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Clinical History:

A 4-month-old baby boy was noted to be irritable and was thriving poorly. Imaging included radiographs of both upper limbs (Figures 1 and 2).

Figure 1: Frontal radiograph of the left upper limb

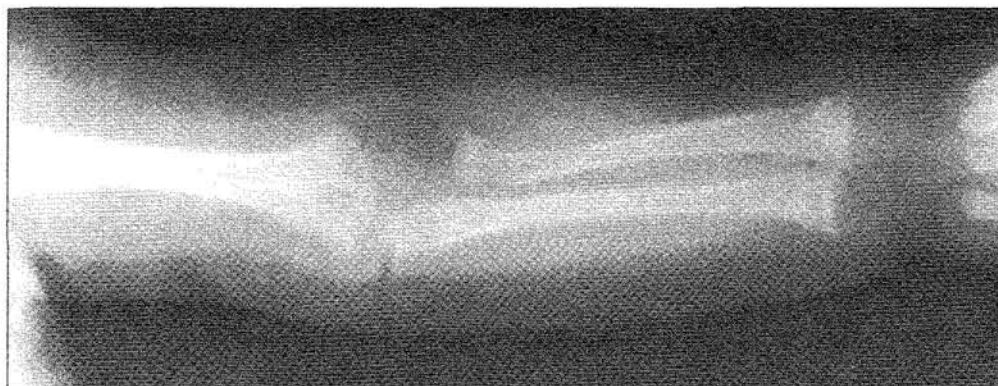


Figure 2: Frontal radiograph of the right upper limb and right upper chest



Answer
on
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What is the diagnosis?

- a) Rickets
- b) Scurvy
- c) Acute leukaemia
- d) Caffey's disease
- e) Non-accidental injury

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Answer:

- a) Rickets

Radiological findings

These radiographs show abnormalities in the metaphyses, epiphyses and diaphyses of the bones of all four limbs:

- **Metaphyses:**

There is fraying, splaying and cupping of the metaphyses of the long bones (**Figures 3–5**). Similar deformities of the anterior rib ends give rise

to the typical “rachitic rosary” (**Figure 4**). Cortical spurs are seen projecting at right angles to the metaphyses to surround the uncalcified growth plates.

- **Epiphyses:**

The growth plates are irregular and widened. The epiphyseal centres are poorly mineralized (**Figures 3–5**).

- **Diaphyses:**

Generalized periosteal new bone formation due to ossification of subperiosteal osteoid is present. This appearance only develops with treatment. The underlying cortex is indistinct (**Figures 3–5**).

Figure 3: Figure is identical to Figure 1 with addition of arrows. There is splaying and cupping of the metaphyses of the distal radius and ulna, with thin metaphyseal spurs (curved arrows). The metaphyses are frayed (small white arrows). The growth plate at the wrist is widened. Generalized periosteal reaction is present (small back arrows)

