

3D volume rendered CT image of the right femur
History: 12 year old boy with right leg pain

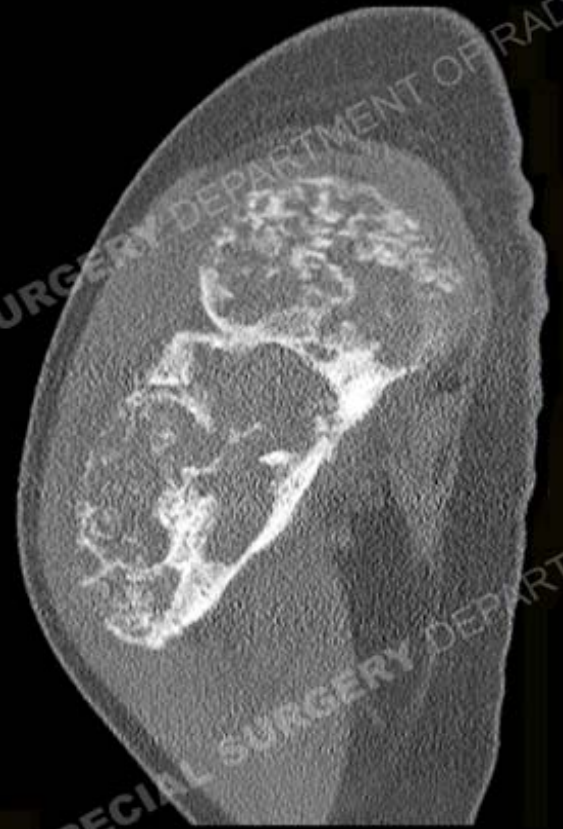




Both radiographs obtained on 9/5/12



Two representative axial and one representative coronal CT image of the underlying osseous architecture



Coronal and sagittal CT images through the area of focal clinical interest



Radiograph of 2010



Radiograph of 2012

Findings

Both the radiographs and CT demonstrate a diffuse process of the bone with expansion, remodelling, and abnormal attenuation/architecture of the bone. In particular, a shepherd's crook deformity is seen of the right femur with other areas of "hazy lucent" or ground glass opacity on the radiographs. On the CT, internal trabecular type architecture is seen with focal areas of a calcified type matrix and other areas of dense sclerosis. When comparing the sets of radiographs, there has been distraction of the right proximal femoral shaft with new cortical disruption. Extensive postoperative changes are seen of the left lower extremity.

