Tumoral Calcinosis: Serial Images to Monitor Successful Dietary Therapy

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Abstract. Tumoral calcinosis involves formation of periarticular calcified soft tissue masses. Experimental evidence suggests a metabolic etiology with dietary restriction of calcium and phosphorus as beneficial therapy. We prospectively monitored serum levels of calcium, phosphorus, alkaline phosphatase, and erythrocyte sedimentation rate (ESR) while successfully treating a patient with tumoral calcinosis. The values were compared with changes on serial radiographic and radionuclide bone and gallium images. Our work suggests using serial serum phosphate levels and the ESR as the most sensitive indications of progress in dietary treatment of tumoral calcinosis.

Key words: Tumoral calcinosis – Soft tissue calcification – Metabolic calcification – Renal tubular dysfunctions – Dietary therapy in skeletal disorders

Tumoral calcinosis results in dense periarticular calcific masses in tissues around the hip, shoulder, and elbow; other joints are less commonly involved. The serum phosphorus is elevated; the serum calcium is normal or only slightly elevated in all cases. The alkaline phosphatase is normal. The incidence is higher in black than white populations, and the frequency is equal for both sexes. A familial component has been noted [7, 9]. Untreated lesions may progress to limit function, disfigure, ulcerate, and undergo secondary infection.

The differential diagnosis is extensive. Benign soft tissue calcifications including bursal calcifications or post-traumatic calcification or ossification (“myositis ossificans”) can be large. Patients with renal transplants and those with collagen diseases can develop large calcified masses. Metastatic calcification in soft tissues occurs in systemic metabolic conditions with and without hypercalcemia. Additional and less likely considerations are chondrosarcoma or malignant degeneration of osteochondroma.

Case Report
A 45-year-old Greek man complained of a large painful mass in the left shoulder with complete restriction of glenohumeral motion. The mass had been present for nine years. Initial treatment with multiple cortisone injections gave symptomatic relief but no decrease in size. Resection of the mass was attempted twice and the mass recurred within a few months each time.

Previously, the patient had had three similar lesions: at age six years a nodule on the left foot resolved spontaneously, at age fourteen years a mass on the medial aspect of the right ankle drained a "white, chalky liquid" and was resected; and at age sixteen years a similar mass on the lateral aspect of the right knee was excised. None of the lesions recurred. There was no history of trauma at the involved sites. The patient said his parents, three siblings, and only child had never been similarly afflicted.

Clinical evaluation revealed localized pain, a palpable mass, and limitation of motion at the shoulder. Initial radiographs (Fig. 1A) showed a large, amorphous periarticular calcification surrounding the left proximal humerus with no bone or joint destruction. Gallium-67 citrate and 99mTc-methylene diphosphonate (99mTc-MDPA) (Fig. 1B) bone images showed an intense radionuclide uptake in the mass. Admission laboratory values included normal serum calcium, normal alkaline phosphatase, elevated serum phosphate, and elevated erythrocyte sedimentation rate (ESR) (Table 1). Tubular phosphate resorption was slightly elevated at 88.5% (normal 76-87%), urea nitrogen and serum creatinine were normal.

The lesion was biopsied and the patient started a strict diet which allowed 600 mg phosphate daily (the patient's average intake was 2,000 mg, and the recommended average intake is 800 mg) and 300 mg calcium (patient's average intake was 1,225 mg, recommended intake is 800 mg). Intake of 60 ml Maalox and 120 ml Amphogel daily was also required. His serum phosphate levels immediately started to decrease and by three months were within the normal range (Table 1). His ESR dramatically decreased while his calcium remained unchanged. The patient still complained of considerable pain in the shoulder. Clinical assessment of size of the mass and glenohumeral motion showed no improvement.

Between six and eight months after initiation of therapy the
Fig. 1A, B. A.S. prior to initiation of dietary therapy. A Left shoulder radiograph on admission, shows a large periarticular calcification surrounding the left glenohumeral joint. The underlying bones and glenohumeral joint space appear intact. B Initial bone and gallium images, posterior views, show a lesion corresponding well to the calcified mass in Fig. 1A

Table 1. Serum levels with dietary restriction

<table>
<thead>
<tr>
<th>Date</th>
<th>Phosphate (2.8–4.2 mg/dl)</th>
<th>Calcium (8.4–10.1 mg/dl)</th>
<th>ESR (0–20 mm/h)</th>
<th>Alkaline phosphatase (10–85 IU)</th>
</tr>
</thead>
<tbody>
<tr>
<td>29 August 78</td>
<td>5.5</td>
<td>9.4</td>
<td>97</td>
<td>45</td>
</tr>
<tr>
<td>Phosphate deprivation diet started</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>17 October 78</td>
<td>4.5</td>
<td>9.3</td>
<td>95</td>
<td>54</td>
</tr>
<tr>
<td>5 December 78</td>
<td>4.1</td>
<td>9.8</td>
<td>86</td>
<td>61</td>
</tr>
<tr>
<td>9 March 79</td>
<td>4.7</td>
<td>9.5</td>
<td>48</td>
<td>55</td>
</tr>
<tr>
<td>16 May 79</td>
<td>3.9</td>
<td>9.9</td>
<td>30</td>
<td>65</td>
</tr>
<tr>
<td>23 July 79</td>
<td>3.5</td>
<td>9.7</td>
<td>50</td>
<td>61</td>
</tr>
<tr>
<td>18 September 79</td>
<td>2.7</td>
<td>10.1</td>
<td>40</td>
<td>62</td>
</tr>
<tr>
<td>10 November 79</td>
<td>3.3</td>
<td>9.7</td>
<td>45</td>
<td>59</td>
</tr>
<tr>
<td>Amphogel stopped; diet less strictly adhered to</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>29 November 79</td>
<td>5.2</td>
<td>9.4</td>
<td>–</td>
<td>43</td>
</tr>
</tbody>
</table>

Patient reported gradual relief of pain. He had a significantly increased range of motion at the glenohumeral joint, with softening and disappearance of the mass to clinical examination. At 15 months, radiographs and radionuclide images showed a marked decrease in size and intensity of the shoulder lesion (Fig. 2). In addition, however, a right hilar and paratracheal mass was noted which proved on biopsy to be metastatic oat cell carcinoma. During subsequent chemotherapy, the patient abandoned the strict low phosphorus diet; the serum phosphate immediately increased while calcium remained within the normal range.

Discussion

Classic therapy for tumoral calcinosis has been complete surgical resection; however, the recurrence rate is high. Accordingly, surgery should be regarded as palliative, and critical structures should not be sacrificed in attempted resection. The use of cortisone and other anti-inflammatory agents has not been proven successful [9]. Therapeutic radiation has been shown to be of no value [4, 9].

Many etiologic possibilities have been suggested but recent and convincing experiments [8] have shown that parathormone levels are normal in patients with tumoral calcinosis. Similarly, they have normal gastrointestinal absorption of calcium; maintenance of the normal serum calcium and positive calcium balance are the normal direct renal tubular effects of hyperphosphatemia. Some data suggest that the etiology of tumoral calcinosis is related to a specific enhancement in renal tubular phosphate resorption, probably originating in the proximal tubule. Based