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CASE REPORT

Osteoarthritis – a rare indication for atlantoaxial fusion. A case report and review of the literature

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The authors report a case of C1-C2 fusion which was performed in a 64-year-old woman with unilateral atlantoaxial osteoarthritis, who consulted because of incapacitating occipital pain and decreased cervical rotation without neurological deficit. The diagnosis of unilateral C1-C2 osteoarthritis was confirmed by an open-mouth radiograph, a bone-scan, a computerised tomography scan, and magnetic resonance imaging. C1-C2 fusion was performed using a computer assisted navigation system and posterior instrumentation. This resulted in marked relief of pain, and distinct improvement in quality of life. The prevalence of atlantoaxial osteoarthritis is about four per cent in patients suffering from peripheral or spinal osteoarthritis. However, many primary-care providers are not familiar with this entity. C1-C2 fusion remains rare, due to the difficulties related to the diagnosis of atlantoaxial osteoarthritis, its established non-operative treatment options, and the fact that it has not received adequate attention in the orthopaedic literature. However, based on review of the literature, it can be stated that C1-C2 fusion is an effective and safe procedure providing distinct reduction of pain and increased quality of life in case of failure of conservative treatment.

INTRODUCTION

The prevalence of atlantoaxial osteoarthritis (AAOA) was reported as 4% in 705 consecutive outpatients who had peripheral or spinal osteoarthritis *(7)*. Nevertheless, AAOA is still considered to be a rare cause of pain and instability *(1,5)*.

The diagnosis of AAOA is difficult, due to unspecific symptoms such as occipital neuralgia and vertigo *(2,3)* and to the fact that many primary-care providers are not familiar with it *(3,15)*. Because of the variety of non-operative treatment options (physical therapy, immobilisation, medication, and anaesthetic injections) with varying but unpredictable degrees of improvement, it was concluded in the past that there was no specific therapy for symptomatic AAOA *(7)*.

As a result of this unsatisfactory situation, various surgical treatment options were initiated, with the first successful arthrodesis for AAOA reported in 1978 (according to Ghanayem *et al (5)*). This publication was soon followed by other reports, displaying various surgical procedures to obtain fusion in case of AAOA with incapacitating pain *(2-9,14,15)*. According to several case series, fusion leads to distinct pain relief *(3,5,6,9,15)*. However, the indication for fusion in case of AAOA remains rare as this entity has not received adequate attention in

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Fig. 1. — Open-mouth radiograph revealing OA of the left C1-C2 facet joint with narrowing of the joint space, subchondral sclerosis, peripheral osteophyte formation, and erosions of the joint.

the orthopedic literature. This is the reason why we report the present case of unilateral AAOA as an indication for atlantoaxial fusion. We will also provide a review of the pertinent literature with special reference to diagnosis, indication, surgical procedures, and results.

CASE REPORT

A 64-year-old woman was admitted to the authors' hospital in July 2002. She complained of incapacitating occipital pain. There was no history of trauma. She had been treated conservatively (physical therapy, medication, and injections) for five years without relevant improvement.

On physical examination, cervical rotation to the left was painful and markedly limited, whereas pain was not affected by flexion and extension, as would be expected in association with degenerative changes in the subaxial (C3-C7) cervical spine. Neurological deficits were not present.

An open-mouth radiograph (fig 1) and a computerised tomography-scan (CT-scan) showed osteoarthritis (OA) of the left atlantoaxial facet joint with narrowing of the joint space, subchondral sclerosis, peripheral osteophyte formation, and erosions of the joint. C1-C2 instability was excluded by lateral flexion / extension radiographs.

Because of a history of breast cancer with ablation of the breast in May 2001, magnetic resonance imaging (MRI) was performed to exclude a cervical metastasis or an infection. MRI also demonstrated a C5-C6 disc herniation without compression of the cord, and disc protrusion at C3-C4 and C4-C5. A bone scan, which was carried out for the same reason, revealed focal uptake unilaterally at C1-C2.

As a preoperative test, a local anaesthetic was injected into the left atlantoaxial facet joint under fluoroscopic control ; this relieved the pain completely.

