Case Report

Transoral surgery for myelopathy caused by rheumatoid arthritis of the cervical spine

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Spontaneous forward luxation at the atlanto-axial level is a well-known and common complication of rheumatoid arthritis. The distance between the articular surface of the odontoid process of the axis and the corresponding articular surface of the atlas should not normally exceed 3 mm. in adults; a larger distance indicates luxation. Conlon, Isdale, and Rose (1966) found luxation between atlas and axis in 25 per cent. of 333 consecutive patients of rheumatoid arthritis. Luxation at this level initially leads to compression of occipital nerve roots causing radiating pain in the occipital region. Increasing luxation is known to result in medullary or arterial compression with severe neurological deficit and the outcome may be fatal. Smith, Benn, and Sharp (1972) found that 30 of 150 patients with atlanto-axial luxation already had signs of medullary and/or arterial compression or developed them during the period of observation.

The surgical treatment of atlanto-axial luxation in rheumatoid arthritis thus seems well justified. The method of choice seems to be posterior exposure, reposition, and fusion between the occiput and the cranial part of the cervical spine. Fusion with bone grafts only entails prolonged immobilization, which may be harmful, particularly in a patient with rheumatoid arthritis. Over the last 3 years we have treated eleven patients with rheumatoid atlanto-axial luxation, using a technique permitting early mobilization. Our results are encouraging and have been reported elsewhere (Brattström and Granholm, 1973). The method used is a further development of the technique described by Scoville, Palmer, Samra and Chong (1967) for metastatic disease of the spine and requires the use of wire, acrylic plastic, and bone chips to obtain fixation and fusion.

The prerequisite for this forward atlanto-axial luxation is a destruction of the ligaments between C1 and C2. Rheumatoid arthritis may, however, cause a much more wide-spread lesion in adjacent bone, mainly in the joint facets, leading to a downwards luxation of the atlas on the axis. Smith and others (1972) found downwards luxation in nine of their 150 cases of rheumatoid atlanto-axial luxation. In this condition, the odontoid process may protrude through the foramen magnum and further reduce the space for the spinal cord. In one case recently treated by us this complication resulted in paraplegia due to pressure on the medullary structures intracranially. This case is of interest, not only because this complication may not be too uncommon, but also because such patients may be treated surgically. In the case described below, the odontoid process was first removed by a transoral route, and fixation of the occiput to the cervical spine was then achieved in a second procedure.

Case report

A 65-year-old woman, in whom the diagnosis of rheumatoid arthritis had been suspected at the age of 23, had had joint changes limited to the hands and knees in the early decades of the disease, but in recent years there was also increasing involvement of the hips, elbows, and shoulders. The erythrocyte sedimentation rate had been raised for 40 years and serological tests were found to be positive. She had been treated with steroids, both systemically and locally.

For the last 12 months she had occipital pain and radiating pain in both arms, and for 6 months before her present admission to the neurosurgery department, she had been confined to a wheelchair because of difficulty in walking. She had undergone hip surgery 4 months before the most recent admission to hospital. During the last 2 or 3 months she had gradually developed increasing weakness of both legs, spastic reflexes, and spinal automatism. For the last week she was bedridden and could not empty her bladder.
Examination

All this was confirmed on admission and a loss of sensation to pinprick and touch below the level of D9 was also demonstrated. Radiographs of the cervical spine seemed to show a fracture of the odontoid process, but the abnormal location of this process could also have been due to the fact that the entire axis was severely tilted dorsally. The forwards luxation in the atlanto-axial joint was measured to about 10 mm. It was more difficult to measure the downwards luxation, but the odontoid process protruded for several millimetres through the foramen magnum. Severe decalcification was noted in the entire cervical spine. This set of radiographs is unfortunately not suitable for reproduction.

Progress

The patient was placed on a Stryker frame on admission and skeletal traction was applied by means of Blackburn tongs. X-ray examination on the following day indicated that the dorsal tilting of the axis was reduced but that the odontoid process still protruded intracranially (Fig. 1).

The position of the bone structures did not change for the next 5 weeks, although the patient was subjected to traction with increasing load throughout this period of time. During the first 2 weeks there was neurological improvement, as sensibility returned, and she could start to move her legs voluntarily again. The return of function was, however, not complete, and as this was thought to result from compression of medullary structures by the odontoid process, it was decided to perform an anterior decompression by the transoral route.

