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Authors
SEEGER, LL
BUTLER, DL
ECKARDT, JJ
et al.

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Tumoral calcinosis-like lesion of the proximal linea aspera

Leanne L. Seeger, M.D. 1, Debra L. Butler, M.D. 1, Jeffrey J. Eckardt, M.D. 2, Lester Layfield, M.D. 3, and John S. Adams, M.D. 4

1 Department of Radiological Sciences, 2 Department of Surgery, Division of Orthopedic Surgery, 3 Department of Pathology, and 4 Department of Medicine, Division of Endocrinology, UCLA School of Medicine, Los Angeles, California, USA

Abstract. Tumoral calcinosis is presently a poorly defined disease. In its classic form, it consists of multiple large foci of benign mineralization in the soft tissue adjacent to bone near large joints. Patients are generally of African descent and are adolescents or young adults at presentation. Both metabolic and traumatic etiologies have been proposed. We report six adult Caucasian patients with lesions that pathologically resembled tumoral calcinosis. All lesions were small (less than 3 x 3 cm) and were located along the proximal linea aspera of the femur. All patients presented with pain. Because of the atypical patient population and the unusual size and location of the lesions, we refer to this process as a "tumoral calcinosis-like lesion." A typical radiographic appearance and location, together with appropriate clinical history, can strongly suggest this diagnosis.

Key words: Tumoral calcinosis Linea aspera lesion

Tumoral calcinosis is a poorly understood condition in which benign calcifications form in soft tissue. These foci are usually found in the vicinity of large joints but do not involve the synovium itself [4, 6, 16, 23, 30-32, 36, 38]. It has also been reported that the lesions do not involve adjacent bone [4, 6, 16, 21, 23, 29, 36, 38]. Although a metabolic etiology has been proposed, some authors feel that the condition may occasionally be related to trauma [5, 10, 15, 30, 36, 39, 42, 44]. We describe six patients with symptomatic soft-tissue calcification adjacent to the superior aspect of the linea aspera and adherent to the underlying bone. Five patients underwent surgical excision of the mass. Although these lesions occurred in an atypical patient population and an unusual location, they resembled classic tumoral calcinosis pathologically.

Materials and methods

The six patients described in this report were seen between 1986 and 1989. Initial clinical and radiographic evaluation was obtained outside our institution. Although the lesional location and appearance made a diagnosis of malignancy unlikely, all patients were referred for oncologic evaluation of presumed osteosarcoma. Patient ages ranged from 47 to 65 years (mean age, 55 years). Three patients were male, and three were female; all were Caucasian. The lesions were right-sided in three patients and left-sided in three.

Five patients presented within 2 months of the onset of pain in the posterior thigh and/or hip region, and one reported a 2 year history of left posterior hip pain. One patient recalled a specific incident of trauma to the hip region. Another, with a history of alcoholism, could not deny recent trauma. Two patients had undergone recent steroid injection for presumed trochanteric bursitis on the side of the lesion. No patients reported a family history of tumoral calcinosis, and the lesions were unifocal in all cases.

Laboratory studies were limited. Serum calcium level was obtained in only one patient and was normal. Serum alkaline phosphatase level was measured in only one patient and was mildly elevated (122, normal = 30-105 IU/l). Wintrobe erythrocyte sedimentation rate in the same patient was also elevated (180, normal range = 0-10). Serum phosphorus and parathyroid hormone levels were not measured in any of the patients.

Plain films were obtained in all patients, computed tomography (CT) scans in four, technetium methylene diphosphonate Tc-99m (99mTc-MDP) radionuclide bone scans in three, and magnetic resonance (MR) images in two.

Four patients underwent excisional biopsy at our institution, and one underwent biopsy elsewhere. Symptoms resolved after surgery in these five patients, and there has been no evidence of recurrence with 5-42 months of follow-up. The sixth patient is pain-free at 36 months.

