



History: 82 year old man with long standing bilateral hip pain. Patient with prior cervical spine surgery and no history of previous trauma.

LEFT CROSS TABLE

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Neutral lateral radiograph



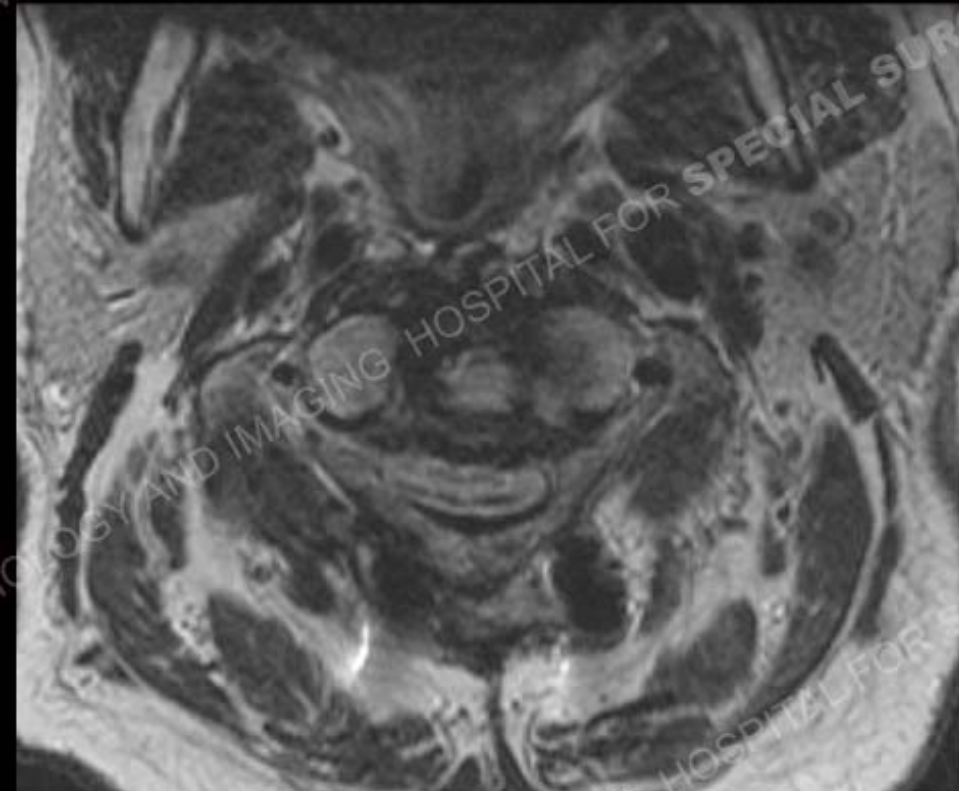
Flexion lateral radiograph



T1 Sagittal

T2 Sagittal

IR Sagittal



T2 axial at C1/C2 junction

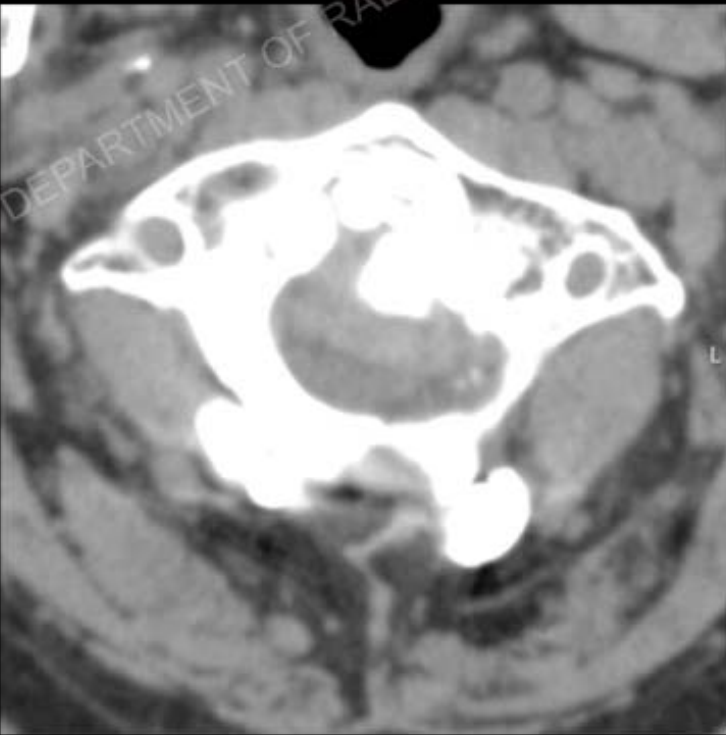


