Unicameral Bone Cyst with Epiphyseal Involvement: Clinicoanatomic Analysis

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Summary: Epiphyseal involvement of a unicameral bone cyst (UBC) is rare. This anatomic setting represents a distinct clinical and radiographic entity. This study reports a new case and analyzes the clinical and biological behavior of seven additional UBCs with epiphyseal involvement from the literature. We report the first successful treatment of this variant with methylprednisolone acetate. The average age was 20.1 years with a male to female ratio of 1.3:1. Anatomic location: proximal femur (4), proximal humerus (2), and proximal tibia (2). Both age and location were atypical when compared to the classic metaphyseal location. Radiographically, all lesions presented a characteristic involvement of the epiphysis and metaphysis in various proportions. The epiphyseal plates were judged closed versus open in 50%, respectively. Follow-up ranged from 9 months to 3 years. Six cases healed following a single curettage (three with and three without bone graft). There were no late complications of fracture, deformity, shortening, or avascular necrosis. Recurrence was 0%. No secondary procedures were required. We conclude the age, location, and radiographic appearance is atypical and diagnosis is difficult, but the biological behavior is less aggressive and the prognosis more favorable than the typical, metaphyseal UBC. Curettage with or without bone graft has a high success rate. We recommend aspiration and intralesional methylprednisolone as the initial management. We hypothesize that epiphyseal UBCs have a better prognosis than metaphyseal location alone due to the older age, atypical location, and the potential of the epiphysis to reossify. Key Words: Epiphysis—Unicameral bone cyst—Methylprednisolone acetate—Proximal humerus—Proximal femur—Benign bone tumor (childhood).

Unicameral bone cyst (UBC) is a benign lesion found in the long tubular bones of children and adolescents. Classically, they originate in the metaphysis, abut the growth plate, and later may involve the diaphysis. This results from the continued longitudinal bone growth of the epiphysis beyond the site of the cyst (11). UBCs rarely involve the epiphysis. Jaffee first described UBC, noting a single example of epiphyseal involvement in a 42-year-old man (11). Cohn reported three cases of UBC that had evidence of penetration of the physis at surgery (6). Other reports of epiphyseal involvement by UBC, including treatment and results, have been described (2,3,7,10,18,22). We have recently treated a solitary bone cyst with extensive epiphyseal involvement in the proximal femur of a 14-year-old boy. Treatment consisted of biopsy, aspiration, and injection of methylprednisolone acetate. Curettage or bone grafting was not performed. The purpose of this paper is to report this case and to review the literature.

A review of the literature has revealed a total of
seven cases of UBC with epiphyseal involvement (Table 1) (2,3,7,10,11,18,22). All of these reports have been of isolated cases (1942–1980). To our knowledge, this study is the first report to analyze the overall manifestation and behavior of unicameral bone cysts with epiphyseal involvement (EUBC). This paper presents a retrospective analysis of the above cases in order to answer the following questions:

1. What are the clinical and radiographic characteristics of UBCs with epiphyseal extension?
2. What is the clinical significance (biological behavior) regarding growth disturbance, deformity, recurrence, and avascular necrosis?
3. What is the appropriate clinical management of this rare variant?

**CASE REPORT**

A 14-year-old male complained of exercise-related pain in the right hip for 3 months not responsive to salicylates. There were no other musculoskeletal or constitutional complaints. There were no cutaneous abnormalities. Physical examination showed that both legs were of equal leg lengths without atrophy. The gait was normal; the Trendelenburg test was negative. The range of internal and external rotation on the right hip was limited by 30°. The remainder of the musculoskeletal examination was normal. Laboratory studies, including CBC, serum electrolytes, calcium phosphorus, and serum alkaline phosphatase were within normal limits.

**Radiographic Evaluation**

Radiographs of the right hip (Fig. 1) demonstrated an extensive radiolucent defect of the femoral neck and capital femoral epiphysis, partly circumscribed by a narrow margin of sclerosis. The cortex showed thinning but no expansion, cortical breakthrough, periosteal reaction or fracture. The femoral neck was of normal length without deformity. The acetabulum, pelvis, and joint space were normal.

A 99mTc-MDP bone scan (Fig. 2) revealed no increased uptake in the lesion, but a subtle decrease in the femoral neck. There was normal activity in the growth plates (signifying an open, active growth plate) without evidence or hyperemia or soft tissue extension.

