ATLANTO - AXIAL SUBLUXATION IN RHEUMATOID ARTHRITIS*

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Rheumatoid arthritis has a high morbidity but a low mortality. One cause of death in this disease is compression of the cervical cord following atlanto-axial subluxation. This makes the finding of atlanto-axial subluxation of great importance, although the relationship of small amounts of subluxation to symptoms and subsequent progress has not been established. Similarly, the importance of a chance finding of subluxation is uncertain.

In this study, the entire rheumatoid population of a hospital clinic has been surveyed both clinically and radiologically to determine the incidence of this abnormality.

Material

76 consecutive out-patients with rheumatoid arthritis were studied; they represented the population served by the hospital. Their disease satisfied the American Rheumatism Association criteria for rheumatoid arthritis. Seventy of the patients studied had at least one of the following additional features: positive rheumatoid factor tests, erosions, or nodules.

Control subjects were selected from the physical medicine department clinics, the physiotherapy department, and the hospital beds. No person with headache or clinical evidence of rheumatoid arthritis was included in the control group. The 28 control subjects were matched with the rheumatoid patients for age and sex.

Method

Details of the apparent duration of the disease, of systemic steroid therapy, nodules, and radiological changes were recorded. Erosive changes were recorded as absent, present, or severe, the last if one or more bones in the hands or feet were seen in radiographs to be so eroded as to have an unrecognizable contour. Symptoms referable to the cervical spine were enquired after, namely pain or stiffness in the head or neck either spontaneously or on movement, and paraesthesiae on

neck movement. The range of neck movement was assessed in three planes using a goniometer to measure the deflections of a pointer attached to the head. Signs thought to be referable to the upper cervical spine were sought with no prior knowledge of the radiological findings.

(1) "Clunking" was recorded as present if on flexion and extension of the neck a distinct jerk in the normally smooth movement could be seen or felt.

(2) The "palate sign" was recorded as positive if the index finger of the examiner feeling the posterior pharyngeal wall could detect abnormal separation of the anterior arch of the atlas from the body of the axis during neck flexion. The correct site for the index finger tip was found by sliding the finger along the dorsum of the tongue till it met the posterior pharyngeal wall, then by moving it up and down until the ligament between the anterior arch of the atlas and the body of the axis could be felt. The opposite hand then eased the patient's neck alternately into flexion and extension, whilst the examining finger felt for changes in the relationship of the two bones (Fig. 1, opposite).

(3) Abnormal laxity was judged by cradling the occiput on the thenar aspect of the examiner's left hand with the ring and small fingers supporting the dorsal processes of the atlas and axis. During gentle flexion of the upper cervical spine the relative movement of the occiput, atlas, and axis can be felt. Antero-posterior laxity was detected by attempting to lift the patient's head without causing forward flexion (Sharp and Purser, 1961). These two tests were performed with the patient supine.

A selective neurological examination was performed on the patients with special reference to the presence of long tract signs.

Radiography of the atlanto-axial joint was confined to a mid-line lateral tomogram taken in full active flexion of the upper cervical spine (Fig. 2a). To achieve a mid-line film, three or more cuts at 0.5 cm. intervals were taken and one film was selected for measurement. All radiographs were taken with a tube to film distance of 70 cm., the mid-line cut varying from 13.5 to 21 cm. from

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Fig. 1.—The "palate sign" is positive if the examiner's finger can feel abnormal separation of the anterior arch of the atlas from the body of the axis during flexion of the patient's neck.

Fig. 2a.—Lateral tomogram.

Fig. 2b.—Line drawing prepared by tracing Fig. 2a. The meaning of the letters is as follows:

- (a) atlanto-axial separation (mm.).
- (b) spinal canal depth (mm.).
- (c) tilt of odontoid peg (in degrees) anterior or posterior to line perpendicular to foramen magnum line.
- (d) thrust of odontoid peg (mm.) above or below foramen magnum line.

the film. The resulting magnification factor varying from 1.24 to 1.43:1 has been ignored (maximum difference 1.15:1). Measurements were made from a tracing of the radiograph (Fig. 2b). A line was drawn passing through the centre of the facing surfaces of the anterior and posterior arches of the atlas. The distance "a" from the adjacent surfaces of the anterior arch and odontoid peg, and "b" from the odontoid peg to the posterior arch were measured. A line was also drawn across the lower border of the foramen magnum (fora-

men magnum line). The number of degrees of tilt "c" anteriorly or posteriorly of the line bisecting the odontoid peg to a line perpendicular to the foramen magnum line was recorded. The distance "d" of the tip of the odontoid peg above or below the foramen magnum line was measured.