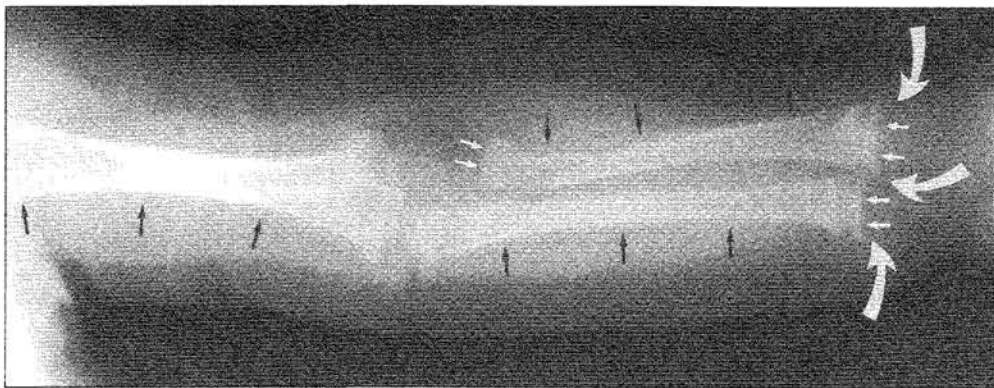
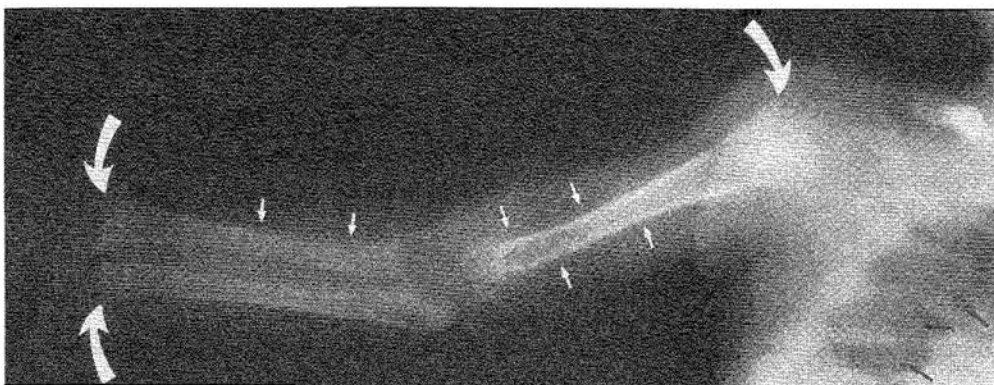
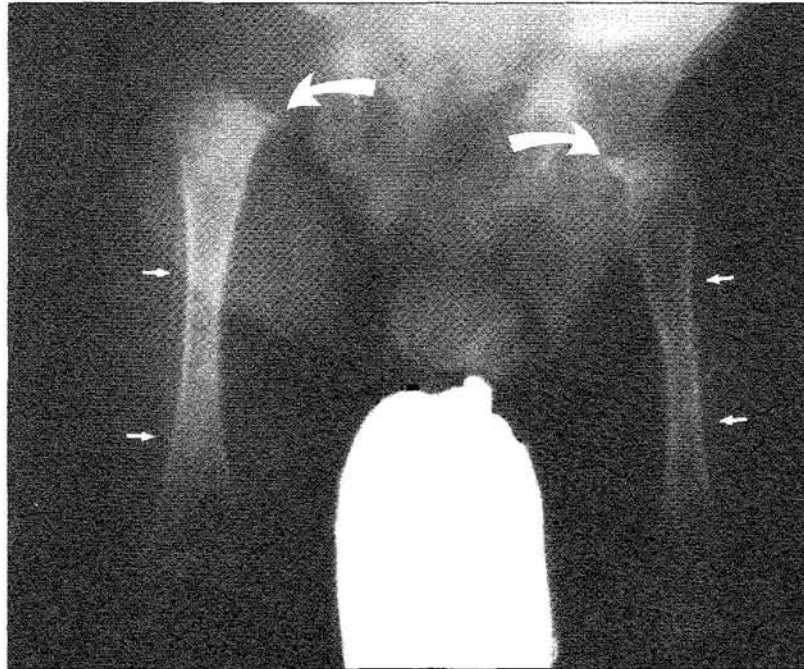


Figure 4: Figure is identical to Figure 2 with addition of arrows. Metaphyseal splaying, cupping and spurs (curved arrows) are best seen at the proximal humerus and distal forearm bones. Periosteal reaction affects the shafts of all the long bones (small white arrows). Rachitic rosary deformities of the anterior rib ends are arrowed (small black arrows)



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Figure 5: Frontal radiograph of both femurs. Metaphyseal widening and spurs (curved arrows), as well as generalized periosteal reaction (small white arrows), are seen



Discussion

Rickets is the childhood form of osteomalacia which occurs during enchondral bone growth. The radiographic appearances are due to increased uncalcified osteoid in the immature skeleton. Early changes include indistinctness of the metaphyseal margin, progressive fraying, and widening of the growth plate due to lack of normal calcification in the zone of provisional calcification. Bones subjected to stress such as the ankles, knees and wrists, are particularly involved. The uncalcified osteoid which is produced is deposited in the subperiosteal location, causing elevation of periosteum and indistinct cortical outlines. With treatment, mineralization of the subperiosteal osteoid gives rise to the bone-in-bone appearance seen in our patient.

