

Answer to this month's Radiological Conference

Answer:

b) Caffey's disease

Radiological findings

The initial radiographs taken when the baby first presented with leg swelling at 5 months of age show thick well-defined periosteal reaction involving the anteromedial aspect of the diaphysis of the tibia. The metaphysis and the epiphysis are relatively spared. No underlying bony erosion or destruction can be seen. No associated soft tissue mass is detected (**Figures 4 and 5**). Subsequent radiograph taken when the baby was 8 months old shows resolving periosteal reaction with a laminated appearance (**Figure 6**).

Figure 4: Frontal view of right lower limb taken when the baby was 5 months old with addition of an arrow. Well-defined thick smooth cortical thickening involving the diaphysis (arrow) with relatively sparing of the metaphysis (arrowhead)



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Figure 5: Lateral view of right lower limb taken when the baby was 5 months old with addition of an arrow. Anterior cortical thickening with no associated soft tissue swelling or underlying bony destruction was seen (arrow)



Figure 6: Frontal view of right lower limb taken when the baby was 8 months old with addition of an arrow. Cortical thickening which appears slightly decreased in size and laminated (arrow)



Discussion

Caffey's disease

Caffey's disease (Infantile Cortical Hyperostosis) is an uncommon disease which almost always affects infants less than 6 months of age. The exact etiology is unknown but viral infection, immune disorder and genetic predisposition have been proposed to be responsible for this disorder. An autosomal dominant pattern of inheritance with incomplete penetrance and variable expressivity has been identified. Sporadic cases are less common. Familial cases are the major pattern of this disorder nowadays.

Clinically, patients have fever of acute onset, hyperirritability and tender soft tissue swelling. The clinical course is variable, and usually the clinical and radiological features subside in a period ranging from a few months to a few years. Occasionally, it can recur or persist as an active disease, thus causing a delay in skeletal development. Complications are uncommon and include bowing deformity and osseous bridging of adjacent bone. Investigations may show raised erythrocyte sedimentation rate, alkaline phosphatase and white cell count.

Single or multiple bones may be affected. Mandible, clavicles, ulnae and ribs are common sites. Other tubular bones such as tibiae, fibulae, femora, radii, metacarpals and metatarsals are less commonly involved. Vertebrae and phalanges are rarely affected. Symmetric and sequential involvement are usual, although tubular bones and scapulae tend to have asymmetric involvement. The cortical hyperostosis typically affects the diaphysis with sparing of the epiphysis. During healing, the cortical thickening appears laminated with increased porosity. The radiograph in the appropriate clinical setting is usually diagnostic. Isotope bone scan is sometimes used to document the multiple sites of involvement. Magnetic resonance imaging can provide better delineation of the bony and soft tissue involvement but usually does not offer additional benefit in the management.

The differential diagnosis for bilateral symmetric periosteal thickening includes scurvy, leukaemia, neuroblastoma, hypervitaminosis A and long term administration of prostaglandin E_1 . Rickets and scurvy can be differentiated by their metaphyseal and epiphyseal changes. Hypervitaminosis A is rare in infants less than

1 year of age. Differential diagnosis for marked periosteal reaction affecting a single bone includes osteomyelitis, non-accidental injury and malignant tumours such as Ewing's sarcoma and osteogenic sarcoma.

Ewing's sarcoma

Ewing's sarcoma rarely affects infants, being more common in the first and second decades. Ewing's sarcoma may have laminated periosteal reaction simulating Caffey's disease, but lytic or permeative bony destruction in the meta-diaphyseal region is rare in Caffey's disease. Pure periosteal thickening without bony destruction and the very young age of patient are features against Ewing's sarcoma.

Osteomyelitis

Osteomyelitis typically affects the metaphysis in children. In infants, it can spread to the epiphysis through the transepiphyseal vessels that cross the growth plate. The periosteal reaction can be prominent and extends to the diaphysis. Osteolytic destruction is common. The diaphyseal location of the periosteal thickening and absence of bony destruction make osteomyelitis less likely.

Leukaemia

Acute childhood leukaemia usually affects children between 2 and 5 years of age. About 80% are lymphoblastic in origin. Isolated periostitis due to cortical involvement by tumour cells is infrequent. More common radiological findings are diffuse osteopenia, osteolytic destruction and radiolucent and radiodense metaphyseal bands. Metastatic neuroblastoma may produce an identical appearance.

Non-accidental injury

Non-accidental injury is the third most common cause of death in children. The child is usually less than 2 years old. Multiple asymmetric fractures in different stages of healing are typical. Both shaft or metaphyseal fractures can occur, although metaphyseal corner fracture due to a twisting injury is more characteristic. Unusual sites of fracture such as the scapulae, hands, feet and

spine; and unusual severity of injuries, for example, depressed or multiple skull fractures should raise the suspicion of non-accidental injury. Non-accidental injury tends to have extensive periosteal reaction due to subperiosteal haemorrhage simulating Caffey's disease. Other differentiating features include presence of fractures and extension of periosteal reaction to the epiphysis, both of which are usually present in non-accidental injury but not in Caffey's disease. ■

Further readings

1. Resnick D. *Diagnosis of Bone and Joint Disorders*. Philadelphia: WB Saunders; 1995;2247-2258,2326-2365,4436-4442.
2. Chapman S, Nakielny R. *Aids to Radiological Differential Diagnosis*. 3rd ed. London: WB Saunders; 1995;38-41.
3. Dahnert W. *Radiology Review Manual*. 3rd ed. Baltimore: Williams and Wilkins; 1996;35-36,74.
4. Yochum TR, Rowe LJ. *Essential of Skeletal Radiology*. 2nd ed. Baltimore: William and Wilkins; 1996;1193-1204.
5. Eisenberg RL. *Clinical Imaging- an Atlas of Differential Diagnosis*. Philadelphia- New York: Lippincott-Raven; 1997;748-757.