Computed tomography (Fig. 3) showed a lucent, nonexpansile lesion of the femoral head and neck without extraosseous extension or matrix formation, with an internal absorption coefficient of fluid. The combined radiographic studies suggested a nonaggressive lesion. The preoperative differential diagnoses included atypical UBC, atypical chondroblastoma, giant cell tumor, unusual aneurysmal bone cyst, or eosinophilic granuloma.

**TABLE 1. Epiphyseal unicameral bone cyst: literature review**

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age/sex</th>
<th>Location</th>
<th>Relationship to epiphyseal plate</th>
<th>Status of plate</th>
<th>Rx</th>
<th>Status</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>11</td>
<td>42/M</td>
<td>Distal femur</td>
<td>—</td>
<td>Closed</td>
<td>—</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>6.5/F</td>
<td>Proximal tibia</td>
<td>Metaphyseal/epiphyseal</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>22/F</td>
<td>Proximal femur</td>
<td>—</td>
<td>Closed</td>
<td>Curettage; bone graft</td>
<td>Healed 3 years, no deformity</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>15.3/F</td>
<td>Proximal humerus</td>
<td>Metaphyseal/epiphyseal</td>
<td>Open</td>
<td>Curettage; bone graft</td>
<td>Healed 18 mos, no Fr</td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>24/M</td>
<td>Proximal humerus</td>
<td>Metaphyseal/epiphyseal</td>
<td>Closed (tomograms)</td>
<td>Curettage; no graft</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>17/M</td>
<td>Proximal femur</td>
<td>Epiphyseal</td>
<td>Closed</td>
<td>Curettage; bone graft</td>
<td>Healed 6 mos, asymmetric</td>
<td></td>
</tr>
<tr>
<td>Malawer and Markley (1982)</td>
<td>14/M</td>
<td>Proximal femur</td>
<td>Metaphyseal/epiphyseal</td>
<td>Open</td>
<td>Intrallesional methylprednisolone alone; no curettage or graft</td>
<td>FROM healed (80%) 2 years</td>
<td></td>
</tr>
</tbody>
</table>

*FROM, full range of motion.

Fr, fracture.

The femoral neck was approached through an anterolateral incision. An 18-gauge needle was inserted into the lesion, and clear, yellow fluid was aspirated. Manometric measurements showed 16 cm H₂O pressure. Pulsation of the column synchronized with the patient’s pulse. A Valsalva maneuver was performed, and a direct variation of the pressure was noted. Biopsy of the wall through a limited 0.5 x 0.5 cm window in the anterior cortex confirmed a cystic cavity with no underlying primary neoplasm.

Histologically, the lining showed islands of acellular eosinophilic material, interspersed by a thin fibrous stroma containing a few vessels (Fig. 4). There were no giant cells, histiocytes, or hemosiderin. The clinical and histological diagnosis of UBC was based on the finding of a cavity containing yellow fluid lined by a thin fibrous membrane. Curettage and bone graft were not performed. Methylprednisolone acetate, 240 mg, was instilled under direct vision via the cortical window, and the wound was closed. The dose was determined by the amount required to fill the cyst cavity.

RESULTS AND FOLLOW-UP

Postoperatively, the patient was maintained nonweight bearing for 6 weeks. Full weight bearing was begun at 2 months. Twenty-four months following surgery, he was fully ambulatory and participating in all activity. Examination of the hip showed a full range of motion and a normal gait.

Radiographic Evaluation

Radiographs were obtained at 2 months, 6 months, 8 months, 12 months (Fig. 5), and 18 months (Fig. 6) after surgery. Serial radiographs showed rapid thickening of the bony cortex and secondarily progressive centripetal ossification. Heterotopic bone formation in the abductor muscles was noted and considered secondary to the surgery. Twelve months after surgery, 80% of the defect had reossified. There was complete reossification of the epiphysis and remodeling of the calcar, with premature obliteration of the epiphyseal plate and subsequent femoral neck shortening (decrease articular trochanteric distance) but without deformity or avascular necrosis and with maintenance of a normal joint space (Fig. 6A).
FIG. 3. Computed tomography scans at the level of the capital epiphysis (A) and at the level of the femoral neck (B). Apart from a solitary central calcification, no osseous or cartilaginous matrix is present. There is no soft tissue mass. The cortex of the femur is thinned but intact. The joint space and acetabulum are normal. Note the surrounding zone of reactive sclerosis indicating a benign lesion.

