1–2 (*asterisk*) in the deep portion of the operative field. **D–F:** Corresponding postoperative axial (D and E) and sagittal (F) CT scans demonstrating the extent of resection. Figure is available in color online only.

mass that may exhibit septations or fluid levels due to the layering of calcified fluid.^{2,13,17–19,29} MRI typically demonstrates intermediate- to low-signal intensity of T1/T2 sequences and little to no contrast enhancement, as was seen in our patient (Fig. 1).

Although TC of the spine can be confused with other more malignant pathologies,⁸ the primary differential diagnosis of systemic TC includes the crystal deposition arthropathies gout and pseudogout. The clinical history and histological diagnosis typically distinguish these entities, with gout demonstrating negatively birefringent crystals under polarizing light, pseudogout demonstrating positively birefringent crystals, and TC demonstrating non-birefringent crystals. Grossly, TC often has a chalky white texture associated with retained calcified fluid and a honeycomb appearance surrounding the lesion; both of these features were readily apparent in our patient (Fig. 2). TC is not usually associated with the adjacent bony destruction seen in patients with crystal pseudarthropathies.^{3,26} It has been noted that incomplete resection of TC in other locations can lead to recurrence and that continued clinical and radiographic follow-up for these patients is essential in the years following an operation.^{10,20}

TC of the Cervical Spine and the CVJ

Although TC typically affects the hip joints and the joints of the upper extremities, spinal involvement has been described.²⁶ Cases involving both the vertebral bodies and the posterior elements have been reported, with most cases involving the lumbar spine.^{3,8,28,32,33} Twelve cases have been reported that involved the cervical spine, and most of these patients presented with neck pain and/or symptoms of spinal cord compression (Table 1).^{1,4,8,15,}

^{20,23,25,26,30} Only 3 cases of TC involved C-1 and/or C-2; the remaining cases involved the middle to lower cervical spine. Most patients with cervical TC have been treated with posterior decompression with or without fusion; none have been treated with transoral decompression.

Of the 3 reported cases of TC affecting the C1–2 levels, only 1 patient had TC involving the anterior elements at these levels. Chang et al.⁴ reported the case of a 44-year-old woman who presented with posterior neck pain and headaches and was found to have a periodontoid mass causing C-2 nerve root and spinal cord compression. Although the TC in this case involved the odontoid and ventral components of C1–2, the lateral nature of the mass and the associated neural compression prompted a posterior decompression and occipital to C-5 fixation.

The other 2 cases of TC affecting C1–2 were also treated with posterolateral decompression, 1 with and 1 without fusion. Mooney and Glazier²³ reported the case of a 17-month-old boy who presented with torticollis and was found to have a calcified mass at the lateral aspect of C-1, where it was encroaching on the spinal canal. The lesion was resected via a far-lateral approach, followed by a partial C-1 laminectomy with no fusion performed. Durant et al.⁸ reported the case of a 78-year-old man who underwent posterior decompression and fusion for presumed rheumatoid pannus; however, the histopathology revealed TC. No other details regarding patient symptoms or imaging were provided in their report.

Of note, our literature review identified several cases of calcium deposition at the CVJ that were thought to represent TC but were not confirmed by histology.^{3,16,21} The report by Kokubun et al.¹⁶ has been cited as one of the original descriptions of TC in the cervical spine; however,

TABLE 1. Published reports of tumoral calcinosis of the cervical spine

Authors & Year	Pt Age (yrs)	Level	Position	Clinical Presentation	Treatment
Agarwal et al., 1993	45	C5–6	Posterior	Pain, weakness	Laminectomy
Ohashi et al., 1996	12	C3–6	Posterolateral	Torticollis	Surgical excision
Mooney & Glazier, 1997	1.5	C1–2	Lateral	Torticollis	Posterolateral decompression
Durant et al., 2001	78	C1–2	NS	NS	Posterior excision/fixation
	61	C7–T1	NS	NS	None
	48	C3–4	Posterolateral	NS	Posterior laminectomy & fusion
	70	C4–5	Posterior	NS	Laminectomy
Matsukado et al., 2001	54	C2–4	Posterior	Cervical pain, weakness	Posterior decompression/laminoplasty
Qadri et al., 2005	51	C7–T1	Posterior	Myelopathy	Laminectomy & excision
Teng et al., 2006	59	C3–6	Posterior	Neck pain/weakness	Laminectomy & fusion
Jackson et al., 2007	29	C6–T2	Posterior	Pain, paresthesia	Anterior plus posterior decompression & fusion
Chang et al., 2013	44	C1–2	Anterior	Headache	C-1 laminectomy, occiput–C5 fixation
Current report	60	C1–2	Anterior	Dysphagia	Transoral decompression

NS = not specified; Pt = patient.

the histological diagnosis was actually consistent instead with calcium pyrophosphate dehydrate deposition disease. This highlights the difficulty in diagnosing these lesions and emphasizes the need for awareness of this pathology. Our case is the fourth reported case of histologically confirmed TC affecting C1–2, and the first case involving treatment with transoral decompression.