Results

Radiography

The most important radiological feature of atlanto-axial subluxation is the size of the gap between the anterior arch of the atlas and the odontoid peg (Fig. 2*b*, *a*). In control subjects this did not exceed 2 mm. Checks of the accuracy of the measurement were made. The tracings of the radiographs were measured by two independent observers, fifteen radiographs were re-traced and re-measured, and twelve patients were x-rayed on subsequent occasions and the radiographs re-measured. The maximum error was 0.5 mm. [In two patients the change was much greater, an increase of 1.5 and 3.5 mm. respectively. Presumably this represented increase in actual subluxation]. Thus, taking separation of 3 mm. or over as abnormal, nineteen patients (25 per cent.) had atlanto-axial subluxation. Fig. 3 demonstrates the percentage incidence of atlanto-axial separation in the patient and control groups ($\chi^2=20.30$; $p \leq 0.001$).

The depth of the spinal canal represents the space available for the spinal cord (Fig. 2 "b"), and it

Fig. 3.—No control subject had atlanto-axial separation of as much as 3 mm.

might be anticipated that those patients with shallow canals would sustain spinal cord injury with smaller amounts of subluxation. The measurements of depth are recorded in Table I.

The tip of the odontoid peg may be tilted forwards or backwards relative to its base, and when tilted backwards seems more likely to impinge on the spinal cord. This tilt has been measured in degrees of tilt forwards (+ve) or backwards (—ve) in relation to a line at right-angles to the foramen magnum line (Fig. 2 "c"). Owing to the difficulty of choosing the central axis of the odontoid peg (by joining two points midway between anterior and posterior surfaces), this angle could not be measured more closely than 5°. The results are recorded in Table I.

The distance of the tip of the odontoid peg above (positive) or below (negative) the foramen magnum line was measured. An error of 3 mm. was not uncommon if one or other of the bony landmarks was not clear, but in general the measurement was reproducible with an error not greater than 2 mm. These results are presented in Table I as the thrust.

Symptoms and Signs

Half the patients admitted to neck pain, and six of these had subluxation. There were thirteen subluxations in the group with no pain (Table II).

TABLE II
PRESENCE OF NECK PAIN RELATED TO
SUBLUXATION

Neck pain		Present	Absent
Total Patients		38	38
Subluxation	No.	6	13
	Per cent.	16	34

Neck stiffness and paraesthesiae on neck movement bore no relationship to subluxation. The measure-

TABLE I
RADIOGRAPHICAL MEASUREMENTS

Groups Examined		Depth of Spinal Canal (mm.)			Tilt of Odontoid Peg (°)						Thrust of Odontoid Peg (mm.)			
		15-19	20-24	25-30	-8 and Over	Up to -8	Up to -4	Up to +4	Up to +8	+8 and Over	Over -6	Up to -6	Up to -3	Positive
Patients	No.	12	43	21	12	10	19	20	8	7	13	34	25	4
	Per cent.	16	56	28	16	13	25	26	11	9	17	45	33	5
Controls	No.	2	18	8	2	4	9	5	6	2	6	17	5	0
	Per cent.	7	64	29	7	14	32	18	21	7	21	61	18	0

TABLE III
NECK SIGNS RELATED TO SUBLUXATION

Neck Signs	Clunk		Laxity		Palate	
	Positive	Negative	Abnormal	Normal	Positive	Negative
No. of Patients	6	70	31	45	7	69
No. Subluxed	2	17	6	13	5	14

ments of the range of neck movement was reproducible to 5° if checked immediately, but variations of up to 15° in a single movement were found if the ranges were re-measured on a subsequent occasion. These measurements are therefore not presented, but there was no obvious difference in range between those patients with and those without subluxation.

The results of examination for the three signs thought to be indicative of atlanto-axial subluxation are presented in Table III. The "clunk" sign was often sought with the patient supine. As this sign may depend upon gravity more positive results might have been obtained with the patient sitting. The "palate" sign (Fig. 1) was the most reliable, there being only two "false positive" results. On the other hand the sign was recorded as negative in twelve patients with subluxation. These three signs were all sought on at least two occasions for each patient (except for one who failed to re-attend and another who died from intercurrent illness).