Comment

In planning the treatment of this lesion, the preoperative radiographic evaluation was important in demonstrating the limits of the lesion and its nonaggressive character. The unusual, extensive involvement of the epiphysis made a specific diagnosis difficult, even though many of the lesion's characteristics suggested a bone cyst. Additional possible diagnoses included chondroblastoma, giant cell tumor, eosinophilic granuloma, aneurysmal bone cysts, or a well healed infection (21). The "cold" character on radionuclide bone scan argued against fibrous dysplasia (9). An intraosseous lipoma should have shown a typical low attenuation value by computed tomography. A bone cyst was ultimately diagnosed by the finding of a cystic cavity, containing clear yellow fluid, and lined by a thin fibrous membrane.

CLINICOANATOMIC ANALYSIS AND LITERATURE REVIEW

An extensive review of the literature noted seven cases in addition to the present case of UBC with epiphyseal involvement (Table 1). We analyzed the age, sex, location, treatment, rate of recurrence, and deformity in order to determine the clinical behavior and the appropriate management of this variant.

Age and Sex Distribution

Age at the time of diagnosis ranged from 6.5 to 42 years with an average of 20.1 years. Males outnumbered females 4:3. The average age was notably higher than those reported of classic metaphyseal lesions (1,2,4,10,14–16,19).
Location of Lesion

There were four (50%) femoral, two (25%) proximal humeral, and two (25%) proximal tibial lesions. This is an unusual distribution. Neer reported a reverse ratio of 2:1, proximal humerus:proximal femur, in a review of 250 simple bone cysts (17). He found the tibia to be a rare site (less than 4%). Two of eight cases of EUBCs involved the tibia.

Radiological Relationship and Status of Physis

All lesions (except Case 7) involved the metaphysis and epiphysis. The degree of involvement varied from predominantly metaphyseal to equivalent degrees of epiphyseal and metaphyseal involvement.

Four of the seven cases (Cases 1, 3, 5, and 6) were in older individuals whose epiphyseal plates were closed. Three reported cases (Cases 2, 4, and 7) in addition to our case (Case 8) had an open physis. In one case, the epiphyseal plate was obliterated by the lesion and the effects of old trauma.

Treatment and Results

Follow-up ranged from 9 months to 3 years. Results of the earliest case (Case 1) were not reported.

Six of the remaining cases reviewed were treated with curettage, three with and three without bone grafting. All lesions healed. Bone grafting did not offer any apparent advantage. There were no differences between those with open or closed epiphyses regarding final outcome, irrespective of treatment. There were no recurrences, late fractures, deformity, avascular necrosis, or secondary procedures required. No patient developed leg-length discrepancy. Humeral shortening in Case 5 was attributed to old trauma. Case 7, a femoral head lesion, presenting with subarticular collapse and early degenerative changes, was cured with curettage and bone graft. This was the only case of articular collapse. Thus, open epiphyseal plates did not appear to increase the incidence of late complications.

DISCUSSION

Unicameral bone cysts are classically metaphyseal and adjacent to the physis in origin but may later involve the diaphysis. Seventy percent will remain juxta-epiphyseal (17). The epiphysis is rarely involved. This study reports one new case of a UBC with epiphyseal involvement and retrospectively analyzed seven cases from the literature.

Clinical Considerations

The average age of patients with UBC in tubular bones is 9 to 15 years (1,3,4,10,14,16,19,20) with a male to female ratio of 2 to 3:1. In the present series, the average was older, 20 years, and the male to female ratio was decreased to 1.3:1. The proximal humerus and proximal femur (2:1) are the most common sites representing 75% of most large series. In contrast, the present series showed four femoral and two proximal humeral lesions, a reverse of the expected ratio. In addition, two patients (Cases 3 and 4) had proximal tibia lesions, whereas tibial lesions were much less common in other series. Neer noted that less frequent locations were characteristically found in adults (19). Boeseker et al. found no proximal tibia lesion in 145 simple UBC reviewed (3). He noted only one UBC with epiphyseal involvement in a review of the 145 UBCs seen at the Mayo Clinic during a 47 year period. Therefore, we concluded that the age, sex, and anatomic location of EUBCs are atypical and do not lead to a correct diagnosis from clinical or radiographic criteria alone. Surgery is necessary to establish the correct diagnosis. Aspiration is a simple diagnostic technique.

