Rheumatoid C₁–C₂ dislocation: pathogenesis and treatment reconsidered

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SYNOPSIS The commonly accepted mechanism of rheumatoid C₁–C₂ dislocation is the incompetence of the transverse ligament of C₁ vertebra. The presence of rheumatoid granulation tissue between the anterior arch of C₁ vertebra and the odontoid process is an important factor. This might explain why the dislocation becomes irreducible when the disease progresses. In a 55 year old man, transoral removal of the rheumatoid granulation tissue made the previously irreducible C₁–C₂ dislocation reducible. Stabilization of the reduced C₁–C₂ articulation was accomplished at a second operation by posterior fusion of the occiput to C₁, C₂, and C₃ vertebrae.

Although dislocation of the first or second cervical vertebra (‘C₁–C₂ dislocation’) is a well-recognized complication of rheumatoid arthritis (Conlon et al., 1966), the exact mechanism producing the dislocation is not yet fully understood.

The most common type of C₁–C₂ dislocation in rheumatoid arthritis is the forward dislocation of C₁ on C₂ vertebrae. The diagnosis is made when, in radiographs of the lateral cervical spine, the distance between the anterior arch of C₁ vertebra and the odontoid process is greater than 2.5 mm in females and 3 mm in males (Conlon et al., 1966).

During the early stages of the disease, the distance between the anterior arch of C₁ vertebra and the odontoid process varies with head position, being greatest with the head flexed and minimal or normal with the head extended. As the disease progresses, the difference between head flexion and extension becomes less and less obvious, and the distance between the anterior arch of C₁ vertebra and the odontoid process eventually becomes fixed and irreducible even under traction.

Incompetence of the transverse ligament of C₁ vertebra has generally been accepted as the cause of forward dislocation of C₁ on C₂ vertebra. Martel and Abell (1963) demonstrated that the transverse ligament ‘had areas of degeneration and increased vascularity . . . were lax and resulted in extreme mobility of the atlanto-odontoid joints’. This was the cause of a fatal dislocation in a 61 year old white female with a 20 year history of rheumatoid arthritis. In a 53 year old female who died of rheumatoid C₁–C₂ dislocation, Webb et al. (1968) found the transverse ligament of C₁ vertebra destroyed to such a degree that it could not be identified.

In the presence of a lax or ruptured transverse ligament, C₁ vertebral body slips forward on C₂ vertebra in flexion, but the displacement of the odontoid process remains limited. The odontoid process does not reach the posterior arch of C₁. Werne (1957) proposed that the alar ligaments limit the extent of backward dislocation of the odontoid process when the transverse ligament is destroyed. Were this hypothesis correct, the forward dislocation of C₁ on C₂ vertebra should be reducible with the head placed in extension. However, frequently the reduction remains incomplete and the gap between the anterior arch of C₁ vertebra and the odontoid process with the head in extension remains excessive, although less than when the head is flexed. We postulate that this failure to reduce the dislocation might be explained by the presence of rheumatoid granulation tissue between the anterior arch of C₁ vertebra and the odontoid process. In a 65
year old white male who died of rheumatoid $C_1$–$C_2$ dislocation reported by Gleason and Urist (1965), there was a mass of rheumatoid granulation tissue resulting in an increased space between the anterior arch of $C_1$ vertebra and the odontoid process. Although the presence of the rheumatoid granulation tissue is mentioned in this (Gleason and Urist, 1965) and one other (Bland, 1967) necropsy report, this information has not been applied to solve the clinical problems.

Recently we have seen a quadriparetic patient with a fixed $C_1$–$C_2$ dislocation. A mass of granulation tissue was removed from between the anterior arch of $C_1$ vertebra and the odontoid process and the dislocation subsequently became reducible.

CASE REPORT

(E.T. 68–64) A 55 year old American Indian male was admitted to the Madison VA Hospital on 29 May 1971 with quadriparesis and urinary retention of two days' duration.

The patient had had rheumatoid arthritis for 15 years and had been confined to a wheelchair for three years. On the morning of 27 May 1971, after a fit of coughing he felt his neck snap and a few hours later was unable to feed himself at the breakfast table and pain was noted in the back of his neck. He remained in bed most of the day and could not void. That evening he was admitted to a local hospital and was catheterized. The next day the patient was transferred to VAH Madison.