T2 Sagittal

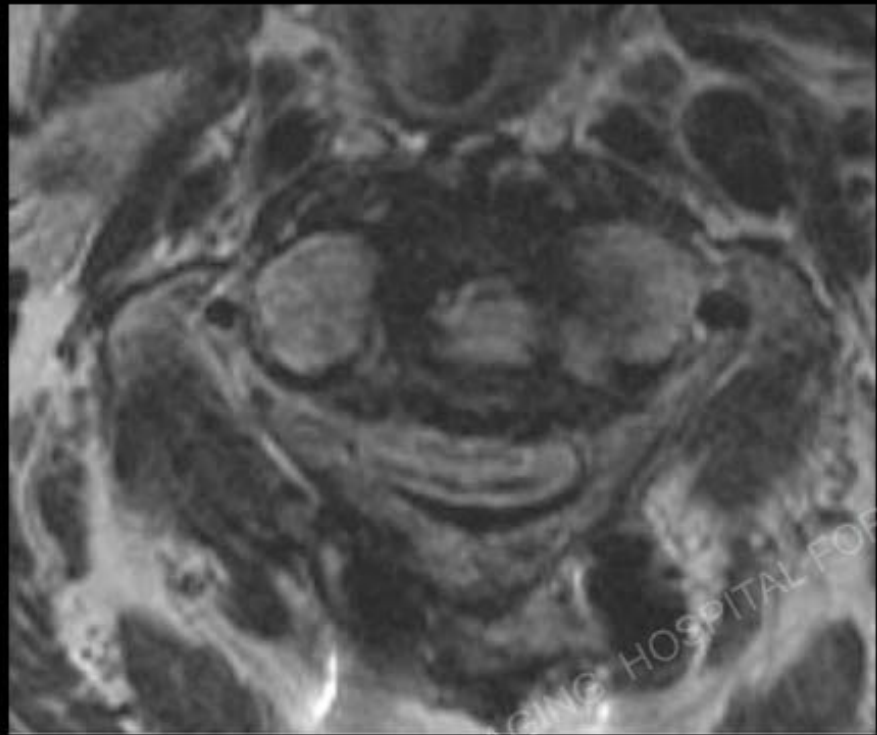


Sagittal reformations of CT study about the midsagittal line

DEPARTMENT OF RADIOLOGY AND IMAGING HOSPITAL FOR ...



Axial CT image at C1/C2 junction

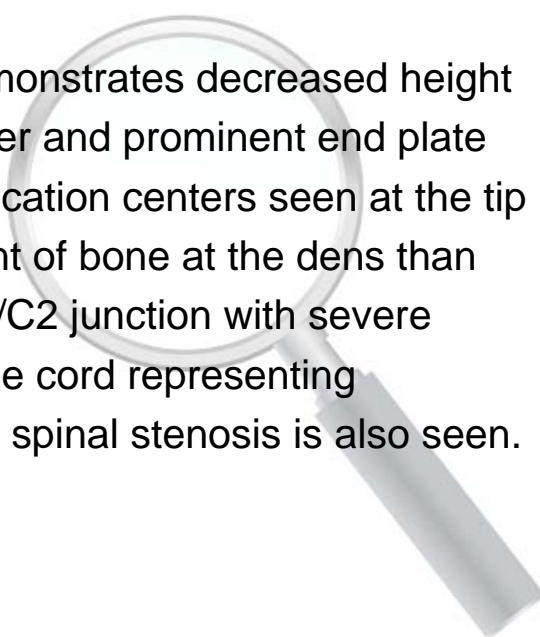


Axial T2 image at C1/C2 junction

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.



Deformed/Dysplastic femoral heads with severe arthrosis.



