Pain and Deformity of the Index Finger in a 41-year-old Woman

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History and Physical Examination

A healthy 41-year-old right-hand-dominant woman presented with a chief complaint of slowly progressive deformity and pain in her right index finger. She reported a traumatic injury to her right index finger 5 years earlier, for which she never sought medical attention, and noted subsequent enlargement of the area. Two weeks before presentation, she experienced pain when a distraction force was applied to the involved digit. She denied any fever, chills, weight loss, or systemic illness. Her medical history otherwise was negative, including no personal history of malignancy or musculoskeletal disorders. Her social history was negative for tobacco, alcohol, or intravenous drug abuse.

Examination of her right hand revealed a fusiform deformity, swelling, and moderate, tenderness to palpation of her right index finger localized over the proximal

phalanx. Range of motion was full at the metacarpophalangeal joint. There was a 10° flexion contracture at the proximal interphalangeal joint with range of motion limited to 10° to 20° at the proximal interphalangeal joint and 0° to 40° at the distal interphalangeal joint. There were no significant angular deformity or rotational deformity of the digit, no pain or instability elicited on axial loading, and no overlying skin changes. Capillary refill was brisk and there were no sensory or motor deficits. Laboratory analyses, including complete blood count, serum chemistry panel, and coagulation panel, were within normal limits.

Radiographs of the right index finger were obtained on initial evaluation (Fig. 1).

Imaging Interpretation

Lateral (Fig. 1A) and anteroposterior (Fig. 1B) radiographs of the right index finger showed an expansile, lytic lesion involving the distal half of the proximal phalanx with cortical thinning, sclerosis along the proximal margin, and extension to the articular surface. There was no evidence of intralesional mineralization, cortical disruption, or fracture through the lesion. An MRI was not ordered, given the long-standing presence of the lesion, its relatively benign course, and its nonaggressive radiographic features.

Based on the history, physical examination, laboratory tests, and radiographic findings, what is the differential diagnosis?

Differential Diagnosis

Enchondroma
Unicameral bone cyst

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Each author certifies that all investigations were conducted in conformity with ethical principles of research, and that informed consent for participation in the study was obtained.

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Fig. 1A–B (A) Lateral and (B) anteroposterior views of the right index finger show an expansile, lytic lesion of the proximal phalanx with cortical thinning and marginal sclerosis. There is an absence of intralesional mineralization, cortical disruption, and fracture through the lesion.

Aneurysmal bone cyst
Giant cell reparative granuloma
Intraosseous ganglion cyst
Giant cell tumor
Osteoblastoma
Metastatic disease

The lesion was excised and histologic studies of the lesion were performed (Figs. 2–4).

Based on the history, physical examination, laboratory tests, radiographic findings, and histologic studies, what is the diagnosis and how should this lesion be treated?

Histology Interpretation

Intraoperative findings revealed a bluish cystic lesion with paper-thin cortices (Fig. 2). The resected surgical specimen consisted of an ovoid portion of bone with a yellow-tan external surface. Sectioning was achieved without decalcification, revealing a central cystic space containing serosanguinous fluid and flaky grey-white material. The entire specimen was decalcified, after which cross sections of the lesion were submitted for routine histologic

Fig. 2 An intraoperative photograph of the lesion shows its bluish, cystic appearance with paper-thin cortices.

Fig. 3 An histologic specimen examined in a low-power field shows marked thinning of the cortical bone with central acellular eosinophilic amorphous material (Stain, hematoxylin and eosin; original magnification, 2 $\times$).

processing. Sections were cut at 4 to 5 μ m and stained with hematoxylin and eosin and modified trichrome stains.

Microscopic examination showed marked thinning of the cortical bone (Fig. 3), with an accumulation of acellular eosinophilic amorphous material in the center of the specimen. The trichrome stain showed a pattern consistent with collagen blending into elements of woven bone. Elsewhere there was evidence of hemorrhage. Some of the material was smeared on a slide and examined with compensated plane polarized microscopy. The analyzed sample did not show birefringence. There was no organized cyst lining but rather a few layers of flattened fibroblasts and occasional histiocytes along the border of the cystic spaces (Fig. 4).

Fig. 4 An histologic specimen examined in a high-power field shows the cyst lining contains flattened fibroblasts with an occasional histiocyte. (Stain, trichrome; original magnification, 250 $\times$).

Diagnosis

Solitary (unicameral) bone cyst.

Discussion and Treatment

The differential diagnosis for our patient included enchondroma, aneurysmal bone cyst, giant cell reparative granuloma, intraosseous ganglion cyst, or less likely, osteoblastoma, giant cell tumor, or metastatic disease.