At the time of admission he was obtunded. A cataract was present in the right eye; the left eye ground was normal. An extensive rheumatoid pro-

FIG. 1. (a) Preoperative radiograph of cervical spine showing dislocated $C_1$ on $C_2$ vertebra. The dislocation was not reducible. A halo tong was used to maintain the head position during and after operation. (b) Postoperative cervical spine radiograph showing successful reduction of the $C_1$–$C_2$ dislocation. Halo tong support has been removed.
cess had deformed shoulders, elbows, wrists, knees, and ankles. There was flexion contracture at the knees. The patient was barely able to raise his legs from the bed and move his toes. The upper extremities were paralysed and flaccid. Sensory examination showed a sensory level with loss of pain and temperature from the neck down with preservation of vibration, position, and light touch sensation in all the extremities and the trunk.

He showed evidence of urinary tract infection caused by *Klebsiella* and *Proteus*. The blood haemoglobin was 12.9 g/100 ml and haematocrit 37.2%. Serum protein level was 6.6 g and albumin 2.8 g/100 ml. The sedimentation rate was 50/36 mm in the first two hours. The Waaler-Rose test was positive at 1:1280. No cold agglutinins were detected. The antinuclear antibody test was also negative.

The cervical spine radiograph showed a considerable amount of C₁-C₂ dislocation. The odontoid process was eroded but not separated from the body of C₂ vertebra. The distance between the anterior arch of C₁ vertebra and the anterior margin of the odontoid process was 15 mm, and the distance between the posterior arch of C₁ vertebra and the posterior margin of the odontoid process was only 12 mm (Fig. 1a).

The patient was immediately placed on a Stryker frame with Crutchfield tong skull traction. It then became obvious that the dislocation was not reducible even with neck extension and weight loading. However, the discomfort in the neck disappeared, and the neurological findings were not made worse by the traction.

He remained on the Stryker frame and in traction for the next five months. The dislocation remained basically unchanged but the neurological findings showed some improvement. He became able to raise both arms; limited use of fingers returned.

Removal of the posterior arch of C₁ vertebra was considered in the hope of decompressing the spinal cord (Alexander et al., 1953; Dastur et al., 1965). However, this would have resulted in increased instability of C₁ over C₂ vertebra and result in possibly fatal medullary injury. In view of our hypothesis that the failure to achieve reduction is due to overgrowth of rheumatoid connective tissue between the anterior arch of C₁ vertebra and the odontoid process, it was thought that the treatment of choice to reduce the dislocation without removing any bone was to remove the tissue between the anterior arch of C₁ vertebra and the odontoid process through a transoral approach.

On 10 November 1971, Crutchfield tong traction was discontinued. A halo tong was used to maintain the head position. A cervical myelogram showed no tissue growth posterior to the odontoid process; the tectorial membrane was smooth and without bulges. On 29 November 1971 a tracheostomy was performed and the next day a transoral decompression was carried out under a Zeiss dissecting microscope. The soft palate was split at the midline. The tubercle of the anterior arch of C₁ vertebra was found to be prominent; the normal anatomy of atlanto-odontoid synovial structure was not present. There was tissue herniation above and below the anterior arch of C₁ vertebra and the space between it and the odontoid process was densely packed by the same tissue. Four grams of this tissue were removed piecemeal under the dissecting microscope. It had a white appearance and firm consistency and was easily cracked by the rongeurs. Microscopically it consisted of a mixture of fibrous tissue and bone trabeculae. Round cell infiltration and necrosis were prominent (Fig. 2).

After removal of the tissue between the anterior

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**FIG. 2.** Rheumatoid tissue removed at time of surgery from the space between anterior arch of C₁ vertebra and the odontoid process. In some areas it showed bone trabeculae invaded by rheumatoid tissue (above), while in others it was mainly connective tissue with palisading fibroblasts, round cell infiltration, and necrosis (below).
arch of C1 vertebra and the odontoid process, and with the patient still under general anaesthesia and under direct vision of the dissecting microscope, the patient’s head was readjusted. Movement was seen at both anterior arch of C1 vertebra and the odontoid process and the distance between them was reduced to 4 mm. The reduced position was again maintained by the halo tong.