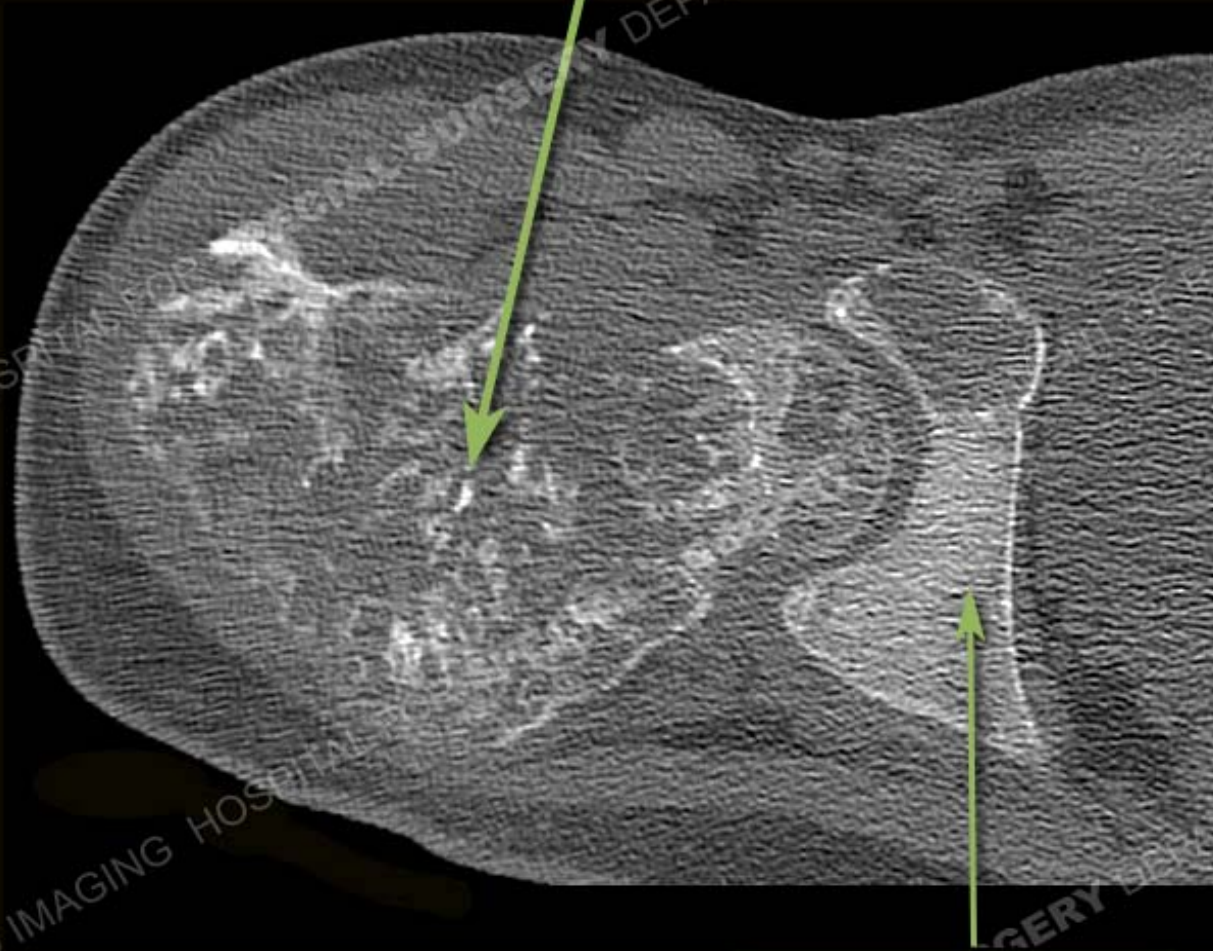


Shepherd's crook deformity

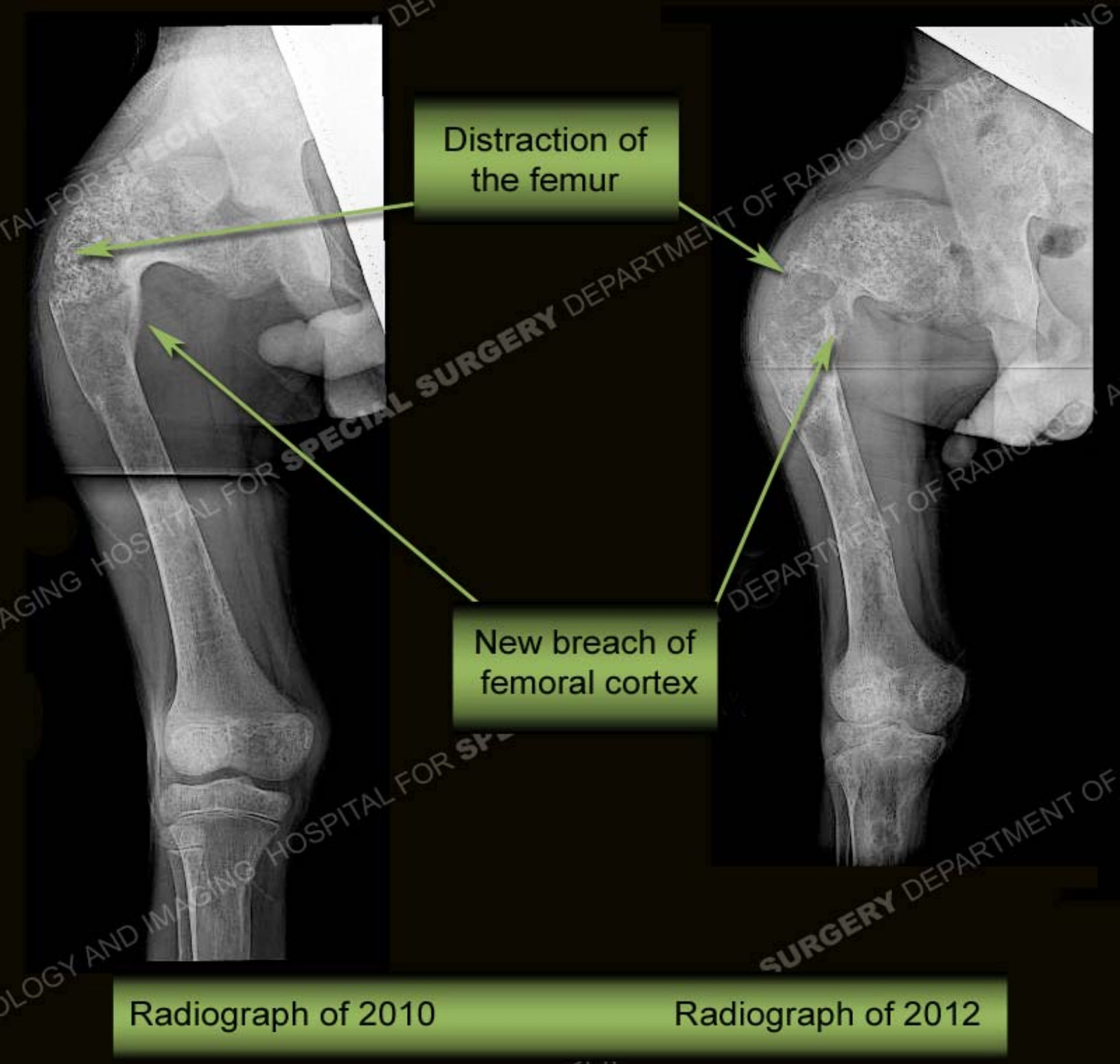
Ground glass opacification



Marked expansion of the bone with internal, trabecular type architecture



Marked sclerosis



Distraction of the femur

New breach of femoral cortex

Radiograph of 2010

Radiograph of 2012

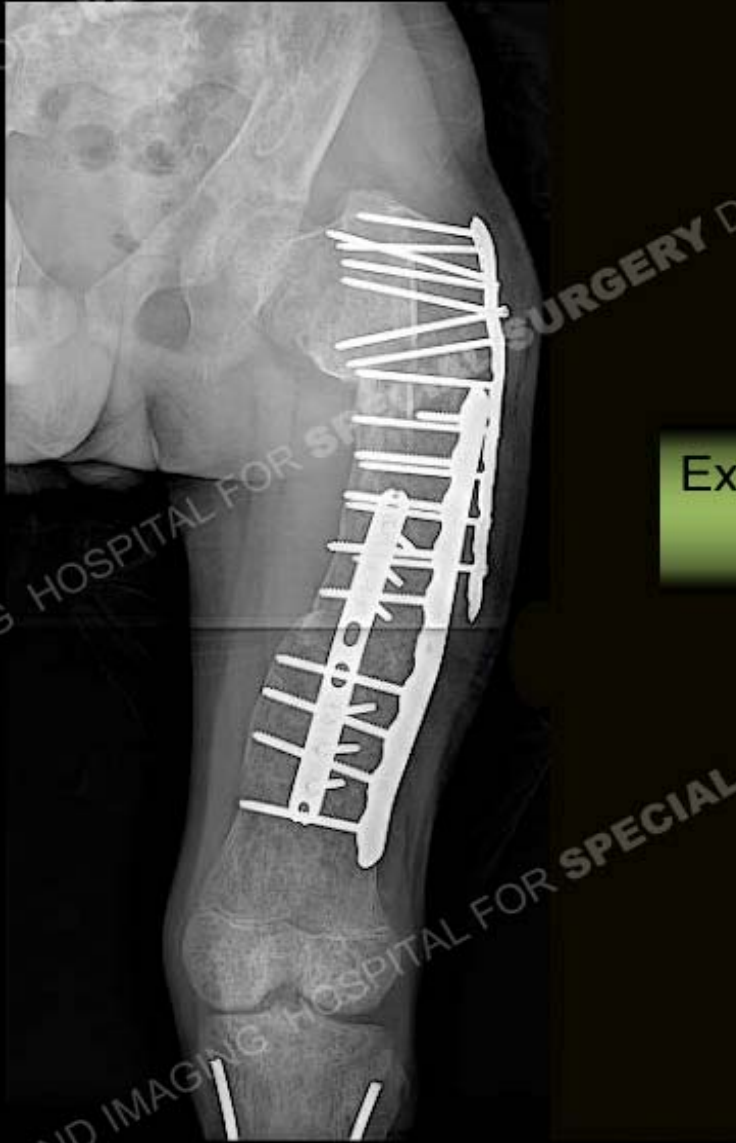
Breach of the cortex



Diagnosis: Polyostotic fibrous dysplasia with pathological fracture

- Fibrous dysplasia is a noninherited abnormality of the bone forming mesenchyme where osteoblasts do not mature normally and normal bone is replaced by immature, woven bone and fibrous tissue. This accounts for the areas of abnormal density of the bone inclusive of the ground glass, trabecular type architecture, and sclerosis. Although the ground glass architecture is classic, there can be an array of abnormal appearance of the bone as in this case. This abnormal bone is structurally weak allowing for expansion, remodelling, and pathologic fracture as in this case.
- Other complications such as malignant degeneration are exceedingly rare with precocious puberty (McCune Albright syndrome) and intramuscular myxoma (Mazabraud syndrome) being more common albeit still rare associations. Even when a discrete fracture is not present, the patient may have skeletal pain thought to be related to bony remodelling or underlying endocrine abnormalities. As seen in this case, multiple fractures may occur requiring multiple sites of fixation.





Extensive fixation for prior left femur fracture



IM rods of left tibia without a clear previous fracture. Fracture may have completely remodelled or alternatively rods may have been placed for pain/prophylaxis.

Resources:

Resnick and Kransdorf. Bone and Joint Imaging. 2005.