An additional CT-scan for the VectorVision® computer assisted surgery (CAS) system permitted precise operative planning by determining the anatomy of the first and second cervical vertebrae and the course of the vertebral arteries. Surgery was performed in September 2002, using the CT-scan based CAS system (Spine 5.0 software) and the Neon® spinal system. CAS systems had already been demonstrated to be safe and reliable devices providing increased accuracy in screw positioning *(10,11)*. The spinal system used was a modular rod-screw device for posterior cervical stabilisation. In case of C1-C2 fusion, it consists of transarticular screws, rods, rod/screw connections providing a high angular stability, and special atlas claws. *In vitro* biomechanical testing confirmed rigid internal fixation in case of C1-C2 fusion, which does not depend on bone graft and sublaminar wiring *(12,13)*.

The surgical procedure was performed under general anaesthesia using a Mayfield clamp for head positioning and a standard posterior midline approach. The posterior arches of C1 and C2 were exposed. After CAS system calibration – revealing an accuracy of 1.0 mm – two K-wires were placed transarticularly using two additional distal skin incisions (fig 2). The positioning of both K-wires was controlled under fluoroscopy (fig 3). The screws and atlas claws were inserted and connected by the rods. After harvesting cancellous autografts from the posterior iliac crest, the posterior aspect of both C1 and C2 was decorticated and packed with cancellous bone.

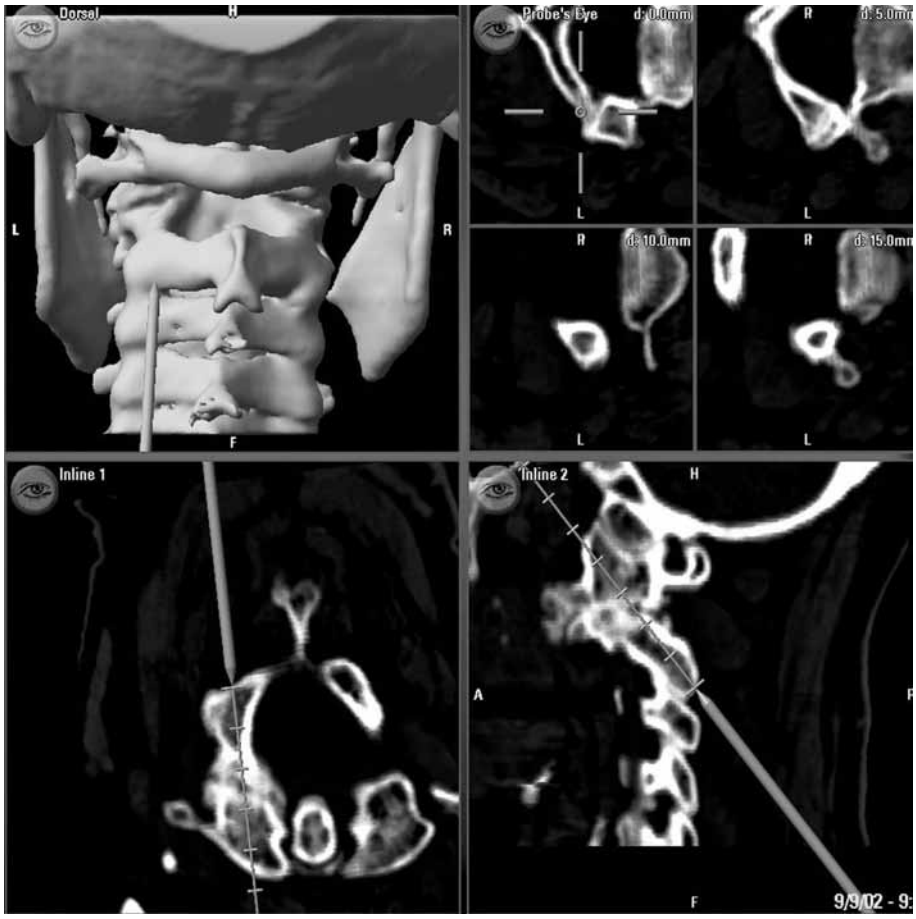

Fig. 2. — Placement of the K-wires by means of the CAS system.

Postoperative anteriorposterior and lateral radiographs (fig 4) as well as a CT-scan confirmed the correct position of the implant. Following surgery, the patient was supplied with a rigid cervical orthosis for one week and with a soft cervical collar for another week.