Enchondroma is a benign, cartilage-forming lesion of the metadiaphysis thought to arise from failure of physal cartilage cells to undergo apoptosis. Radiographically, enchondromas are well-circumscribed lesions with areas of mineralization that have classic “arcs and rings”, or popcorn-like appearance [21, 23]. In patients with multiple enchondromas (Ollier’s disease), the lesions can result in substantial disability, and the risk of malignant transformation to chondrosarcoma is 25%, whereas malignant transformation of a solitary enchondroma is rare [19]. In Maffucci syndrome, patients have multiple enchondromas, hemangiomas, and a nearly 100% risk of having skeletal or visceral malignancy develop [19]. The clinical, radiographic, and histopathologic distinctions between enchondroma and low-grade chondrosarcoma are not easily made. Pain, degree of endosteal scalloping, and location are important determining factors, as low-grade chondrosarcomas tend to be more symptomatic, have pronounced endosteal scalloping, and occur much more commonly in proximal locations. Chondrosarcoma more often is painful, has more intense radiotracer uptake on bone scintigraphy, and more cellular atypia than enchondroma [14]. Management of an asymptomatic solitary enchondroma is

observation. If the lesion becomes symptomatic, intralesional curettage and allograft bone packing are performed. In the hands and feet, fracture can occur through the lesion. In such cases, surgery should be delayed until there is radiographic evidence of healing, after a period of immobilization. In our patient, enchondroma was easily excluded based on the absence of chondrocytes in the resected specimen.

Aneurysmal bone cyst is an expansile, cortically based lesion with an unclear etiology, although trauma has been implicated in the surface variety. The lesion is mostly blood-filled, with fluid-fluid levels evident on MRI aiding in the diagnosis but not necessarily differentiating it from a unicameral bone cyst [12]. Histologically, an aneurysmal bone cyst contains multiple fibrous septae and large dilated vascular lakes surrounded by a giant cell-rich matrix with hemosiderin deposits [4]. Although cystic degeneration of other lesions could mimic unicameral bone cysts, the absence of vascular lakes or any true cyst lining, and the presence of amorphous eosinophilic material are all features supporting the diagnosis. Treatment consists of excision of surface aneurysmal bone cysts and curetting, burring, and thermal or chemical ablation of medullary lesions with or without internal fixation. Local recurrence is common, particularly when located in the spine. It is of paramount importance to differentiate an aneurysmal bone cyst from telangiectatic osteosarcoma, as wide excision and adjuvant chemotherapy are mandated for the latter.

Giant cell reparative granuloma (GCRG) is a benign, reparative lesion predominantly encountered in the skull, mandible, and facial bones, and also in the short tubular bones of the hands and feet [16]. As with an aneurysmal bone cyst, it most likely represents a response to traumatic intraosseous hemorrhage, and histologically, the solid variant of an aneurysmal bone cyst and GCRG are indistinguishable, both lacking the vascular lakes of a classic aneurysmal bone cyst [15]. Treatment is curettage and bone graft and recurrence is rare.

An intraosseous ganglion cyst is a subchondral lesion of tubular bones with expansile features, but can be excluded here on the basis of its typically viscous, gelatinous contents also found in its more common soft tissue counterpart [22]. Gross pathologic examination of our patient’s lesion revealed a serosanguinous low-viscosity liquid containing flaky-white material. Histologically, an intraosseous ganglion cyst is thick-walled with myxoid change, unlike the thin-walled septae of the unicameral bone cyst. The clinical course of intraosseous ganglion cyst, which most commonly is encountered in the carpus, is usually benign, most often presenting as an incidental finding or as mild, dull wrist pain [22].

A giant cell tumor is an eccentric, metaphyseal, radiolucent lesion with a geographic margin that abuts

subchondral bone. The most common locations are the distal femur, proximal tibia, and distal radius. Depending on the stage, a giant cell tumor can present with cortical thinning or breakthrough and may contain a soft tissue component. A giant cell tumor occurs most commonly in middle-aged persons and is more common in women. Although a giant cell tumor is a benign lesion, 2 to 6% of patients have pulmonary metastasis, especially associated with local recurrence [3]. A chest radiograph should be made on initial evaluation and radiograph or CT of the chest performed when evaluating a local recurrence. Treatment is intralesional or marginal to wide excision. The histologic appearance of a giant cell tumor includes multinucleated giant cells with nuclei that are identical to the nuclei of the surrounding stromal cells. Histologic examination excluded a giant cell tumor as the diagnosis in our patient.

An osteblastoma is a rare benign tumor of bone, accounting for less than 1% of all primary skeletal tumors [13]. The two most common locations, the proximal tibia and the posterior elements of the spine, account for almost half of all osteblastomas, but solitary and multifocal osteblastomas have been reported in the hand [1]. Radiographic appearance is variable, but usually consists of an expansive well-circumscribed lytic lesion with sclerotic margins. Microscopically, an osteblastoma is characterized by a cellular, highly vascularized stroma of immature bone surrounded by osteoblasts [1], and therefore is excluded in our patient.