Results

Imaging studies

In all patients, radiographs revealed soft-tissue mineralization that could not be separated from the underlying posterior femoral cortex. All lesions were located in the subtrochanteric region at the insertion of M. glutaeus maximus and/or adductor magnus. The calcifications were dense and well defined in two patients (Fig. 1) and were amorphous in four (Fig. 2). All lesions measured...
Fig. 1 A–C. A 48-year-old woman with pain in the right posterior thigh of 7 weeks' duration. There was no history of trauma. A Anteroposterior radiograph of the hip reveals dense calcification superimposed over the proximal shaft (arrow). B Close-up of John-son lateral radiograph shows that the calcification is posterior and contiguous with the underlying cortex. C CT scan. The calcification is well defined, and there is no associated soft-tissue mass. Underlying cortex is normal. Biopsy revealed tumoral calcinosis.

Fig. 2 A–C. A 47-year-old man with history of pain in the right lateral hip of 6 weeks' duration. There was a history of hip trauma 4 months previously. A Johnson lateral radiograph reveals amorphous calcification adjacent to the posterior aspect of the proximal femur. B 99mTc-MDP bone scan, anteroposterior view. There is markedly increased radionuclide uptake in the region of the lesion. Biopsy revealed tumoral calcinosis. C CT scan. The adjacent outer cortex is eroded, but the endosteal surface is intact.
Fig. 3. Aggregates of granular to amorphous material with focal fibrohistiocytic response. (H and E, ×75)

Fig. 4. Amorphous and focally calcified debris surrounded by palisading epithelioid histiocytes and reactive fibroblasts. (H and E, ×75)

Fig. 5. Reactive new bone formation and fibrohistiocytic infiltrate surrounding granular debris at lower left. (H and E, ×75)

less than 3×3 cm in size. CT study confirmed that all lesions were contiguous with the underlying femoral cortex, and the medullary cavity was normal. In five patients, the superficial cortex was eroded adjacent to the mineralization, but the endosteal surface was intact. Radionuclide bone scans of three patients showed focal moderate (one patient) to markedly increased uptake (two patients) in the region of the lesion. MR sequences obtained in two patients included spin echo sagittal T₁-weighted images and axial T₁- and T₂-weighted images. In one patient the lesion was evident in the sagittal images as a well-defined, low to intermediate signal intensity mass immediately adjacent to the cortex. Even retrospectively, the lesions were difficult to see on axial images. MRI demonstrated normal underlying marrow in both patients.

Pathologic findings

The appearance of the specimens from all patients was similar. On gross examination the bone specimens obtained by curettage contained fibrous tissue and chalky to cheesy, yellow-white, friable material. Frequently, this material lay in a partially cystic space surrounded by fibrous tissue and a bony capsule.

Microscopically, all specimens were characterized by aggregates of amorphous to finely granular material that was light pink to pale blue on hematoxylin and eosin staining (Fig. 3). These aggregates were frequently surrounded by a fibrohistiocytic reaction that included foreign body type giant cells (Fig. 4). In other areas, the granular material lay in dense fibrous tissue with little or no inflammatory response. The larger collections of debris were surrounded by reactive bone formation and a cellular fibrous stromal reaction running between the bony trabeculae (Fig. 5). The material did not show birefringence with polarized light.

Discussion

Tumoral calcinosis is the descriptive term given to extraosseous, nodular, calcified masses [33, 41]. Deposits are usually associated with bony prominences in the region of a joint, may be multiple, and may become symp-
moral calcinosis, the etiology of this condition remains debated and is poorly understood. Most patients reported in the literature were adolescents or young adults and were from the tropical and subtropical regions of Africa [1, 22, 30, 31, 42]. Hyperphosphatemia has been found in some individuals, but serum calcium and parathyroid hormone levels are reported normal [3, 21, 22, 24, 26, 32, 34, 35, 38, 45]. An apparent inherited disorder of phosphate and vitamin D metabolism has also been described [8, 19, 28, 34, 37, 38, 48]. Trauma has been implicated as a cause in selected patients [5, 10, 15, 30, 36, 39, 42, 44]. Although surgical specimens of the patients reported here revealed findings typical of tumoral calcinosis, the patients' age and race are atypical, and the lesions were solitary, unusually small, and in an atypical location. For this reason, we refer to this condition as a "tumoral calcinosis-like lesion."

