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Painful Ulnar Lesion in a 16-year-old Boy

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History and Physical Examination

A 16-year-old boy presented with swelling and a firm painful mass located in the middle of his right forearm 5 weeks after a fall from his bicycle. Immediately after this fall, he had experienced pain, and a contusion of his right forearm was diagnosed in the local emergency department. Gradually, the forearm swelling increased. The pain was mild and did not interrupt sleep. The patient was referred for a second evaluation 3 weeks later in a local public health center. A radiographic assessment of his right forearm (Fig. 1) was performed and the radiographs were erroneously interpreted as normal by a general practitioner, who advised rest.

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Five weeks postinjury, the symptoms continued and the swelling increased. Physical examination in our hospital revealed a firm mass in his middle forearm, predominantly in the ulnar side, over an area approximately 7×7 cm. There was tenderness to palpation and the overlying skin was apparently normal. He was not febrile, had no palpable lymph nodes, and had full motion in the neighboring joints. There was no medical history. Calcium, phosphorus, alkaline phosphatase, and parathormone levels were normal.

Plain radiography (Figs. 2, 3A), bone scan (Fig. 3B), multidetector computed tomography (MDCT) (Fig. 4), and MRI (Fig. 5) of the right forearm were performed.

Based on the history, physical examination, and imaging studies, what is the differential diagnosis?

Imaging Interpretation

Plain radiographs (lateral, anteroposterior, and focused) 3 weeks after injury showed a lytic lesion in the diaphysis of the ulna with expansion into the soft tissues along the radial margin with faint peripheral calcification. In addition, there was periosteal reaction along the medial margin of the lesion, which had an onion skin pattern (Fig. 1). The overall appearance was suggestive of an aggressive lesion. The plain radiograph 5 weeks after injury showed interval progression of the lesion with destruction of the lateral cortex, a more extensively calcified lacy pattern in the area of soft tissue extension, a Codman's triangle, and more obvious lytic change in the medullary cavity (Fig. 2A).

The bone scan, performed 7 weeks after injury, showed increased uptake of the lesion (Fig. 2B). There were no other foci of abnormal radionuclide uptake elsewhere in the skeleton. The MDCT, performed 8 weeks after injury, showed a lytic lesion in the ulna with destruction of the

Fig. 1A–C (A) Lateral, (B) anteroposterior, and (C) focused plain radiographs, taken 3 weeks postinjury, show a soft tissue mass with faint calcification on the radial side (open arrows) and a periosteal reaction on the contralateral side (thin arrows).

Fig. 2A–B (A) A plain radiograph, taken 5 weeks postinjury, shows a parosteal lesion with inhomogeneous calcification of the soft tissue element (open arrows), periosteal reaction with Codman triangle

(solid white arrow), and a permeative pattern in the medullary area (black arrows). (B) A bone scan performed 7 weeks postinjury shows increased uptake of the lesion.

Fig. 3A–E The MDCT performed 8 weeks after injury shows (A) the fluid-fluid level (solid arrows) in the matrix of the soft tissue elements of the mass. (B) A larger cavity with fluid-fluid level (arrow) also is seen. (C) The calcified matrix (open arrows) and cortical lysis

(thin arrow) are evident. (D) The longitudinal reconstruction and (E) the three-dimensional reconstruction show the cortical shell of the extraosseous component of the lesion.

Fig. 4A–B (A) The oblique sagittal and (B) the transverse T2-weighted turbo spin echo MR images, taken 8 weeks postinjury, show the solid component of the extraosseous lesion (open arrows) and the

cysts in the lesion, with fluid-fluid levels (solid arrows). The marrow infiltration has a cystic appearance with well-defined borders (thick arrows).

lateral cortex and a large associated expansile soft tissue mass. The area of soft tissue expansion had internal calcification, which appeared somewhat chondroid and had a thin continuous peripheral calcified rim. On closer inspection, there was evidence of fluid-fluid levels in the soft tissue component (Fig. 3).

MRI was performed 8 weeks after the injury. Sagittal and axial T2-weighted images showed complex, predominantly T2 hyperintense signal in the area of soft tissue extension with areas of cystic change and confirmation of the fluid-fluid levels observed on the CT. The intraosseous component of the lesion was cystic with

well-defined, low-signal borders with the adjacent normal marrow (Fig. 4).

Differential Diagnosis

- Giant cell tumor
- Aneurysmal bone cyst
- Giant cell reparative granuloma
- Chondroblastoma
- Ewing's sarcoma
- Osteosarcoma

Fig. 5A–C (A) A low-power photomicrograph shows multiple spaces in association with solid areas. Spaces contained blood or were empty (Stain, hematoxylin and eosin; original magnification, $\times 40$). (B) A higher magnification photomicrograph shows numerous osteoclast-like giant-cells, prominent benign osteoid, which tend to anastomose, and reactive new bone formation. The solid areas contain fibroblast-like cells with benign features and scattered lymphocytes (Stain, hematoxylin and eosin; original magnification, $\times 100$). (C) No nuclear atypia or immature cells forming osteoid were observed (Stain, hematoxylin and eosin; original magnification, $\times 400$).

An open biopsy of the lesion was performed 2 months after the injury (Fig. 5). A new radiograph, taken the day before definitive surgical treatment of the lesion (12 weeks

Fig. 6 A plain radiograph, taken 12 weeks after injury, shows a calcified parosteal lesion on both sides of the ulnar diaphysis.

postinjury), showed a calcified parosteal lesion on both sides of the ulnar diaphysis (Fig. 6).