On 6 January 1972 a posterior occipito-C1-C2-C3 vertebral fusion was carried out using gauge 20 wires and acrylic plastic reinforcement. The halo tong was removed on the 10th day after fusion. A postoperative radiograph of the cervical spine showed the distance between the anterior arch of C1 vertebra and the odontoid process to be 4 mm and the distance between the posterior arch of C1 vertebra and the odontoid process to be 22 mm (Fig. 1b). The patient was discharged on 27 January 1972. At the time of discharge he was in a wheelchair and able to use his arms and hands for daily activity of living. He was able to stand, but could not walk because of the flexion contraction of the knees and extensive rheumatoid deformities of the feet.

**DISCUSSION**

Review of necropsy reports of 12 patients who died of dislocation of C1 on C2 vertebra due to rheumatoid arthritis (Davis and Markley, 1951; Martel and Page, 1960; Martel and Abell, 1963; Gleason and Urist, 1965; Verjaal and Harder, 1965; Bland, 1967; Webb et al., 1968) shows that incompetence of the transverse ligament of C1 vertebra was found in only three patients (Martel and Abell, 1963; Webb et al., 1968). Of these three patients, two died of upper cervical cord compression from the displaced odontoid process (Martel and Abell, 1963; Webb et al., 1968). In the third patient (Webb et al., 1968), although the transverse ligament was incompetent, the cause of death was due to a sudden collapse of the lateral body of C1 vertebra with bilateral vertebral artery occlusion and the vertical upward displacement of the odontoid process which penetrated the medulla. Thus in only two patients can the incompetent transverse ligament be related to the patient’s death.

In all 12 patients, the fundamental abnormality was a chronic synovitis involving almost all the synovial joints of the cervical spine with the most severe change at the synovial joints between C1 and C2 vertebrae. Significant rheumatoid granulation tissue was found between the anterior arch of C1 vertebra and the odontoid process on six occasions (Gleason and Urist, 1965; Bland, 1967), in the retro-odontoid space on two occasions (Martel and Abell, 1963; Gleason and Urist, 1965), and between C4 and C5 vertebrae on one occasion (Bland, 1967). In one patient backward dislocation of C1 vertebra due to rheumatoid process was associated with a large intraspinal meningioma attached to the body of C2 vertebra (Verjaal and Harder, 1965).

These unpredictable findings suggest that a complete diagnostic evaluation by myelography is certainly needed to plan the surgical strategy if surgical intervention is elected. A routine posterior decompression or fusion without myelographic study could be disastrous in these patients.

There are also three necropsy reports of immediate death due to the sudden collapse of the lateral body of C1 vertebra or the occipito condyle with medullary injury by the odontoid process (Davis and Markley, 1951; Martel and Page, 1960; Webb et al., 1968). Extensive bone erosion, fragmentation, and softening involved not only the odontoid process, the anterior arch and lateral body of C1 vertebra but the occipital bone as well.

The fact that in rheumatoid arthritis resulting in C1–C2 intervertebral dislocation the anterior arch and the lateral body of C1 vertebra may also be weakened by the rheumatoid process creates a serious contraindication to the posterior decompression by removal of the posterior arch of C1 vertebra. The experiences of Alexander et al. (1953) and Dastur et al. (1965) show the high operative risk that is associated with posterior decompression even in non-rheumatoid C1–C2 dislocation.

Of considerable interest is a report by Sukoff et al. (1972) concerning a patient who had an irreducible rheumatoid C1–C2 dislocation and was treated initially by posterior decompression with removal of the posterior arch of C1 and fusion of C2 vertebra to the occiput. The fusion was solid but the dislocation remained unreduced. Six months later quadriparesis recurred and a transoral removal of part of the anterior arch of C1 vertebra together with the rheumatoid granulation tissue and the odontoid process was performed to prevent further neurological deterioration. Had the transoral ap-
approach been performed first as in our case the pathological process would have been corrected and the C1–C2 dislocation reduced. A simpler posterior fusion, rather than decompression, may then be advised for stabilizing the reduced C1–C2 vertebral articulation.

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REFERENCES