Hydroxyapatite crystals constitute the mineral component of tumoral calcinosis with calcium phosphate and calcium carbonate in fluid suspension [1, 18, 20]. It is therefore not surprising that this condition is observed with greater frequency in patients with systemic disorders in which the "calcium-phosphate" product is elevated, particularly in hyperphosphatemic disorders such as vitamin D intoxication, chronic renal failure, and hyperphosphatemic tumoral calcinosis [4, 9, 14, 25]. The latter disorder appears to be an autosomal dominant disease with variable penetrance, characterized by inappropriately avid renal phosphate retention, inappropriately high serum levels of the hormonal form of vitamin D (1,25-dihydroxyvitamin D), and a unique hypoplastic dental lesion [27, 46].

Tumoral calcinosis in the presence of a normal circulating phosphate concentration is, however, the rule rather than the exception [4, 6, 13, 17, 39, 43]. Hyperphosphatemia is reported to occur in only one-third of patients, and in them tumoral calcinosis may be a distinct clinical entity [37]. No known underlying disorder of mineral ion homeostasis was evident in any of our six patients. This finding strongly suggests that local tissue factors (i.e., pH, inflammatory cytokines, etc.) were responsible for initiating calcium-phosphate deposition. It is clear that once a nidus of hydroxyapatite is formed, further salt deposition is favored [33].

The histologic differential diagnosis for the lesions seen in our patients includes tumoral calcinosis, myositis ossificans, calcific bursitis, and calcific tendinitis. The lesions contained large geographic areas of granular or amorphous calcification, modest numbers of chronic inflammatory cells surrounding the calcifications, and bands of fibrosis like those seen in tumoral calcinosis. Pathologically, they are distinct from true heterotopic bone formation such as that which occurs in myositis ossificans, because tumoral calcinosis lacks the protein matrix scaffolding characteristic of bone [33]. These lesions can, however, contain regions of reactive bone formation [30, 31, 40]. Myositis ossificans is characterized by a prominent zonal pattern of bone formation and maturation. Peripheral maturation of bone is present, whereas central portions of the mass consist of immature spindle cell proliferation. This feature was not present in our specimens. In addition, amorphous calcific debris, as seen in our material, is not a component of myositis ossificans. Although the lesions were associated with lamellar and woven bone, this bone was either residual cortical bone or reparative bone and did not constitute part of the primary pathologic process.

Calcific bursitis does contain calcified debris similar to that seen in our patients, but it arises within a bursa and is associated with synovial tissue and cystic areas, findings which were not present in our material. Calcific tendinitis has many pathologic similarities to our lesions, but it usually contains areas of degenerating tendon, larger numbers of chronic inflammatory cells, and a more disorganized pattern of fibrosis. The lesions of calcific tendinitis are usually smaller and more superficial than those of tumoral calcinosis and do not increase in size. In addition, the invasion of cortical bone is unusual for either tendinitis or tumoral calcinosis.

In general, tumoral calcinosis lesions do not invade the cortex of the underlying normal bone [4, 6, 16, 21, 23, 29, 36, 38]. A number of cases of underlying bone involvement have, however, been reported [13, 36, 44]. In our series of six patients, all lesions were adherent to the underlying bone, and CT scan revealed the presence of superficial cortical erosion in five, a highly unusual and rarely reported feature of classic tumoral calcinosis. Despite this bony involvement, we believe that these lesions appear very similar to tumoral calcinosis.

In our set of patients, thin-section CT was the imaging procedure of choice for defining the lesional extent and evaluating the underlying cortex. The lesions were difficult to define on MR images, even with a 5-mm slice thickness, probably because of the low sensitivity of MRI for small soft-tissue calcifications and subtle cortical erosions. Increased radionuclide uptake on the bone scan is a sensitive but nonspecific finding and could reflect either neoplasm or trauma [2, 7, 11, 12, 47].

The typical location of these tumoral calcinosis-like lesions and their characteristic appearance on plain films and CT scans allow this entity to be included in the differential diagnosis of juxtacortical mineralization in the appropriate clinical setting. This radiographic appearance should, however, not delay biopsy confirmation if malignancy is suspected.

References

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