Surgical Procedures

The Stryker frame was used as an operating table, and the patient was operated on during skeletal traction and in the supine position. An oral endotracheal tube was inserted. The soft palate was split in the midline up to the hard palate to expose the epipharyngeal space. The retropharyngeal tissues in this space were divided in the midline from the caudal end of clivus and for 4 cm. caudally. The odontoid process was visualized above the anterior arch of the atlas. The availability of the operative field is shown in Fig. 2.

FIG. 2 Drawing indicating principal structures involved during transoral removal of odontoid process. It is necessary to split the soft palate, but the odontoid process is then accessible through the opening between the clivus and the anterior arch of the atlas.

The base of the odontoid process was separated from the body of the axis by means of an air-drill and the process was removed piece-meal. A small portion of the apex of the process, which was strongly adherent to the dura, was left in place (Fig. 3, overleaf). The dura was severed at the end of the procedure, and the extradural space was covered with Oxycel to control the ensuing cerebrospinal fluid leakage. The tissues of the epipharyngeal space and the soft palate were closed with adsorbable stitches and finally a tracheostomy was performed because of an insidious swelling of the tissues. The patient was tube-fed for 10 days and was given antibiotics for the same period of time because of the cerebrospinal fluid leakage during surgery.

FIG. 1 Lateral x-ray of base of skull and the cranial part of cervical spine. The odontoid process protrudes for several millimetres intracranially. Arrow indicates apex of process.
The tracheostomy was maintained for 3 weeks and she was kept in skeletal traction during this period. After 3 weeks, a posterior fusion was done from the occipital bone to C2 and C3, using wire, pins, and acrylic plastic. The procedure was performed under general anaesthesia with the patient prone and in a cerebellar headrest. The occipital bone and the cranial part of the cervical spine were exposed through a straight midline incision. Four burr-holes were placed in the occipital bone and a heavy wire was passed up and down through these, anterior to the lamina of the atlas and around pins in the spinal processes of C2 and C3 (Fig. 4). The wire was tightened to accomplish reposition, whereupon burr-holes, wire, pins, and spinal processes were covered with acrylic plastic. The wound was closed in separate layers.

Postoperative course
The patient was found to have improved neurologically as soon as she recovered from anaesthesia after the first operative procedure. Figs 1 and 3 are pre- and post-operative radiographs. The tracheostomy was closed and the skeletal traction was removed on the first day after the second procedure, whereupon she was mobilized without external support. The very long period of inactivity during skeletal traction (8 weeks) had further reduced the power of joint movement, but the neurological improvement progressed as she was given physiotherapy.

Discussion
This case is of interest not only as a reminder of the possibility of atlanto-axial dislocations in rheumatoid arthritis, which are quite common and can be corrected surgically, but also as an example of the possibilities of treatment in the progressive course of certain cases of rheumatoid arthritis. Transoral removal of the odontoid process is probably not very frequently performed, but this technique has been advocated by Southwick and Robinson (1957) and by Fang and Ong (1962). This method was also used by Sukoff, Kadin, and Moran (1972) in a case of rheumatoid arthritis in which the odontoid process was removed 6 months after a posterior fusion occiput to C2. A lengthy and technically difficult procedure in our case was rewarded by an unexpectedly smooth postoperative course.

We have found that signs of medullary or radicular compression are frequently underestimated in this disease, so that most cases are not referred for treatment until they have severe neurological deficit. Sensibility to touch and pin-prick is always difficult to estimate, and the examination is even more difficult if the patient has been subjected to numerous surgical procedures on the joints of the arms and legs with ensuing reduction of sensibility around scars. Medullary or radicular compression ultimately leads to paralysis, but muscular weakness is common in chronically ill patients with rheumatoid arthritis because of inactivity and stiffness. Reflexes may be
absent or impaired around severely affected joints. It is quite common, however, that pain from radicular compression is described as a ‘new’ sensation. It thus seems appropriate to pay more attention to the complaints of the patient than to the doubtful indications of neurological examination.

**Conclusion**

Rheumatoid arthritis is frequently complicated by atlanto-axial luxation, which can be corrected surgically. The paper describes in detail the treatment of a woman in whom it proved necessary to remove the odontoid process transorally before a posterior fusion could be performed. The main symptom when treatment started was spastic paraplegia, and this gradually improved after surgery.

**References**

Brattström, H., and Granholm, L. (1973) Der Orthopädie, 2, 118 (Chirurgie der Halswirbelsäule bei Patienten mit rheumatoider Arthritis)


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