

Progressive non-infectious anterior vertebral fusion (Copenhagen disease)

Published on 27.05.2015

DOI: 10.1594/EURORAD/CASE.12686

ISSN: 1563-4086

Section: Musculoskeletal system

Area of Interest: Musculoskeletal spine

Procedure: Imaging sequences

Imaging Technique: MR

Special Focus: Calcifications / Calculi Case Type:
Clinical Cases

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Patient: 39 years, male

Clinical History:

A 39-year-old male patient was referred to our hospital due to a long history of kyphosis and left-sided chest pain.

Imaging Findings:

An MRI study of the whole spine was performed, which showed prominent upper and mid-thoracic kyphosis and anterior fusion between the T4 and T9 vertebrae. Narrowing of the intervertebral discs at these levels, more marked anteriorly, was also demonstrated, as well as compensatory cervical hyperlordosis. There was no cord abnormality noted and the bone marrow appeared to be of normal signal intensity. The MRI findings were consistent with progressive non-infectious anterior vertebral fusion, or Copenhagen disease.

Discussion:

Progressive non-infectious anterior vertebral fusion (PAVF) is a rare, early childhood spinal disorder of unknown aetiology, which is characterized by the gradual onset of thoracic and/or lumbar spine ankylosis, often in conjunction with kyphosis. PAVF was first described in 1949 in a 14-year-old male patient by Knutsson [1]. A series of 26 patients was reported in 1991 by The University Hospital of Copenhagen and the term "Copenhagen syndrome" is also sometimes used to denote this condition [2]. A study of three cases by Hughes et al [3] was the first to discuss the importance of MR imaging in depicting the disease. MR imaging in that study demonstrated anterior disc narrowing and anterior endplate irregularity progressing to fusion. Associated bone oedema or fatty replacement was seen in the endplates as well.

The prevalence of PAVF is unknown, but approximately 80-100 cases have been reported (predominantly isolated ones). Women seem slightly more affected than men. The disease is often discovered in young children while investigating a spinal deformity, as it is in most cases clinically manifest by thoracolumbar kyphosis due to progressive anterior vertebral ankylosis in the thoracic and/or lumbar areas, which extends with time to the posterior parts. PAVF usually presents with mild pain, stiffness of the neck and/or back with developing thoracolumbar kyphosis. Often, however, PAVF is asymptomatic.

Neurological abnormalities are exceptional. PAVF can occur isolated or, less frequently, as part of a syndrome. Syndrome-associated manifestations include facial dysmorphism, absence of one cervical vertebrae, radio-ulnar synostosis, exostosis, generalized overgrowth and split cord malformation.

The radiological features are rather distinctive and involve narrowing of the anterior part of the disc space with associated erosion and irregularity of the anterior end-plates. The narrowing develops with disc space obliteration

and bony ankylosis anteriorly via a thick bony ridge [4]. The posterior aspect of the disc is relatively spared in the early stages but in some cases the narrowing may extend posteriorly with complete posterior fusion. The process may affect one level or several contiguous levels.

Spinal bracing may reduce or partially reverse the deformity. Prognosis is fairly good as it does not involve vital issues. However, most patients complain of low back pain and junctional thoraco-lumbar pain.

Differential Diagnosis List: Progressive non-infectious anterior vertebral fusion, Synspondylism, Spondylothoracic dysplasia

Final Diagnosis: Progressive non-infectious anterior vertebral fusion

References:

- Knutsson F (1949) Fusion of vertebrae following non-infectious disturbance in the zone of growth. *Acta Radiol* 32 (5-6):404-6 (PMID: [15402751](#))
- Andersen J, Rostgaard-Christensen E (1991) Progressive noninfectious anterior vertebral fusion. *J Bone Joint Surg Br* 73 (5):859-62 (PMID: [1894681](#))
- Hughes RJ, Saifuddin A (2006) Progressive non-infectious anterior vertebral fusion (Copenhagen Syndrome) in three children: features on radiographs and MR imaging. *Skeletal Radiol* 35(6):397-401 (PMID: [16328382](#))
- Smith JR, Martin IR, Shaw DG, Robinson RO (1986) Progressive non-infectious anterior vertebral fusion. *Skelet Radiol* 15(8):599-604 (PMID: [3810179](#))

Figure 1

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Description: Sagittal T2-weighted FSE sequence shows anterior fusion between the T4 and T9 vertebrae (arrows) with associated narrowing of the intervertebral discs anteriorly. Prominent upper and mid-thoracic kyphosis is also demonstrated. No cord abnormality identified. **Origin:** Department of Radiology, Stanmore Hospital

Figure 2

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Description: Sagittal T1-weighted SE sequence demonstrates the anterior fusion (white arrows) and normal vertebral bone marrow signal. Irregularity of the superior endplate is also noted (black arrow).

Origin: Department of Radiology, Stanmore Hospital