**Spondyloepiphyseal Dysplasia (SED)**

By Hospital For Special Surgery [2]

**Clinical History:** An 82-year-old man with longstanding bilateral hip pain. Patient has prior cervical spine surgery and no history of previous trauma.
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Figures 2, 3

Figure 4: Neutral lateral radiograph; Figure 5: Flexion lateral radiograph
Figure 6: T1 Sagittal; Figure 7: T1 Sagittal; Figure 8: IR Sagittal
Figure 9: T2 axil at C1/C2 junction; Figure 10: T2 Sagittal

Figures 11 and 12: Sagittal reformations of CT study about the midsagittal line
**Findings:** Radiographs of the pelvis demonstrate markedly abnormal/dysplastic femoral heads with severe arthrosis of both hips, remodelled acetabulae, and foreshortened femoral necks with coxa vara deformity. Cervical spine radiographs demonstrate previous occipito-cervical fusion with poor delineation of the upper cervical spine architecture. However, noted is a marked widening of the atlantodental interval when comparing the neutral to the flexion views. Cross sectional imaging of the cervical spine demonstrates decreased height of the vertebral bodies with increased AP diameter and prominent end plate irregularities. There are multiple non-fused ossification centers seen at the tip of the dens and with an overall decreased amount of bone at the dens than typically seen. Severe stenosis is seen at the C1/C2 junction with severe compression of the cord and high signal within the cord representing edema/myelomalacia. Underlying developmental spinal stenosis is also seen.
Figure 16: Deformed/Dysplastic femoral heads with severe arthrosis; Foreshortened necks with coxa vara

Figures 17 and 18: Marked widening of atlantodens interval with flexion; Previous posterior fusion; Decreased height, increased AP diameter and irregular endplates of vertebrae
Figures 19, 20: Non-fused multipartite tip of the dens with overall hypoplasia of tip of the dens; Abnormal vertebrae as described previously

Figures 21, 22, 23: Kinking or severe stenosis with compression of the cord at C1/C2
Figures 24, 25: Severe stenosis of central canal with severe compression of the cord

Diagnosis: Spondyloepiphyseal Dysplasia (SED)

Discussion: SED is an inherited dysplasia that involves the ends of the bones or epiphyses and the spine. It comes in two variants, congenita (present at birth) and tarda which has a normal appearance at birth and then develops at 4 years of age and older. Given the underlying dysplasia there is premature osteoarthritis which in this patient may have been neglected. In the spine, there is typically a hypoplastic dens which leads to spinal instability and as in this patient leads to fusion to help prevent a catastrophic event. The presence of an os odontoideum or non fused tip of the dens may be seen but is not as typically present.

The vertebral bodies are decreased in height and at times may be completely flat yielding platyspondyly. Ovoid or trapezoidal bodies in the pediatric patient typically than yield vertebrae in the adult with decreased height, increased AP diameter, and end plate irregularities as seen here. Severe stenosis or C1/C2 kinking may be found as compared to the typical cervicomedullary kinking found in achondroplasia. In this patient, no myelopathic symptoms were present, astonishingly so.

Imaging of the other appendicular structures would have shown mutliple areas of epiphyseal dysplasia and advanced arthrosis.

Resources: Resnick. Diagnosis of Bone and Joint Disorders. 4th Ed. 2002
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