Metastatic disease is extremely rare in such acral locations (0.1% of all skeletal metastatic lesions), and when it does occur, it is most commonly lung or renal carcinoma [7]. This diagnosis is unlikely given the insidious presentation of this lesion and the absence of systemic disease, and histologic examination failed to show such cells.

Enchondroma, giant cell tumor, giant cell reparative granuloma, osteblastoma, and metastatic disease were excluded by routine histologic examination, leaving the three cystic lesions in our differential diagnosis. An intraosseous ganglion cyst typically contains viscous material similar to its soft tissue counterpart, with a thick-walled lining sometimes showing myxoid change. Aneurysmal bone cysts contain multiple fibrous septae that may contain blood or appear empty and are surrounded by a giant cell-rich matrix. Ossification and dystrophic types of calcification are often present. Solitary or unicameral bone cysts contain serosanguinous fluid and have a thin membranous lining, with fibrin-like material and calcification that has been likened to cementum, a specialized calcified substance covering the root of a tooth. Histopathologic analysis in this case confirmed the diagnosis of unicameral bone cyst.

A unicameral or simple bone cyst is a benign, solitary lesion of bone that is overwhelmingly more common in the long bones of the skeletally immature, with a 2.5:1 male to female ratio reported [24]. The etiology remains unclear and even controversial but may represent a dysplastic process, possibly in response to trauma, or a local vascular abnormality causing venous obstruction. Jaffe and Lichtenstein categorized lesions as either active or latent, with active lesions favoring the area near the physis and latent lesions having a more diaphyseal location [9]. An involutional phase following pathologic fracture also has been described [24]. The classic “fallen fragment” sign representing fracture of the thin cortical margin into the cystic portion of the lesion, is characteristic, although not pathognomonic, of a unicameral bone cyst [17]. Greater than 90% of unicameral bone cysts are located in the proximal humerus or proximal femur [9, 11]. Other common locations include the proximal tibia, pelvis, calcaneus, and cuboid. Unicameral bone cysts also have been described in the lumbar spine, scapula, patella, metatarsals, and proximal fibula. The location of the lesion in our patient was unusual. We know of three other reported cases of unicameral bone cysts in the hand, two in skeletally immature patients and the other in a 22-year-old man [5, 8, 10].

The most common presentation of a unicameral bone cyst is pain after fracture through the lesion, or less commonly, as an incidental finding [24]. Prognosis is age-dependent, with recurrence or symptomatic lesions being more common when patient age at presentation is younger than 10 years [2]. There are two reported cases of malignant transformation, with Ewing’s sarcoma and chondrosarcoma arising from biopsy-proven unicameral bone cysts [6, 20].

Treatment approaches for unicameral bone cyst have included expectant management, curetting and bone grafting, and intralesional injection with methylprednisolone or saline. Scaglietti et al. introduced injection with methylprednisolone in the 1970s, reporting a low recurrence rate at 1 to 3 years followup, particularly in patients with more skeletal growth remaining [18]. However, this required multiple series of injections, and their results have not been consistently reproduced. More recently, treatment of unicameral bone cysts with trochar trephination, saline irrigation, and filling of the cyst cavity with either bone graft or bone graft substitute has yielded promising results [24]. Regardless of the treatment method, recurrence of the lesion remains the most frequently encountered complication. Lesions of the proximal femur and to a lesser extent those of the subfacet region of the calcaneus should be managed with surgery because of the high risk of pathologic fracture in these areas.

In our patient, because the lesion was progressively symptomatic and the degree of deformity had resulted in

severe limitation of motion at the proximal interphalangeal joint, we elected to perform staged reconstruction with a tricortical iliac crest bone autograft and proximal interphalangeal joint fusion 8 weeks after the index procedure. Alternatives for treatment may have included expectant management or intralesional curettage with allograft bone packing. The patient underwent excision with placement of a cement spacer and percutaneous pinning to maintain length of the resulting defect. Postoperatively, the patient wore a static metacarpophalangeal block splint and was allowed active metacarpophalangeal range of motion under the supervision of a therapist. At her final postoperative visit, the patient was asymptomatic, with a well-healed incision and full range of motion of her metacarpophalangeal joint. Radiographs revealed complete incorporation of the iliac crest graft and osseous union of her proximal interphalangeal joint. She had returned to work without restrictions.

Atypical radiographic features of a unicameral bone cyst in this patient include the subchondral extension of the lesion and the sclerotic margin. Although articular involvement is unusual for a unicameral bone cyst, reported cases include this finding [2]. The reactive sclerosis could be attributed to the patient's remote history of traumatic injury, and represent a healed fracture through the lesion. This case is unique in that it shows unicameral bone cysts can present in an older population and in less classic locations, resulting in substantial joint deformity and functional limitation.

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