Foreshortened necks with coxa vara

Marked widening of atlantodens interval with flexion

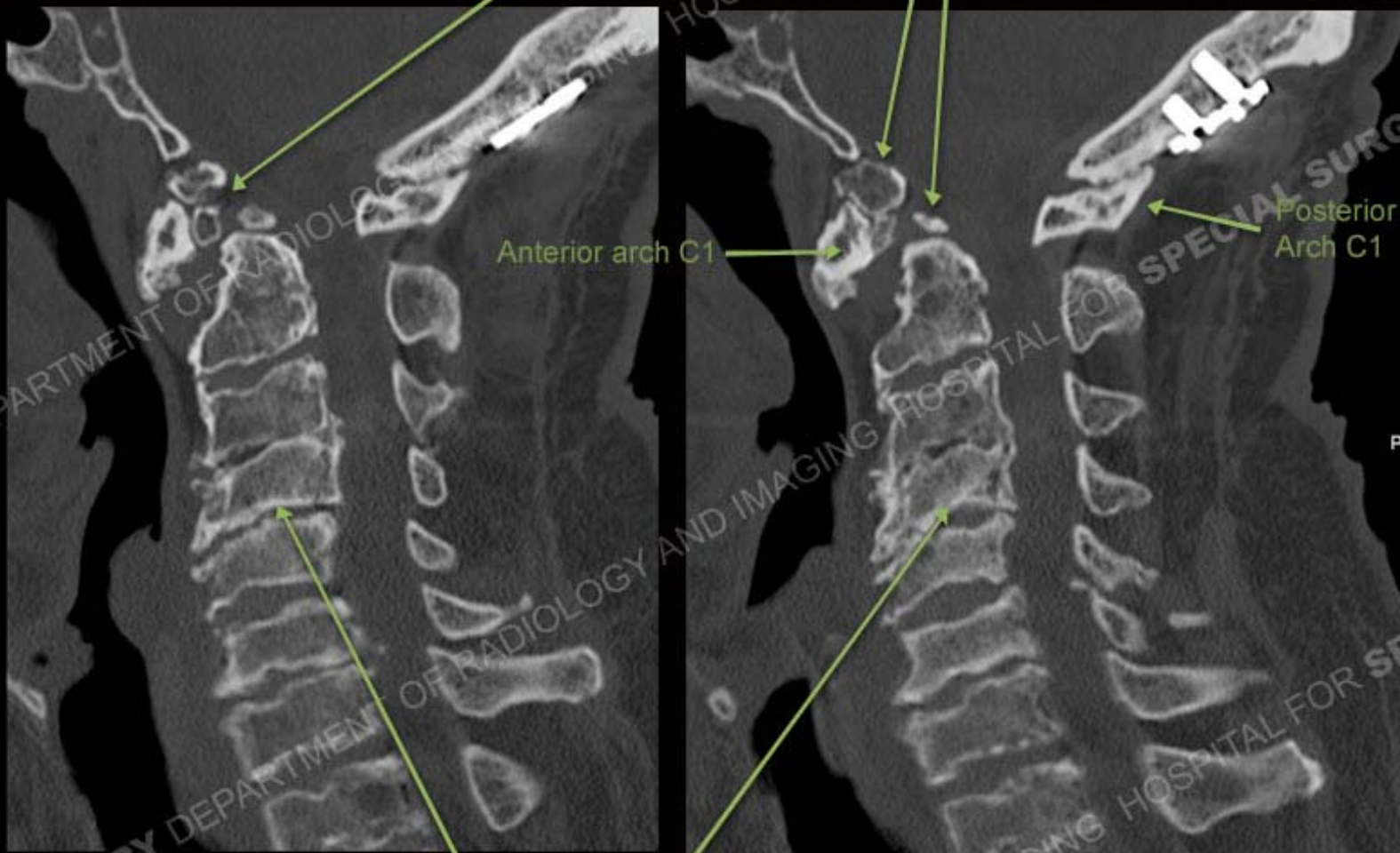


Previous posterior fusion



Decreased height, increased AP diameter, and irregular endplates of vertebrae

Non-fused multipartite tip of the dens (odontoideum) with overall hypoplasia of tip of the dens.