Biological Activity and Hemodynamic Considerations

Biological and hemodynamic correlates of "activity" in addition to radiological location have been described by Enneking (8) (1977). Latent cysts were characterized by a pressure on manometric measurement equal to venous pressure (6–10 cm H2O) with a nonpulsatile column. Our hemodynamic measurements correspond to an active cyst (16 cm H2O and pulsatile). This is the first reported pressure determination of an EUBC. Cohen (6) (1977) briefly reported three proximal femoral lesions with apparent perforation of the physis at surgery, but no further clinical or hemodynamic data were given. While one can speculate that increased cyst pressure may cause local erosion of the epiphyseal plate with subsequent epiphyseal extension, we suspect this is not the main determinant, since most active cysts (therefore increased pressure) (8) do not clinically involve the epiphysis. Epiphyseal involvement has been postulated to result from trauma to the
growth plate with secondary extension (18). This cannot be substantiated by this review. The pulsatile, elevated pressure measurement we obtained may suggest a vascular etiology. Previously venous obstruction had been proposed by Cohen (5,6). Johnson suggested an etiology related to a small angiomyxofibrillar lipoma (12).

Role of Methylprednisolone

Recurrence following curettage of UBC have ranged from 17 to 50% (17). Specifically, UBCs of the proximal femur are difficult to treat and often result in growth disturbance and pathologic fractures with subsequent coxa vara and ischemic necrosis (13). Boeseker (1968) noted a 28% recurrence rate of proximal femoral UBCs (3). Thus, we chose to treat our patient with aspiration and methylprednisolone as described by Scaglietti et al. (19) in order to avoid this anticipated high rate of morbidity and recurrence.

Scaglietti et al. (1979) reported 72 bone cysts treated by topical injection of methylprednisolone (19). Seventy-two percent of the lesions involved
the humerus or proximal femur. Radiographic findings showed reconstitution of the cortical bone of the cyst wall (similar to our patient). Progressive formation of new bone was noted at 6 months with opacification of the cyst cavity. Complete obliteration of the cavity took 3 years. The epiphysis, unlike in our patient, usually continued normal growth, and Scaglietti stressed that no damage to the epiphyseal cartilage resulted from injection of steroids. We hypothesize the premature closure of the epiphyseal plate in the present case was due to the healing process, i.e., reossification of the involved physis and epiphysis (Fig. 6).

Scaglietti noted in all cases with a positive radiological response that no fracture occurred. No patient required surgical treatment. Our patient similarly showed a good response, with early thickening of the cortical walls and opacification of the cavity with resultant early ambulation and maintenance of a good range of motion (Fig. 5).

CONCLUSIONS

Unexpectedly, all six reported cases in the present series healed with a simple curettage (with or without bone graft). More importantly, there were no late sequelae of fracture, deformity (except Case 7), or avascular necrosis. Joint destruction and recurrence was not a problem. Interestingly, the status of the physis (open or closed) did not influence the final outcome.

EUBC is a rare lesion and may be difficult to diagnose. The differential diagnosis should include chondroblastoma, giant cell tumor, eosinophilic granuloma, intraosseous lipoma, and aneurysmal bone cyst (21). We believe that the clinical behavior of a EUBC is to heal with simple curettage. The expected tendency for recurrence, deformity, and avascular necrosis is less than we anticipated. We hypothesize that these lesions have a better prognosis than metaphyseal location alone due to the older age of these patients (a good prognosticator in simple UBC), their atypical locations, and the increased potential of the epiphysis to reossify. We believe EUBCs represent a distinct clinicopathologic entity with a less aggressive behavior and a more favorable prognosis than the classic, metaphyseal UBC, although its etiology is unknown. Further hemodynamic evaluation of both the metaphyseal and epiphyseal variant is warranted and may elucidate an underlying mechanism of epiphyseal extension.

SUMMARY

This paper reports a new case of a unicameral bone cyst with epiphyseal involvement and reviews seven cases from the literature. The clinical and biological behavior were analyzed.

1. An exact radiologic diagnosis is difficult. Radiologic imaging aids by demonstrating a nonaggressive, solitary osseous lesion confined to bone. Aspiration is recommended as a simple confirmatory diagnostic technique.

2. UBC (epiphyseal variant) presents in an older age group, often after epiphyseal closure, without expansion or cortical destruction and involves both the metaphysis and epiphysis.

3. These lesions are less aggressive than the metaphyseal UBC and can be expected to heal with simple curettage, with or without bone graft.

4. There is a low morbidity following curettage. Postoperative fracture, deformity, and avascular necrosis did not occur.

5. We recommend intralesional methylprednisolone acetate following aspiration with protective bracing as the initial management and would reserve curettage for recurrent lesions or for diagnosis when aspiration is nonconfirmatory.

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REFERENCES


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