Her complaint about pain decreased markedly according to a visual analog scale (7.4 six weeks preoperatively ; 2.5 two weeks postoperatively ; 1.4 one year postoperatively). Analysis of quality of life using the SF-36 (36-Item Short-Form Health Survey) six weeks preoperatively and two weeks, and one year postoperatively, revealed a distinct improvement (physical functioning : 75 / 85 / 90, bodily pain : 22 / 84 / 74, role-physical 25 / 50 / 50,

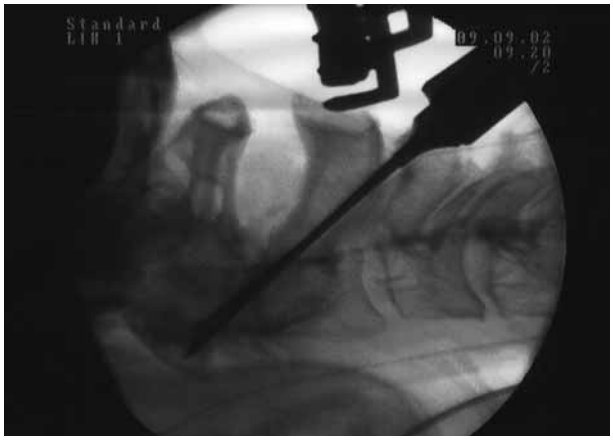

Fig. 3. — Intra-operative fluoroscopy confirming the correct position of the K-wires.

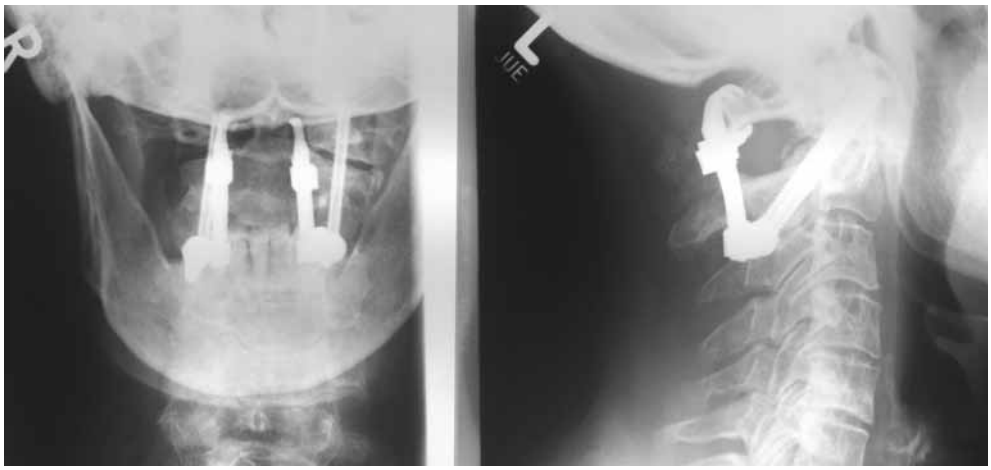

Fig. 4. — Postoperative AP and lateral radiographs.

general health : 40 / 67 / 47, vitality : 40 / 60 / 60, social functioning : 50 / 75 / 88, role-emotional : 0 / 67 / 100, mental health : 60 / 68 / 84) as did the Oswestry scale (34 / 14 / 0). Solid fusion was achieved six months postoperatively, which was confirmed by biplanar radiographs and CT-scan.

REVIEW OF THE LITERATURE

The Medline database was searched for AAOA and C1-C2 fusion. Nine case reports and studies regarding AAOA were identified between 1981 and 2001.

The earliest report originates from Harata *et al* *(8)*. In 1981 they described 31 patients with AAOA (14 females, 17 males), suggesting that this disease might not be particularly uncommon. They reported on characteristic radiological and histological signs and established a classification according to the radiological appearances, displaying three different subtypes and their distribution (12% lateral atlantoaxial, 17% atlantoodontoid, and 71% mixed). Treatment was conservative in all cases except one, a 63-year-old woman with incapacitating occipital pain secondary to OA, who was managed with transoral C1-C2 fusion leading to a marked improvement.