Patients with Subluxation

Subluxation was more common in patients with rheumatoid arthritis of longer duration (Table IV) although in one it was detected after only 19 months of disease. There was no relationship to age (Table

TABLE IV
DURATION OF DISEASE RELATED TO SUBLUXATION

Duration of Disease (yrs)		0-4	5-9	10-14	15-19	Over 20	Total
Total Patients		32	16	12	10	6	76
Subluxed	No.	3	6	3	6	1	19
	Per cent.	9	37	25	60	17	—

TABLE VI
CLINICAL FEATURES RELATED TO SUBLUXATION

Clinical Findings		Sex		Serology		Erosions		Nodules		Total
		Female	Male	Negative	Positive	Mild	Severe	Absent	Present	
Total Patients		50	26	21	55	64	14	54	22	76
Subluxed	No.	10	9	4	15	12	7	11	8	19
	Per cent.	20	35	19	27	19	50	20	36	—

TABLE V
AGE OF PATIENTS RELATED TO SUBLUXATION

Age (yrs)		Under 50	50-59	60-69	Over 70	Total
Total Patients		18	24	24	10	76
Subluxed	No.	4	7	7	1	19
	Per cent.	22	29	29	10	—

V), subluxation being found in patients from 38 to 75 years. Subluxation was found more frequently in patients with positive tests for rheumatoid nodules, or severe erosions, and was also commoner in males (Table VI). Of the seventeen patients on corticosteroid treatment, nine had subluxation.

Neurological Signs

Fifty patients were found to have abnormal neurological signs. In 33 there was absence of one or more abdominal reflex, 22 had tendon reflex abnormalities, twelve had sensory impairment, five motor weakness, and five one or two extensor plantar responses. Only the last was thought to signify long tract involvement, and these five are analysed in Table VII (overleaf). Three had atlanto-axial subluxation, but not of such a degree as to be certain of its causal relationship to the long tract signs. In Case 2 the spinal canal was only 19 mm. deep, possibly compounding the effect of 4.5 mm. of subluxation, and Case 5 had 10° of backward tilting of the odontoid peg, possibly contributing to the neurological damage. All five cases were sero-positive with peripheral joint erosions, the latter being severe in two. Two complained of neck pain, and three were taking corticosteroid drugs. In three patients the odontoid peg showed radiological erosions.

TABLE VII
CLINICAL FINDINGS IN FIVE ARTHRITIC PATIENTS WITH LONG TRACT SIGNS

Case No.	1	2	3	4	5
Age (yrs)	65	58	56	80	60
Sex	F	M	F	F	M
Duration of Disease (yrs)	8	15	10	21	6
Serology	+	+	+	+	+
Erosions*	++	+	+	++	+
Nodules	0	0	0	0	0
Neck Pain	0	0	0	+	+
Corticosteroid Therapy	+	+	+	0	0
Local Signs	+	0	0	0	0
	+	+	0	0	0
	+	0	0	0	0
Radiological Measurements	5 24 0 0	4.5 19 -1 -3	3 27 +2 -10	2 20.5 +2 -1.5	1 26 -10 -3
Odontoid Peg Eroded	+	+	0	0	+

* + Erosions present.
++ Erosions severe.

Discussion

The incidence of atlanto-axial subluxation in groups of rheumatoid patients varies with the selection of the patients, and the technique used. Sharp and Purser (1961) studied 79 patients admitted during one year, and examined lateral radiographs of the cervical spine in flexion in 74 of them. Fourteen (19 per cent.) had abnormal separation of the odontoid peg from the anterior arch of the atlas. Their study differed from the present one in three respects: all patients had needed admission to hospital; the radiographs were not tomographs; the measurement was made from the middle of the posterior surface of the anterior arch of the atlas to the nearest point on the anterior surface of the odontoid process. They also read the cervical spine radiographs of a population sample studied by Kellgren and Lawrence (1956). Of the 186 patients with rheumatoid arthritis in the population, six had atlanto-axial displacements, a prevalence of 32.3 per 1,000. Martel (1961) selected 34 patients because of persistent neck discomfort or severe progressive disease. Lateral tomograms in flexion were taken and 24 were found to have abnormal atlanto-axial separation. Martel (1964) also examined 100 "unselected" consecutive clinic cases, and found 34 with an abnormal atlanto-axial interval; this clinic, however, attracted patients with severe disease. De Sèze, Djian, and Debeyre (1963) took lateral radiographs of the neck in flexion of 57 patients with neck symptoms of 103 referred to

their clinic, and found subluxation in ten. Serre, Simon, Janicot, and Levy (1964) examined sixty in-patients and found 23 with subluxation. Conlon, Isdale, and Rose (1966) examined 333 in-patients (233 with classical rheumatoid arthritis, 85 definite, and fifteen probable); 84 patients (25 per cent.) had subluxation of the atlanto-axial joint.

