Backward luxation of the atlas

Two cases of an uncommon condition

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Neck involvement in cases of rheumatoid arthritis is extremely common, and the radiological features have been investigated by several authors, particularly Martel (1961, 1964), Bland, Davis, London, Van Buskirk, and Duarte (1963), Serre, Simon, Janicot, and Lévy (1964) and Conlon, Isdale, and Rose (1966). The features most characteristic are atlanto-axial subluxation, serial subluxation, and disc narrowing without osteophytosis. Atlanto-axial subluxation may be of any degree, and surprisingly is not usually associated with neurological abnormalities. Such subluxation is characterized by a forward movement of the atlas on the axis. Backward luxation is so rarely seen that the two following cases are thought worthy of report.

CASE 1

A 57-year-old Caucasian male developed generalized joint pains at the age of 40, and since has had widespread rheumatoid involvement. He has had a full conservative regime of treatment, including salicylates, chloroquine, and myocrisin. He now has severe joint deformities, particularly of the hands (with ulnar deviation), feet, elbows, and hips. He has skin pigmentation, evidence of vasculitis in his fingers, and rheumatoid elbow nodules. Just before admission to the Royal North Shore Hospital, Sydney, a large ulcer on the medial aspect of his ankle had broken down. This had been present in 1966 when he had a 6-month course of oral corticosteroids while it healed. He has chronic neck pain radiating to the occipital area made worse by neck movements. The range of neck movement is almost full with only a little loss of rotation and

FIG. 1 Case 1. Lateral view of neck, showing disorganized atlanto-axial joint.
Side flexion, and there are no abnormal neurological signs.

Blood tests show moderate anaemia; erythrocyte sedimentation rate (ESR) 114 mm. in 1 hr; Waaler-Rose test positive at a titre of 1:1,028; serum albumin 3.1 g. per cent., and serum globulin 2.2 g. per cent. Blood urea is 40 mg./100 ml. and the urine shows a trace of albumin.

Joint x rays show erosive changes and subluxation, while x rays of the cervical spine show a backward subluxation of the atlas on the axis. The odontoid process cannot be seen on tomography and the anterior arch of the atlas shows some erosion (Figs 1 and 2).

CASE 2

A 55-year-old housewife had a short attack of arthritis without residua at the age of 20. At the age of 43, she developed arthritis in one wrist and soon experienced widespread joint involvement, with pain, stiffness, and progressive loss of function.

First seen at Queen Elizabeth Hospital, Rotorua, at the age of 47, she has since been followed for 8 years, having numerous minor surgical procedures in addition to a full conservative programme. She had corticosteroids for a period, but did not derive real help from them. Gold and chloroquine were also used for several courses but without modifying her progressive course.

Throughout the period of observation she has had marked elevation of the ESR, and has been strongly sero-positive with rheumatoid nodules and hypergamma-globulinaemia. She is mildly hypertensive and has had evidence of arteritis with leg ulcers and nail-fold lesions.

Her joints now show marked destruction both clinically and radiologically. The neck was first clinically involved 4 years ago; 2 years ago she started to get vertigo on sudden head movements. She complains of some difficulty in controlling her head in the mornings, but has only slight restriction of neck movement, and no abnormal neurological signs.

X rays of the neck since 1962 show separation of the
odontoid peg and instability at the atlanto-axial level (Figs 3 and 4). In extension, backward luxation of the atlas is more marked than with the neck in flexion. There are marked destructive changes at the atlanto-occipital and atlanto-axial joints, and there is some osteoporosis of the anterior arch of the atlas.

Discussion

Non-traumatic spontaneous backward luxation of the atlas on the axis is an extremely rare condition. We have been able to find in the world literature only one previous case, reported by Verjaal and Harder (1965). This patient, also suffering from chronic sero-positive rheumatoid arthritis, was a 66-year-old housewife with widespread joint involvement and a high ESR, who had been treated with gold, salicylates, and pyridine derivatives. She was seen because of severe neck pain; 3 weeks later she developed signs of cord compression which progressed to complete spastic tetraplegia and death. X-ray examination of the cervical spine disclosed absence of the odontoid process and the anterior arch of the atlas with backward displacement of the atlas on the axis. At autopsy these findings were confirmed. There were destructive rheumatoid changes in the atlanto-occipital joints but the atlanto-axial joints were undisturbed. There was also present a soft tissue mass at the C1-C2 level which was felt to be a contributing factor. Verjaal and Harder (1965) did not present evidence to exclude the possibility that it was a rheumatoid nodule, rather than, as stated, a meningioma.

For backward luxation of the atlas to occur the anterior arch of the atlas must be destroyed or congenitally absent, or the odontoid process must be fractured, destroyed, or congenitally absent. In Case 1 the odontoid process could not be demonstrated by tomography, while in Case 2 the separated process was still in normal relationship with the eroded anterior arch. Destruction of the odontoid peg in cases of rheumatoid arthritis of the cervical spine is occasionally seen, but in these cases anterior luxation of the atlas is the rule (Fig. 5).

In the case reported by Verjaal and Harder, the absence of the anterior arch of the atlas and the odontoid peg together with rheumatoid involvement of the lateral joints provides a reasonable explanation of the backward luxation of the atlas, but in our cases, in neither of which was there complete destruction of the anterior arch, the reason why backward rather than forward luxation of the atlas has occurred is obscure.
Summary

Two cases of backward luxation of the atlas in patients with severe chronic rheumatoid arthritis have been described. In neither case were there severe neck symptoms or neurological signs. For backward luxation to occur the odontoid process must be destroyed, fractured, or congenitally absent, or the anterior arch of the atlas must be destroyed or congenitally absent. The factors leading to backward rather than forward luxation are obscure.

References


Résumé

La luxation en arrière de l’atlas

Deux cas de luxation en arrière de l’atlas chez des malades atteints d’arthrite rhumatoïde chronique grave ont été décrits. Dans aucun des cas il n’y avait des symptômes cervicaux ou des signes neurologiques. Pour que la luxation en arrière ait lieu l’apophyse odontoïde de l’atlas doit être détruite, fracturée ou absente de naissance, ou l’arête antérieure de l’atlas doit être détruite ou absente de naissance. Les facteurs qui conduisent à la luxation en arrière plutôt qu’en avant sont obscurs.

Sumario

Luxacion del atlas hacia atrás

Se han descrito dos casos de luxación del atlas hacia atrás, en pacientes con poliartritis reumatoide crónica grave. En ninguno de los dos se observaban síntomas severos en el cuello o signos neurológicos. Para que ocurra luxación hacia atrás, el proceso odontoide debe estar destruido, fracturado o ausente por razones congénitas, o el arco anterior del atlas debe estar destruido o ausente por razones congénitas. Los factores que conducen a la luxación hacia atrás, más que hacia adelante, son inciertos.
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