Tumoral calcinosis presenting as a deformity of the thoracic spine

Case report

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The authors describe a rare case of tumoral calcinosis (TC) of the thoracic spine in a 13-year-old boy with thoracic scoliosis. The patient presented with a 2-year history of back pain. He had no personal or family history of bone disease, deformity, or malignancy. Magnetic resonance imaging revealed a heterogeneously enhancing mass involving the T-7 vertebral body and the left pedicle. Computed tomography findings suggested that the mass was calcified and that this had resulted in scalloping of the vertebral body. The lesion was resected completely by using a left T-7 costotransversectomy and corpectomy. The deformity was corrected with placement of a vertebral body cage and pedicle screw fixation from T-5 to T-9. Pathological analysis of the mass demonstrated dystrophic calcification with marked hypercellularity and immunostaining consistent with TC. This represents the third reported case of vertebral TC in the pediatric population. Pediatric neurosurgeons should be familiar with lesions such as TC, which may be encountered in the elderly and in hemodialysis-dependent populations, and may not always require aggressive resection. (DOI: 10.3171/2011.8.PEDS11193)

Key Words • spinal deformity • tumoral calcinosis • spine

Tumoral calcinosis is a rare pathological entity marked by dystrophic calcification in soft tissue, and is frequently associated with secondary hyperparathyroidism. These patients usually present with hyperphosphatemia, normocalcemia, and focal and multifocal periarticular soft-tissue calcifications, which take the form of calcium hydroxyapatite crystals surrounded by a foreign-body giant cell. Although this entity is rare, an increasing number of TC cases is being identified in patients receiving renal dialysis and in the aging population, in whom TC related to degenerative and inflammatory changes in bone structures develops. A third group of patients have familial TC, which often occurs as painless lesions before the age of 20 years. Patients with familial TC usually have a heritable abnormality of vitamin D metabolism and mutations in FGF23 or components in the FGF23 signaling pathway. In sporadic cases, patients have no metabolic abnormalities related to calcium, phosphate, or vitamin D.

Although reports of TC of the spine are rare, the true incidence may be higher because these lesions are often asymptomatic and difficult to detect on plain radiography. In many cases these lesions are small and occur at sites of inflammation and degenerative changes, prerequisites that will surely increase the presence and detection of these lesions in the aging population. Clinicians and radiologists may misdiagnose these lesions as malignancies. Consequently, large, aggressive resection of these mostly benign lesions is common, and pathologists may interpret the biopsy or surgical specimens as nondiagnostic when they show no malignant features. With a growing geriatric population, clinicians should be familiar with the presentation of and management options for these lesions.

Abbreviation used in this paper: TC = tumoral calcinosis.

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**Case Report**

**History and Examination.** This 13-year-old boy with a 2-year history of midthoracic back pain and rapidly progressing scoliosis was referred to our spine clinic for evaluation and correction of his spinal deformity. The patient’s neurological evaluation revealed 5/5 strength in all muscle groups and no bowel or bladder symptoms. His reflexes were normal, and his sensation to light touch and his proprioception were intact. No clonus was noted, and Babinski reflexes were normal. Initial laboratory values were notable for a calcium level of 8.5 mg/dl. All other laboratory values were within normal limits.

**Neuroimaging.** Admission MR imaging showed a dorsally exophytic mass at the T-7 vertebral body; the lesion was hypointense on both T1- and T2-weighted sequences (Fig. 1 left). The mass had a large posterior extension associated with severe stenosis of the central spinal canal (Fig. 1 right). Contrast CT scanning showed a 2.5 × 3.1 × 2.6–cm densely calcified lesion with a lucent rim and scalloping of the vertebral body. The differential diagnosis based on the imaging findings consisted of metastasis, osteoblastoma of the spine, meningioma, giant cell tumor, and Ewing sarcoma. An evaluation for metastatic disease suggested no other tumor sites.

**Operation.** Given the patient’s rapidly progressing scoliosis and severe spinal cord compression, surgical excision after tumor embolization was recommended. A T-7 costotransversectomy and corpectomy was performed on the left side for complete tumor removal and spinal cord decompression. Vertebral reconstruction and deformity correction was achieved with placement of a 10-mm mesh cage (Synthes) for anterior column support and T5–9 pedicle fixation (Synthes).