Apart from those described above, other findings include flattening and invagination of skull base due to bone softening (craniotabes), delayed dentition, scoliosis and biconcave vertebral bodies. Bowing of long bones in lower limbs usually occurs after the onset

of weight-bearing. The various possible causes of rickets are summarized in the table.

Table 1: Causes of rickets

I. Abnormal vitamin D metabolism:

1. Deficiency: dietary, malabsorption, lack of sunshine
2. Defective conversion to active form due to hepatic or renal disease

II. Abnormal phosphate metabolism:

1. Phosphate deficiency
2. Renal tubular disorder
3. Oncogenic hypophosphataemia
4. Hypophosphatasia

III. Calcium deficiency:

1. Dietary deficiency
2. Malabsorption
3. Calcium chelates

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Scurvy

Scurvy is due to the deficiency of vitamin C, with resulting defective collagen and bony matrix formation. The age of onset is usually at 6-9 months, as maternal vitamin C lasts for the first 6 months of life. Clinically, there is tenderness and weakness of the lower limbs, as well as gum bleeding.

The typical signs of scurvy are:

1. Wimberger's sign: small epiphysis with loss of density and a very thin, sharply-marginated rim.
2. Frankel's white line: a dense white line at the growing metaphysis due to excessive calcification of osteoid in the zone of provisional calcification.
3. Trümmerfeld zone: a lucent metaphyseal band beneath the zone of provisional calcification due to lack of mineralized osteoid.
4. Pelkan spurs: metaphyseal spurs projecting at right angles to the shaft axis.
5. Parke corner sign: metaphyseal corner fractures through the weakened lucent metaphyses.
6. Periosteal reaction: subperiosteal haematoma with calcification of elevated periosteum.

The age of onset and the absence of the first three signs in our patient excludes the diagnosis of scurvy.

Acute leukaemia

Children with acute leukaemia are usually very ill with signs such as fever, splenomegaly and lymphadenopathy. The radiographic features of bony involvement are due to leukaemic infiltrates in various parts of the bone.

1. Metaphyseal translucency is the most characteristic sign. This is a transverse radiolucent band of up to 5 mm in width, crossing the metaphysis at rapidly growing areas such as the knees, wrists, and ankles.

It is due to rapidly proliferating leukaemic cells in the marrow-containing metaphysis.

2. Metaphyseal cortical erosions occur at the medial side of the proximal humeral and tibial shafts and are usually bilateral.
3. Diffuse demineralization is seen in the long bones in 30% of cases. Destruction of fine trabeculae causes the appearance of trabecular coarsening. Involvement of spongiosa and later the cortex causes multiple ovoid osteolytic lesions.
4. Periosteal reaction is smooth, lamellar or sunburst in appearance due to subperiosteal penetration by sheets of leukaemic cells.
5. Sclerotic lesions are seen late in the disease due to reactive osteoblastic proliferation.

Unlike rickets, leukaemic infiltration is usually asymmetrical and does not have a generalized pattern. The lesions are due to bony destruction rather than defective mineralization, and are therefore more irregular and ill-defined.

Caffey's disease (Infantile cortical hyperostosis)

Caffey's disease is a self-limiting entity manifesting as asymmetrical hyperostosis affecting the diaphyses of tubular bones, with sparing of the metaphyses and epiphyses. The distribution of bony involvement thus readily differentiates this condition from rickets. The age of onset is less than 6 months, with a mean of around 9 weeks. Deeply situated tender soft tissue swellings are associated with cortical changes in the underlying bones. Soft tissue swellings appear before the bony changes and may disappear long before bony resolution. The sites of involvement, in decreasing order of frequency, are the mandible, clavicle, ulna and then other sites. The phalanges and vertebrae are usually spared.

Non-accidental injury

The diagnosis of non-accidental injury (or child abuse) should be suspected in any child presenting with

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multiple bone fractures. The most specific fracture types are metaphyseal fractures of the long bones and rib fractures, with 80% of fractures occurring in children less than 18 months old. Unlike rickets, the metaphyses in the abused child are sharply defined and fractures usually occur in bones with normal density. The periosteal reaction is localized around the fracture site while it is generalized in rickets. In non-accidental injury, fractures and other injuries in the body are typically of different ages with poor correlation to a history of injury. ■

References

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