Abnormal vertebrae as described previously

Kinking or severe stenosis with compression of the cord at C1/C2

High signal in the cord

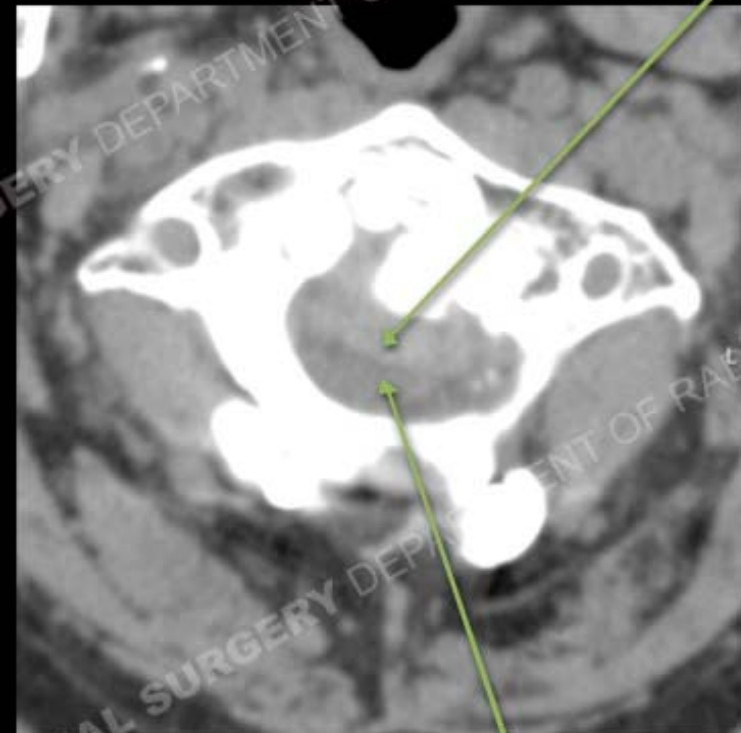


T1 Sagittal

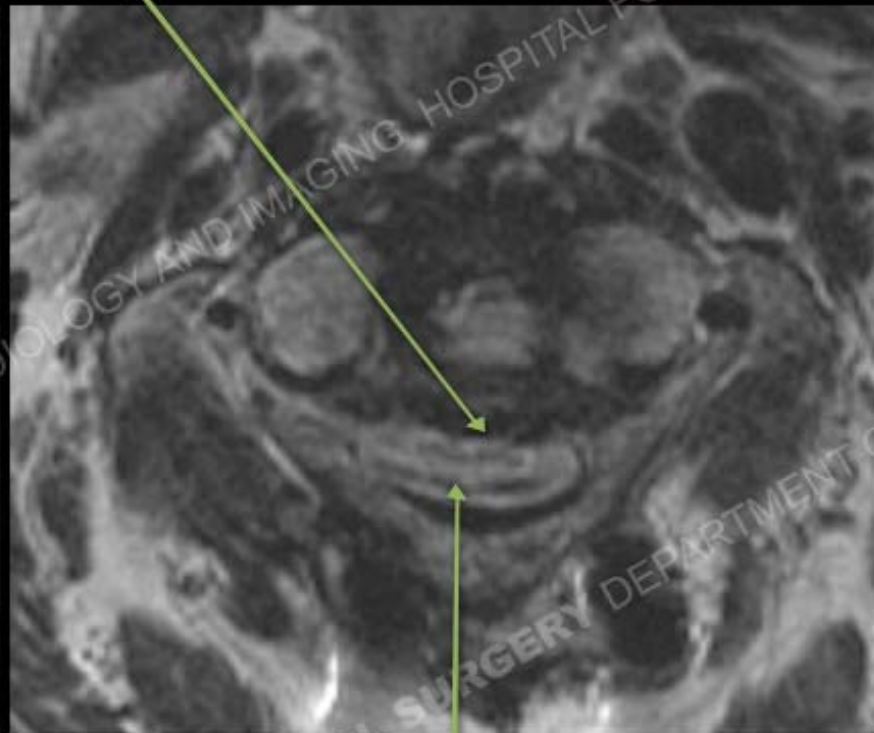
T2 Sagittal

IR Sagittal

Severe stenosis of central canal with severe compression of the cord



Lower attenuation is space available for cord which is severely compromised



High signal within the cord

Diagnosis

Spondyloepiphyseal Dysplasia (SED)

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.



Discussion

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 multiple areas of epiphyseal dysplasia and advanced arthrosis.



Resources

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

<http://emedicine.medscape.com/article/1260836-overview>

