Bones of contention – Real and Imagined Differential Diagnosis

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No disclosure
CHAPTER 8
Differential Diagnosis I: Diseases Simulating Abuse
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Rickets
Congenital Indifference to Pain
Myelodysplasia and Other Neurologic Disorders
Osteomyelitis
Congenital Syphilis
Scurvy
Vitamin A Intoxication
Caffey’s Disease
Leukemia
Drug-Induced Bone Changes
Copper Deficiency
Menkes’ Syndrome (Kinky-Hair Disease)
Inherited Bone Dysplasias
Miscellaneous Conditions

CHAPTER 9
Differential Diagnosis II: Osteogenesis Imperfecta
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Deborah Krakow, M.D.
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CHAPTER 10
Differential Diagnosis III: Accidental and Obstetric Trauma
Paul K. Kleinman, M.D.

CHAPTER 11
Differential Diagnosis IV: Normal Variants
Paul K. Kleinman, M.D.
David S. Kwon, M.D.
Case 1

1 month-old previously healthy boy
Right lower limb pseudoparalisisi
Final diagnosis?
Rickets

Osteomalacia before fusion of the growth plate.
NO Osteoporosis

Imaging:
Loss of the zone of provisional calcification with apparent physeal widening. Unossified cartilaginous rests cause a frayed appearance of the subphyseal metaphysis. Flaring, cupping and splaying of metaphyses as well as demineralization.
Rickets

Physeal widening
Fraying, flaring, cupping and splaying meatphysis
Symmetrical and systemic
Rickets

Fractures in rickets
In association with abnormal bone radiographs

Transverse long-bone fractures, metaphyseal fractures (not CML), anterior-lateral rib fractures

9 mo nutritional rickets. Distal radius and distal ulna transverse fractures

11mo rachitic rosary and rib fractures

Rickets of prematurity is common in babies of fewer than 28 weeks’ gestation and who weigh less than 1.5 kg (3.3 lbs).

The radiographic findings in metabolic bone disease of prematurity are generalized osteopenia and fractures.

Risk factors associated with bone loss and rickets: (1) cholestatic jaundice, (2) prolonged total parenteral nutrition (longer than 3 weeks), (3) bronchopulmonary dysplasia, and (4) prolonged diuretic therapy with furosemide.

Rickets of prematurity and fractures

Fractures most common in infants weighing less than 1500 g. Fractures usually involve the ribs and long bones, with a predilection for the metaphyseal regions.

Osteopenia but no specific radiologic evidence of rickets

3-month-old former premature infant. From Dr Kleinman’s book
1 do
Esophageal at
3 months old
Postoperative NEC
Congenital indifference to pain (CIPA)

Fractures and epiphyseal separations in various stages of repair
Osteomyelitis in young infants may cause multifocal metaphyseal lesions with SPNBF. Systemic signs and symptoms are often lacking.

Distinction from traumatic fracture must be made on clinical grounds.

2 months follow up
Leukemia

4 mo boy with acute respiratory distress
Hepatoseplenomegaly
Leucoeritroblastosis
Menke’s syndrome

Defective gastrointestinal absorption of copper. Long bone metaphyses have spurs with or without fracture. SPNBF may be present, and osteopenia is frequent after 6 months of age.
Osteogenesis imperfecta is a generalized disorder of connective tissue. The disease in bone and other tissues has a large range of phenotypic expression.
Neonate 36 weeks gestation
C section delivery
Craniotabes?
Case 2

2 month’s old girl
Limb pain
Physiological periosteal reaction
Normal variants

• Physiological periosteal reaction

- 1 - 5 months
- < 2mm
- Double cortex
- Bilateral
- Femur and tibia

Quigley AJ. Skeletal survey normal variants, artifacts and commonly misinterpreted findings not to be confused with non-accidental injury. Pediatric Radiology (2014) 44:82-93
Normal variants

- “Step-Off”
- Proximal tibia cortical irregularity
Normal variants

- “Beak”
- “Step-Off”
- “Physiological periosteal reaction”

Quigley AJ. Skeletal survey normal variants, artifacts and commonly misinterpreted findings not to be confused with non-accidental injury. Pediatric Radiology (2014) 44:82-93
Normal variants

“Beak”

“Spur” (lateral distal femur, distal radius and ulna)
Normal variants

“cupping” distal ulna

Quigley AJ. Skeletal survey normal variants, artifacts and commonly misinterpreted findings not to be confused with non-accidental injury. Pediatric Radiology (2014) 44:82-93
Bones of contention – Real and Imagined Differential Diagnosis
Child Abuse
Child abuse: We have problems

Peter J. Strousse
“Child abuse exists. This is the paramount problem.”
The estimated number of deaths from child abuse in the United States in 2013 was 1,520

Denialism of child abuse is a huge problem, and it is getting worse

Tactics of denialists:
1. Manufacture doubt
2. Identify alleged conspiracies
3. Create impossible expectations of research
4. Use false experts
5. Misrepresent logical fallacies
6. Selectively cite the literature
86 children with unexplained fractures during 14 years.

“Several clear patterns emerged:
1. Osteogenesis Imperfecta
2. Newly recognized group of temporary brittle bone disease (*)
3. Vitamin D deficiency rickets
4. Menkes’ syndrome”

(*)(...)Over the years, I have seen several children referred as cases of osteogenesis imperfecta but having a very atypical history. They had a severe disease, including rib fractures and long bone fractures, in the first 12 months of life, but no fractures at all in later childhood (...