Three years later, Ehni and Benner characterised the C1-C2 arthrosis syndrome *(2)*. They reported on seven patients with unilateral AAOA and occipital pain syndrome. Diagnosis was made by open-mouth radiograph, isotope bone-scan, and CT-scan. Occipital neuralgia was attributed to developmental and posttraumatic lesions in the cervico-cranial junction region, with the nerve roots at the first and the second cervical level as the principal pain pathways. The authors advocated anaesthetic and steroid injections for temporary pain relief (n = 4) and dorsal rhizotomy of the nerve root at C2 for permanent pain relief (n = 2). Atlantoaxial fusion was performed in combination with dorsal rhizotomy in one case.

In 1987 Halla and Hardin estimated the prevalence of AAOA as 4% in 705 consecutive outpatients who had peripheral or spinal OA *(7)*. The clinical syndrome of the 27 patients with lateral AAOA was distinctly different from the one seen in patients with subaxial degenerative disease of the spine. Occipital pain, occipital trigger points, and a rotational head tilt, usually associated with collapse of one of the facet joints, were the main symptoms of AAOA. One patient treated with arthrodesis experienced relief of his symptoms. However, due to the variety of treatment options with varying but unpredictable degrees of improvement a lack of apparent specific therapy for symptomatic AAOA was concluded.

Star *et al* reported in 1992 on a series of nine patients with severe occipito-cervical pain secondary to lateral AAOA, which in most cases

had been diagnosed as occipital neuralgia *(15)*. Non-operative treatment was successful in four patients. However, C1-C2 fusion was neccessary in five patients (Brooks arthrodesis n = 2, Gallie arthrodesis n = 3). Four patients were rated excellent, whereas one patient with a poor result following a Gallie procedure needed a second fusion due to pseudarthrosis. In contrast to Ehni and Benner, who considered the C1 and C2 nerve roots as the principal pain pathways *(2)*, Star *et al* focused on the degenerative changes in the joint itself as the principal cause of pain which led to a 56% rate of C1-C2 fusions in all of their AAOA patients *(15)*.

Joseph and Kumar reported in 1994 on four patients with unilateral AAOA *(9)*. Total relief of the occipital neuralgia was obtained in all patients following atlantoaxial fusion by the Gallie technique. Fusion was achieved in all cases.

In 1996 Ehrat *et al* presented the results of eight patients suffering from post-traumatic osteoarthritis of the cervico-cranial area, who were treated with suboccipital C0-C2 fusion *(3)*. Surgery was performed in three using the transarticular posterior screw fixation according to Magerl. Fusion was successful in all cases. A significant reduction of pain and increased quality of life were achieved.

In the same year, Ghanayem *et al* evaluated the results of 15 patients in whom symptomatic AAOA had been treated with posterior fusion (14 patients) or anterior transoral fusion (one patient) (*5*). At the latest follow-up (14 patients available) 13 patients had an excellent result, and one a fair result. One quadriparesis recovered completely.

Song *et al* reported in 1997 on 19 patients with atlantoaxial instability ; one patient underwent a unilateral posterior atlantoaxial transarticular screw fixation *(14)*.

The latest report concerning C1-C2 fusion secondary to OA comes from Fuentes *et al (4)*. In 2001 they published three cases of symptomatic unilateral AAOA. One of these patients was treated by atlantoaxial fusion after failure of conservative treatment, which consisted of CT-scan guided steroid injection.

DISCUSSION AND CONCLUSION

OA of the subaxial (C3-C7) cervical spine is a common finding in elderly patients *(1)*. It is characterised by neck and shoulder pain rather than occipito-cervical pain, which is more typical of C1-C2 OA (*2,3,5,7,9)*. The majority of primary-care providers are not familiar with this difference in the clinical picture *(3,15)*. Consequently, in case of occipito-cervical pain and co-existing subaxial degenerative changes, AAOA may be overlooked *(5)*.

The main clinical symptoms of this entity are severe occipito-cervical pain and marked decrease of cervical rotation (*2-5,7-9,15)*.

In most cases an open mouth radiograph reveals the diagnosis of AAOA *(2-5,8,9,15)*, whereas flexion-extension radiographs *(3,4,5)*, CT-scan *(2-5,15)*, bone-scan *(3,15)*, and MRI (*3-5,15)* can be useful in case of co-existing atlantoaxial instability or suspected tumoral or infectious disease. If diagnosis remains unclear, an anaesthetic injection under fluoroscopy in the affected joint is advocated to confirm the origin of the complaints *(3,8)*.