Dysphagia at the CVJ

As it was with our patient, dysphagia is a common presenting symptom for patients with CVJ pathology. In a previous review of 148 patients with ventral foramen magnum lesions, Dickman et al.⁷ identified 29 patients (20%) who presented with dysphagia prior to surgery. Further examination of these patients revealed that 18 (12%) had uncoordinated swallowing and that 26 (18%) had a decreased gag reflex. Of note, this analysis included all patients who underwent transoral decompression for any pathology, with most of the 148 patients having rheumatoid arthritis ($n = 58$ [39%]), congenital malformations ($n = 37$ [25%]), or chronic dens dislocations ($n = 29$ [20%]). The series included no case of calcium hydroxyapatite, uric acid (gout), or calcium pyrophosphate (pseudogout). In a more recent report from Fenoy et al.,¹¹ 21 patients with calcium pyrophosphate deposition at the CVJ were specifically analyzed. Of these 21 patients, 3 (14%) presented with dysphagia as a prominent symptom, and 6 (29%) were found to have lower cranial nerve deficits. No patient identified in our literature review of TC presented with dysphagia or lower cranial nerve deficits (Table 1).

The Transoral Approach

The transoral approach provides direct access to ventral midline pathology at the CVJ, and it has been well described for extradural lesions in this region.⁷ The transoral approach is best suited for a compressive pathology of the brainstem and spinal cord from the clivus to the level of approximately C-3, and extended transoral exposures (e.g., transoral-translabiomandibular and transoral-transmaxillary) have been adapted for more extensive le-

sions. Numerous variations of the transoral approach have been described in the medical literature, and most cases are performed for basilar invagination from various pathologies.^{6,12,22} Nonetheless, no previous reports exist of the transoral approach being performed for TC, so the current case report adds to the existing indications for this procedure.

The main limitation of the transoral technique is approach-associated morbidity. Complications include, but are not limited to, cerebrospinal fluid leak or fistula formation, meningitis, oral wound dehiscence or wound infection, need for prolonged parenteral nutrition, respiratory failure, cerebrovascular injury, and delayed occipitocervical instability.^{6,7,11,12,22,24} Rates of complications vary greatly among series; however, the incorporation of perioperative broad-spectrum antibiotics, utilization of image guidance, and consideration of simultaneous or staged posterior fixation are relatively recent interventions that have improved the perioperative morbidity associated with the transoral approach. In our current practice, we use broad-spectrum antibiotics (clindamycin and ceftriaxone sodium) for 4 days postoperatively to reduce the oral flora and a feeding tube for the same period to facilitate wound healing. In addition, we use intraoperative imaging and navigation, which we believe facilitates the identification of anatomical landmarks and optimizes the extent of resection of pathology. In the present case, the comparison of dynamic radiographs obtained preoperatively and postoperatively confirmed stability across the CVJ, and thus obviated the need for posterior fixation.

Conclusions

TC is a calcium deposition disorder that can result in tumor-like lesions in the spine, causing local mass effects. Involvement of the CVJ is rare but may be amenable to transoral decompression. In this case of TC at the CVJ, decompression alone was adequate to relieve the patient's dysphagia, and fusion was not required. Neurosurgeons should be aware of this pathology and include it in the dif-

ferential diagnosis of lesions in this region because prompt recognition facilitates its diagnosis and treatment.

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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: Theodore, Mooney, Oppenlander. Acquisition of data: Theodore, Mooney, Oppenlander. Analysis and interpretation of data: Theodore, Mooney, Oppenlander. Drafting the article: Theodore, Mooney, Oppenlander. Critically revising the article: Theodore, Mooney, Oppenlander. Reviewed submitted version of manuscript: Theodore. Administrative/technical/material support: all authors. Study supervision: Theodore.

Supplemental Information

Videos

Video 1. <https://vimeo.com/190255264>.

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