- Medical records were reviewed in 33 infants who were referred for consultation for multiple unexplained fractures in which the parents and other caregivers denied wrongdoing
- In 9 of the infants, radiographic absorptiometry and/or computed tomography bone density studies were performed
- In 25 of them there was a history of decreased fetal movement and/or intrauterine confinement
- In 26 of these infants the diagnosis of temporary brittle bone disease was made
- The results also demonstrate the usefulness of bone density measurements in evaluating the infant with multiple unexplained fractures to help distinguish nonaccidental injury from intrinsic bone disease
Infants less than 6 months of age.
1. Multiple asymptomatic metaphyseal lesions (particularly along the medial aspect of the distal femurs and proximal tibiae as well as in the distal tibiae and fibulae)
2. Pseudodiastasis of the sutures
3. Transverse lucencies through the forearms and ribs
4. Compression fractures of the spine
(...)

Case 4 A 2-month-old girl (Fig. 4) with an African-American mother and Caucasian father presented with a viral respiratory illness. A chest radiograph demonstrated healing rib fractures. Skeletal survey was interpreted as six, possibly eight, fractures highly specific for nonaccidental trauma. The baby had skull and cervical spine fractures. MRI demonstrated small old subdural hemorrhages (...).

The mother’s vitamin D level was 17 ng/ml
The etiology and significance of fractures in infants and young children: a critical multidisciplinary review

Sabah Servaes 1 · Stephen D. Brown 2 · Arabinada K. Choudhary 3 · Cindy W. Christian 4 · Stephen J. Done 5 · Laura L. Hayes 6 · Michael A. Levine 4 · Joëlle A. Moreno 7 · Vincent J. Pulcini 8 · Richard M. Shore 9 · Thomas L. Slovis 10

Consensus statement, supported by the Child Abuse Committee and endorsed by the Board of Directors of the Society for Pediatric Radiology


1. The reported cases
2. Under-reporting of abuse
3. Medical evaluation
4. Laboratory evaluation
5. Vitamin D
   1. <20 ng/ml deficiency
   2. >=30 ng/ml sufficiency
   3. 20-29 ng/ml insufficiency

- Significant metabolic bone disease is not associated with vitamin D levels of 20 ng/ml or greater
- No study has demonstrated that low serum vitamin D level increases susceptibility to bone fractures.
- Levels of 20–29 ng/ml do not result in skeletal fragility
CONCLUSION. The hypothesis that classic metaphyseal lesions are secondary to child abuse is poorly supported. Their histologic and radiographic features are similar to healing infantile rickets. Until classic metaphyseal lesions are experimentally replicated and independently validated, their traumatic origin remains unsubstantiated. (...)

Comment in
Commentary on "a critical review of the classic metaphyseal lesion: traumatic or metabolic?. [AJR Am J Roentgenol. 2014]
SPR Child Abuse Committee Response regarding classic metaphyseal lesion. [AJR Am J Roentgenol. 2014]
Classic metaphyseal lesions. [AJR Am J Roentgenol. 2014]
Reply: To PMID 24370143. [AJR Am J Roentgenol. 2014]
Reply: SPR Child Abuse Committee response regarding classic metaphyseal lesion. [AJR Am J Roentgenol. 2014]

CONCLUSION. The hypothesis that classic metaphyseal lesions are secondary to child abuse is poorly supported. Their histologic and radiographic features are similar to healing infantile rickets. Until classic metaphyseal lesions are experimentially replicated and independently validated, their traumatic origin remains unsubstantiated.
Beverly P. Wood. AJR:202, January 2014:

(...)Ayoub et al., have the potential to negatively affect the welfare of a group of vulnerable children and infants, whose interests pediatricians and caregivers are committed to defend (...).


(...)Scholarly critique of existing evidence is essential, but exclusion of key literature does not constitute healthy scholarship. Given the stakes involved, we think that the approach of Ayoub et al. is less “critical” than dangerous and that children and families deserve better(...)


(...) I encourage other investigators to study classic metaphyseal lesions with sufficient rigor and scholarship to further clarify the morphology and the biomechanics of these distinctive inflicted fractures.(...)
Inclusion criteria:
The medical examiner determined that the infant had sustained a head injury and that the manner of death was a homicide, at least one CML was evident at skeletal survey, CMLs were confirmed at autopsy, and non-CML fractures were also present.

46 consecutive infant fatalities. RADIOLOGIC AND PATHOLOGIC REVIEW

There were no radiographic or pathologic features of rickets in the cohort.
Evaluating Children With Fractures for Child Physical Abuse
Emalee G. Flaherty, Jeannette M. Perez-Rossello, Michael A. Levine, William L. Hennrikus, and the AMERICAN ACADEMY OF PEDIATRICS COMMITTEE ON CHILD ABUSE AND NEGLECT, SECTION ON RADIOLOGY, SECTION ON ENDOCRINOLOGY, SECTION ON ORTHOPAEDICS and the SOCIETY FOR PEDIATRIC RADIOLOGY
Pediatrics 2014;133:e477; originally published online January 27, 2014;
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Thank you

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