In the past it was felt that there was no specific therapy for symptomatic AAOA, because of the variety of non-operative treatment options with variable but unpredictable degrees of improvement *(7)*. Of course, in case of AAOA without neurological deficit or symptomatic instability, an initial therapy consisting of physical therapy, immobilization, medication, and anaesthetic injections, usually is beneficial *(2-8,15*) and therefore strongly advocated. In contrast to this, AAOA with intractable and persistent occipital pain, not responsive to conservative treatment, or with co-existing instability or with a neurological deficit, can be treated with C1-C2 fusion (*3,5,6,9,15)*. In our opinion, the good clinical results are explainable by the hypothesis that degeneration of the facet joints causes the occipital pain. Consequently, we believe that fusion should be the treatment of choice in selected patients. This hypothesis is supported by the fact that fusion almost immediately relieves the pain *(6)*.

The radiographic prevalence of lateral AAOA in the general population is about five percent *(16)*.

This should raise awareness of the entity. Both surgical and non-surgical treatment options need to be considered. Prospective and randomised studies are advocated in order to determine which patients will benefit from conservative or operative treatment.

REFERENCES

1. **Dvorák J.** Epidemiology, physical examination, and neurodiagnostics. *Spine* 1998 ; 23 : 2663-2673.
2. **Ehni G, Benner B.** Occipital neuralgia and the C1-2 arthrosis syndrome. *J Neurosurg* 1984 ; 61 : 961-965.
3. **Ehrat C, Grob D, Dvorák J.** Die posttraumatische atlantoaxiale Arthrose. *Orthopäde* 1996 ; 25 : 542-545.
4. **Fuentes S, Bouillot P, Palombi O, Manera L, Desgeorges M.** Arthrose atloïdo-axoïdienne unilatérale. À propos de trois cas et revue de la littérature. *Neurochirurgie* 2001 ; 47 : 51-54.
5. **Ghanayem AJ, Leventhal M, Bohlman HH.** Osteoarthrosis of the atlanto-axial joints. Long-term follow-up after treatment with arthrodesis. *J Bone Joint Surg* 1996 ; 78-A : 1300-1307.
6. **Grob D.** Surgery in the degenerative cervical spine. *Spine* 1998 ; 23 : 2674-2683.
7. **Halla JT, Hardin JG Jr.** Atlantoaxial (C1-C2) facet joint osteoarthritis : a distinctive clinical syndrome. *Arthritis Rheum* 1987 ; 30 : 577-582.
8. **Harata S, Tohno S, Kawagishi T.** Osteoarthritis of the atlanto-axial joint. *Int Orthop* 1981 ; 5 : 277-282.
9. **Joseph B, Kumar B.** Gallie's fusion for atlantoaxial arthrosis with occipital neuralgia. *Spine* 1994 ; 19 : 454-455.
10. **Richter M, Amiot LP, Neller S, Kluger P, Puhl W.** Computer-assisted surgery in posterior instrumentation of the cervical spine : an in-vitro feasibility study. *Eur Spine J* 2000 ; 9 (suppl) : 65-70.
11. **Richter M, Amiot LP, Puhl W.** Computernavigation bei der dorsalen Instrumentierung der HWS – eine In-vitro-Studie. *Orthopäde* 2002 ; 31: 372-377.
12. **Richter M, Schmidt R, Claes L, Puhl W, Wilke HJ.** Posterior atlantoaxial fixation : biomechanical in vitro comparison of six different techniques. *Spine* 2002 ; 27 : 1724-1732.
13. **Richter M, Wilke HJ, Kluger P, Neller S, Claes L, Puhl W.** Biomechanical evaluation of a new modular rod-screw implant system for posterior instrumentation of the occipito-cervical spine : in-vitro comparison with two established implant systems. *Eur Spine J* 2000 ; 9 : 417-425.
14. **Song GS, Theodore N, Dickman CA, Sonntag VK.** Unilateral posterior atlantoaxial transarticular screw fixation. *J Neurosurg* 1997 ; 87 : 851-855.
15. **Star MJ, Curd JG, Thorne RP.** Atlantoaxial lateral mass osteoarthritis. A frequently overlooked cause of severe occipitocervical pain. *Spine* 1992 ; 17 (suppl) : 71-76.
16. **Zapletal J, de Valois JC.** Radiologic prevalence of advanced lateral C1-C2 osteoarthritis. *Spine* 1997 ; 22 : 2511-2513.