Atlanto-axial instability: a live-threatening disorder in rheumatoid arthritis

A cineradiographic case report

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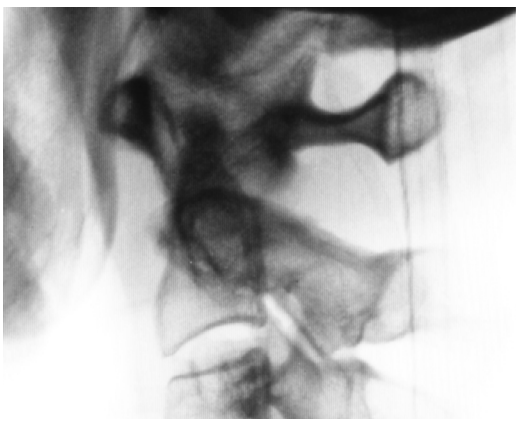
In 1974, a 35-year-old man who had suffered from rheumatoid arthritis (RA) for 14 years was planned for a knee joint replacement at Lund University Hospital. He had no neck pain and no neurological deficits. However, preoperative cervical radiographs showed an anterior atlanto-axial instability with 17-mm anterior atlas-dens interval in flexion (Figures 1 and 2). Håkan Brattström (1923–1999), the father of RA surgery in Sweden, ordered a cineradiography of the cervical spine which showed the instability in a dramatic way and led to caution at the knee arthroplasty. 6 weeks later, an occipitocervical fusion according to the Brattström-Granholm technique (Figure 3) using tension-band wire, bone cement unilaterally and bone chips contralaterally (Brattström and Granholm 1976) was performed. The stabilized neck remained asymptomatic for the rest of the patient's life—he died 27 years later from adenocarcinoma of the lung.

Discussion

Håkan Brattström knew that the earliest symptoms of atlanto-axial instability in RA are usually caused by compression of the occipital nerve roots, resulting in an occipital rhizopathy. He knew that further dislocation may cause compression of the spinal cord, resulting in myelopathy, long tract symptoms and tetraparesis; and that severe dislocation may cause compression of vertebral arteries, with signs of brainstem ischemia—such as vertigo or cranial nerve involvement—and ultimately resulting in respiratory arrest and death (Brattström and Granholm 1976). However, Mikulowski et al.



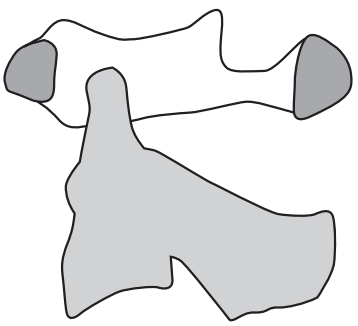


Figure 1. The cervical spine in extension.

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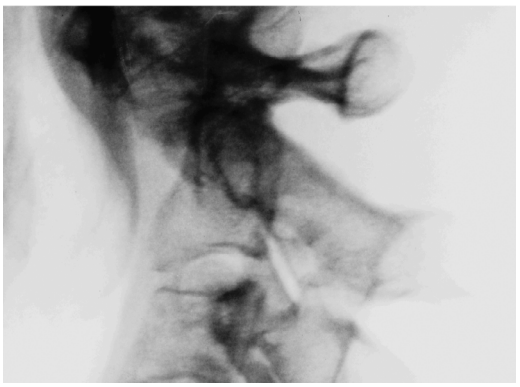

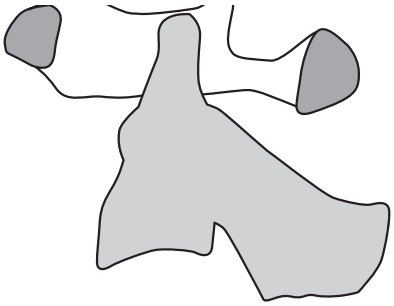

Figure 2. The cervical spine in flexion.

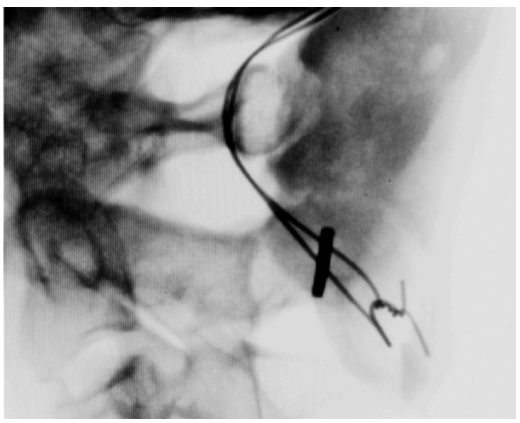

Figure 3. Occipito-cervical fusion according to the Brattström-Granholm technique.

(1975) had just found a high (and not previously reported) incidence of fatal medulla compression in RA patients with atlanto-axial dislocation, and a poor correlation with previous neurological symptoms. Thus, when Håkan Brattström performed the cineradiographic investigation, he was very alarmed when looking at the cineradiography as the patient bent his neck and he later related that he rushed towards the patient with the intention of catching his head!

In 1978, Håkan Brattström allowed me to do a copy of the cineradiography for a lecture. I saved this copy, which—now digitized and edited—is available at http://www.actaorthopscand.org/media/Cineradiography_BM.mov. Looking at this brief movie, it is easy to understand the alarm reaction of Håkan Brattström. Atlanto-axial instability may certainly be a life-threatening disorder in RA, even in the absence of neurological symptoms.

Brattström H, Granholm L. Atlanto-axial fusion in rheumatoid arthritis. A new method of fixation with wire and bone cement. Acta Orthop Scand 1976; 47 (6): 619–28.

Mikulowski P, Wollheim F A, Rotmil P, Olsen I. Sudden death in rheumatoid arthritis with atlanto-axial dislocation. Acta Med Scand 1975; 198 (6): 445–51.