The lesion was pale yellow, soft, and grainy. It was easily scooped from the vertebral body, leaving a cortex of normal-appearing bone. Gross pathological examination of the mass showed markedly hypercellular proliferation of cells with predominantly epithelioid features and extensive dystrophic calcification (Fig. 2A). The lesion demonstrated a mononuclear inflammatory infiltrate, including plasma cells and lymphocytes. Immunostaining revealed uniform strong staining with CD68 (Fig. 2B) and vimentin (Fig. 2C), consistent with a histiocytic origin of the tumor. Outside pathological review confirmed the diagnosis of TC. Workup revealed this to be a case of idiopathic TC.
Postoperative Course. The patient had an uncomplicated hospital course and was discharged wearing an external orthosis. Neurological evaluation confirmed 5/5 strength in all muscle groups, and no bowel or bladder symptoms. Reflexes were normal and sensation to light touch and proprioception were intact.

Follow-up MR imaging findings suggested complete resection of the T-7 lesion with significant correction of the Cobb angle and stabilization with instrumentation (Fig. 3). At his 1- and 18-month postoperative follow-up examinations, the patient was neurologically intact, but his scoliosis appeared to have progressed without any recurrence of the TC (Fig. 4).

Discussion

Analysis of the literature revealed 41 cases of TC of the spine reported between 1952 and 2011.1–13,15–17,20–23 There were 22 cases (54%) of spinal TC in female and 19 (46%) in male patients. The mean age at diagnosis was 55 years, with females being diagnosed at a slightly earlier age (mean 53.3 years, range 12–71 years) than males, who have been diagnosed at a mean age of 56.05 years (range 17–90 years). Most lesions have involved the lumbar spine (46.3%), but lesions in the cervical (36.5%), thoracic (22%), and sacral segments (5%) have also been reported. The severity of presented neurological symptoms has varied between pain and severe neurological deficit. Rheumatoid arthritis, systemic sclerosis, and the Raynaud phenomenon are common comorbid conditions at the time of diagnosis. In the cervical region, these lesions often manifest with torticollis and restriction of neck motion. Although conservative management has been reported as a treatment option in some cases, most lesions advance, requiring surgery within 3 to 6 months. Of the reported cases, resection was successful in 38 (92.7%). There have been no reported cases of recurrence after resection in the spine. This patient represents only the third reported case of TC of the spine in the pediatric population (Table 1).

Conclusions

The incidence of TC is likely to increase as the population ages, especially in conjunction with the diabetes pandemic. Although TC is rare in the pediatric population, neurosurgeons should be familiar with the treatment of these lesions.

Disclosure

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 to the study and manuscript preparation include the following. Conception and design: Theodore, Kalani. Acquisition of data: Kalani, Martirosyan. Analysis and interpretation of data: Kalani, Martirosyan, Little. Drafting the article: Theodore, Kalani, Martirosyan. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors.

TABLE 1: Literature review for demographic data and presentation in pediatric patients with TC

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Age (yrs), Sex</th>
<th>Lesion Location</th>
<th>Comorbid Condition</th>
<th>Preop Diagnosis</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ohashi et al., 1996</td>
<td>14</td>
<td>12, F</td>
<td>cervical spine</td>
<td>torticollis</td>
<td>not reported</td>
<td>resection</td>
</tr>
<tr>
<td>Mooney &amp; Glazier, 1997</td>
<td>2</td>
<td>17, M</td>
<td>C1–2</td>
<td>neuroblastoma, calcified hemATOMa, ganglioneuroma, TC</td>
<td></td>
<td>resection</td>
</tr>
</tbody>
</table>

Fig. 3. Follow-up coronal T2-weighted MR imaging study obtained after resection of the T-7 lesion demonstrating significant correction of the spinal deformity with instrumentation.

Fig. 4. Anterior-posterior (left) and lateral (right) radiographs of the spine showing the intact hardware and progression of the scoliosis at the thoracolumbar junction.
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Approved the final version of the manuscript on behalf of all authors: Theodore. Administrative/technical/material support: Little.

References

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