Based on the history, physical findings, imaging studies, and histology, what is the diagnosis and how should the lesion be treated?

Histology Interpretation

Histologic sections examined under low magnification showed multiple spaces (structures that do not have endothelial lining) in association with solid areas. Spaces contained blood or were empty (Fig. 5A). Higher magnification revealed numerous osteoclast-like giant cells, prominent benign osteoid (as opposed to malignant osteoid that is produced in cases of osteosarcoma) that tended to anastomose, and reactive new bone formation (Fig. 5B–C). The solid areas contained fibroblast-like cells with benign features and scattered lymphocytes. No nuclear atypia or immature cells forming osteoid were noted.

Diagnosis

Solid variant of aneurysmal bone cyst (giant cell reparative granuloma).

Discussion and Treatment

Solid variant of aneurysmal bone cyst (ABC) and giant cell reparative granuloma (GCRG) are different terms describing nonneoplastic lesions with similar histologic features and have been used interchangeably in the pathology literature. GCRG initially was reported as a reactive lesion to interosseous hemorrhage in the jaw in 1953 [8]. The original report on solid ABC was published in 1983 [17], describing lesions characterized by florid fibroblastic or fibrohistiocytic proliferation, osteoblastic differentiation with osteoid production, areas rich in osteoclast-type giant cells, aneurysmal sinusoids, and occasional foci of degenerate calcifying fibromyxoid tissue, histologic features that can be found only in the solid parts of ABCs. Cytogenetic studies have reported translocations involving 16q22 and/or 17p13 are characteristic of ABCs [15]. Findings of a recent study support that at least a subset of GCRGs may be neoplastic and that these lesions

differ cytogenetically from classic giant cell tumors of bone or solid ABC [6]. Further cytogenetic research is necessary to clearly define the pathogenetic relations of these lesions.

The differential diagnosis for our patient included giant cell tumor, ABC, chondroblastoma, Ewing's sarcoma, and osteosarcoma. Giant cell tumors (GCT) are true expansile neoplasms, which erode the cortex and may extend beyond the bone into the soft tissues located mainly in the metaphyses of young adults [14, 19]. In our case, GCT should be included in the differential diagnosis because of the presence of prominent osteoclast-type giant cells, however, the imaging characteristics would be atypical for this lesion. In GCRG, however, these cells are distributed focally and there is no diffuse pattern, which is present in GCT [1, 19]. Bone formation in a GCT rarely is present, as opposed to the case presented. Despite the fact that radiographically the lesion did not match a brown tumor (in hyperparathyroidism), this is mentioned because it is indistinguishable histologically from GCRG [9, 14]. Chondroblastoma can be complicated with ABC and as much as 50% of the cases have stippled calcifications. Histologically, the tumor contains giant cells and sometimes may be misdiagnosed as GCT. However, this is rarely located in the diaphysis [3]. The hallmark of

Fig. 7A–B (A) A plain radiograph, taken immediately postoperatively, shows the lesion after curettage, application of pure alcohol for 10 minutes, and filling of the defect with a synthetic, biodegradable, calcium phosphate-based bone substitute. (B) A plain radiograph, taken 1 year postoperatively, shows no local recurrence.

osteosarcoma is the presence of anaplastic cells with atypical mitoses, which were absent in our case [10, 13, 18]. In addition, osteosarcoma was excluded as a possible diagnosis because of the presence of a mature calcified shell in the soft tissue. Although, this localized painful mass in a 16-year-old boy should raise suspicion of Ewing's sarcoma, a more permeative pattern of marrow involvement and bilateral cortical infiltration should be seen on imaging [4].

ABC is similar histologically to GCRG, particularly in its solid part, but it contains a substantial number of blood-filled spaces, a finding that is not present in GCRP [2, 14].

The solid variants of ABC have been reported in the jaw and in the short tubular bones of the hand, but reports in the long tubular bones are relatively rare [7, 11, 14, 19]. In a large series of 238 cases with ABCs [18], eight were of solid variant and only three were in the long bones. Ilaslan et al. [7] found 32 solid ABC lesions in long bones reported in the English literature and described the morphologic features and varied imaging findings in their 30 cases. A history of trauma was present in only three of their 30 cases. Four lesions had a partially mineralized matrix on plain radiographs or CT, with the remaining 26 being purely osteolytic. Periosteal reaction was observed in only three lesions. The MRI appearance suggested a solid mass except in two cases where mixed solid and cystic areas with fluid-fluid levels were observed. GCRG cases of the short tubular bones of the hands and the feet have a recurrence rate between 23% and 75%, but a better biologic behavior is expected when lesions are located in the long bones [5, 11, 12, 14, 16, 19]. In any case, complete curettage is the suggested treatment.

Our patient was treated by thorough curettage of the lesion and by application, into the empty bone cavity, of gauze saturated with pure alcohol for 10 minutes to destroy cells and blood vessels. The defect then was filled with a synthetic, biodegradable, calcium phosphate-based bone substitute. A forearm splint was applied for 8 weeks to protect the ulna from a pathologic fracture. The wrist was immobilized, but finger, elbow, and shoulder range of motion exercises were encouraged. The histologic examination of the sample taken during the definitive surgery confirmed the initial diagnosis. No local recurrence was observed 1 year after surgery (Fig. 7). Eighteen months after surgery, the patient has had no additional symptoms.

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