Rheumatoid arthritis may narrow the intervertebral discs, erode the vertebral plates, and cause vertebral subluxation or apophyseal erosions (Sharp, Purser, and Lawrence, 1958). The mechanism of the atlanto-axial separation has been studied experimentally by Zinn (1968), who showed that division of the transverse ligament of the atlas enabled only the upper part of the anterior joint space to open. Division or destruction of other ligaments is required for greater amounts of subluxation.

The relationship of neurological complications to atlanto-axial subluxation is not direct. Conlon and others (1966) found upper motor neurone signs in 23 patients with rheumatoid arthritis; four of these had no radiological abnormality of the cervical spine, but eleven had atlanto-axial subluxation. Similarly, of the five patients we found with long tract signs, two did not have subluxation, and in the three who did, the amount was only 3, 4.5, and 5 mm. This emphasizes that other factors may cause serious neurological sequelae of this type. Co-incidental causes of upper motor neurone lesions may be present, or a sub-axial lesion may cause fatal cord

damage (Whaley and Dick, 1968). Sharp (1964) discussed the destructive effects of rheumatoid arthritis lower in the cervical spine, and pointed out that cord damage might occur in these situations. Webb, Hickman, and Brew (1968) described fatal vertebral artery thrombosis in a patient with cervical rheumatoid arthritis.

We have not looked at the lower parts of the cervical spine in this study. It may be that rapidity of development of atlanto-axial subluxation is a serious feature, and that those patients who have survived 5 to 10 mm. of subluxation have passed through the phase of maximum danger. A prospective study of rheumatoid arthritis is needed to evaluate this point, and to clarify which groups most need treatment.

Summary

An out-patient population was examined for atlanto-axial subluxation, and this feature was found in 25 per cent. of patients.

It was relatively commoner in males and occurred regardless of age. It was less common, but not excluded, if the disease had been present for less than 5 years. As expected, it was associated with the presence of severe erosive disease, rheumatoid factor, and nodules. The absence of neck pain by no means excluded subluxation, in fact freedom from neck pain was rather more common in those affected. Of the clinical signs associated with the occipito-atlanto-axial joints only the "palate" sign proved of real value. Five patients with long tract signs were also detected and the significance of the lack of correlation between these two features is discussed. A technique for measuring radiological atlanto-axial subluxation which gives reproducible results is described.

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La subluxation atloïdo-axoïdienne dans la polyarthrite rhumatoïde**RÉSUMÉ**

Un certain nombre de malades externes a été examiné pour la subluxation atloïdo-axoïdienne, et cette lésion a été trouvée chez 25 pour cent des cas.

Elle était relativement plus commune chez les hommes, sans égard pour l'âge. Elle était moins commune, mais pas absente, si la maladie avait été présente pendant moins de cinq ans. Comme on pouvait s'attendre, elle était associée à la présence de la forme sérieuse et érosive de la maladie, du facteur rhumatoïde et de nodules. L'absence de douleur au cou n'excluait nullement la subluxation, en fait l'absence de douleur au cou était plutôt plus commune chez ceux affectés. Des signes cliniques associés aux articulations occipito-atloïdo-axoïdienne seulement le signe du "palais" a prouvé être de valeur réelle. Cinq malades donnant des signes d'atteinte des longs faisceaux ont été aussi vus et la signification du manque de corrélation entre ces deux signes est discutée. Une technique pour mesurer la subluxation atloïdo-axoïdienne radiologique donnant des résultats reproductibles est décrite.

Subluxación atlantoaxil en la poliartritis reumatoide**SUMARIO**

Un grupo de pacientes externos fue examinado en busca de subluxación atlantoaxil, y esta característica fue hallada en un 25 por ciento de los pacientes.

Era relativamente más común en los hombres y ocurría en todas las edades. Era menos común, pero no desaparecía por completo, si la enfermedad había estado presente durante menos de cinco años. Tal cual de esperaba, estaba asociada con la presencia de enfermedad erosiva aguda, factor reumatoide y nódulos. La ausencia de dolor en el cuello de ningún modo excluía la subluxación; de hecho, la falta de dolor en el cuello era quizá más común en las personas afectadas. De los signos clínicos asociados con las articulaciones occipito-atlantoaxiales, solamente el signo del "paladar" resultó ser de verdadero valor. También fueron detectados cinco pacientes con síntomas de tracto largo y se discute el significado de la falta de correlación entre estos dos aspectos. Se describe una técnica para la medición radiológica de la subluxación atlantoaxil que de resultados que se pueden reproducir.