Bulbar symptoms and episodic aphony associated with atlanto-occipital subluxation in ankylosing spondylitis

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SYNOPSIS A patient with intermittent aphony associated with atlanto-occipital subluxation due to ankylosing spondylitis is presented and discussed. The only other case from the literature is reviewed and compared with our patient, where symptoms and signs of episodic low bulbar disease, presumably due to intermittent vascular insufficiency, were relieved by external bracing.

Atlantic-axial subluxation has been well documented in ankylosing spondylitis and rheumatoid arthritis (Kornblum et al., 1952; Vignon and Patet, 1955; Sharp and Purser, 1957; Martel, 1961; Sharp and Purser, 1961; Reitan, 1968). It is often associated with a myelopathy due to the compression of the cervical cord by the dens. Non-traumatic, non-congenital atlanto-occipital subluxation has been recorded in only two instances, both in patients with ankylosing spondylitis, or Marie-Strumpel disease (Coste et al., 1960; Martel, 1961). One of these patients was recorded in detail and presented with symptoms and findings remarkably similar to the case we are reporting. These unusual but striking symptoms, combined with the presence of a rare but specific explanation, suggested the desirability of recording our observations so that others might be aware of its occurrence.

CASE REPORT

(PBBH 123450) This 55 year old white male was admitted because of incapacitating neck muscle spasm and ‘spells’ in December 1969. A history of back and hip pain attributable to ankylosing spondylitis had been present for 21 years. Neck stiffness was first noted by the patient in 1960. Since 1963 posterior cervical pain and stiffness had readily progressed to the point where functional fusion had occurred. To be able to see to the left or right the patient found it necessary to turn his entire body. He had been treated without significant improvement with indomethacin, aspirin, phenylbutazone, infrared irradiation, and a Thomas cervical collar. Three months before admission the patient was treated with cervical halter traction which increased his pain. During the two week period before admission he had noted dizziness while turning in bed, occasional dysphagia, and episodic loss of voice. These attacks seemed to occur after prolonged standing and were preceded by hoarseness and sub-occipital pain. He would then rapidly lose volume until complete aphony ensued. There was no dysthria or dysphasia preceding the aphony. He then found that he could relieve his symptoms within seconds by lying on his right side. He had also complained of occasional difficulty with mastication and movement of his tongue at these times. Infrequently, there was a sensation of coldness in his pharynx. He denied vertigo, diplopia, transient blindness, blurring of vision, or emesis.

On examination his vital signs were normal. His mouth and throat were normal. A mild bilateral exophthalmos was present. His neck was immobile and non-tender and there was no palpable thyroid enlargement. Carotid pulses were full and equal and there were no bruises. His spine was rigid and a moderate dorsal scoliosis was present. The head was held in some degree of forward flexion with rotation to the right. His pupils reacted equally to light and fundoscopic examination was normal. Anisocoria was present, the right pupil being 1 mm larger than the left. Cranial nerves IV-VIII were normal. The gag reflex was depressed bilaterally. Phonation, deglutition, and movement of the palate, uvula, and tongue were normal. Motor examination revealed a
weak left psoas and quadriceps femoris and weak trapezius and deltoids. A mild intention tremor was present on finger-to-nose testing bilaterally. A hypotonic left knee-jerk was present. There was no Babinski response. Peripheral and central sensory examinations were normal.

Radiographs of the cervical, dorsal, and lumbar spine demonstrated the changes of ankylosing spondylitis with calcification and new bone formation in the interspinous and supraspinous ligaments. Cinefluoroscopy of the cervical spine revealed a maximum of 2° of lateral flexion and 1° of flexion-extension, and this was considered to be at the cervicodorsal junction. Lateral tomography of the posterior fossa and upper cervical spine revealed atlanto-occipital subluxation and fusion of the C1, C2, and C3 vertebrae (Figure). The anterior border of the posterior arch of C1 vertebra to the anterior border of the foramen magnum measured 2.5 cm. The top of the dens was 1 cm above Chamberlain's line. Transfemoral vertebral arteriography demonstrated an elongated, tortuous vertebral artery on the right with marked narrowing to a thin, thread-like structure over the 1st cervical vertebra at the foramen magnum and the complete absence of any intracranial component of the left vertebral artery. Carotid angiography showed filling of the posterior cerebral and superior cerebellar circulations. A lumbar puncture could not be performed because of the severe degree of laminar hypertrophy and posterior bone formation.

During his hospitalization he was examined during several attacks of aphony. In the first episode he initially became hoarse, the force of his phonation decreased to a whisper, and within three minutes he was unable to speak although he could form words with his mouth. This was associated with an increase in suboccipital discomfort. At this time there was no change in his deep tendon reflexes and the plantar responses remained flexor. Strength was equal in the upper and lower extremities except as previously noted. His pulse and blood pressure were normal. On protrusion, the tongue deviated to the right. The patient then lay on his right side and within 15 seconds normal volume had returned but he was noted to be somewhat dysarthric, with a persistent XIIth nerve palsy. He recovered fully in one minute.

A second attack was observed while the patient was in a four-poster brace. At this time there was no increase in pain before the attack, but the brace in no way altered the other symptoms and signs of his usual syndrome. He was discharged shortly thereafter in the brace.

He was again admitted approximately three weeks later. He reported good early relief of his symptoms with the brace. There had been less pain and about...
six modified or abortive episodes since his discharge. Two days before admission he noted increasing suboccipital pain with radiation to the shoulders. Neurological findings were unchanged from the first examination. One attack was witnessed, which was again associated with a right XIIth cranial nerve palsy. It was felt that fusion was not indicated as there was no demonstrable cervical mobility. In addition, the distance from the prominent occipital bone to the lordotic cervical spine was felt to be prohibitive for adequate exposure for a stabilizing procedure. Further adjustments were made in the four-poster brace and the patient was discharged. He was re-admitted for a third time for a recurrence of his ‘spells’ in April of the same year. On examination at this time a left sided suboccipital bruit, a mild right XIIth nerve paresis, and a right extensor plantar response was noted at rest. Attempted voluntary movement of the head resulted in an audible clicking sound. Cerebellar, central, and peripheral sensory examinations were again within normal limits. The patient reported less pain, but still had frequent attacks of aphonia. Repeat cervical spine radiographs were unchanged. Atlanto-occipital tomography was repeated and did not demonstrate any changes from those tomograms taken at an earlier date. Daily brace adjustments were made by the orthopaedic consultant and the patient was again discharged. He was last admitted one month later, in May 1970, for further evaluation. Cervical radiographs were unchanged. Additional brace adjustments and fittings were made. A short trunk support with a universal joint on the front chin piece was applied, with adjustable Velcro straps used to connect the front and back head pieces. Better padding and contouring was obtained and he was discharged. After this there was a rapid decrease in the severity and frequency of attacks. By June 1970, the patient was symptom free. In July the XIIth nerve paresis was no longer seen and there was no bruit detectable. Both plantar responses were flexor. In September of the same year he noted mild hoarseness at times but had no other overt attacks and he returned to work. He was last seen in January 1972, and had been working for over a year. There had been no further episodes and his neurological examination was normal. He continued to wear the brace full time.

**DISCUSSION**

Atlanto-occipital subluxation has not been recorded as a complication with neurological sequelae in recent reviews of ankylosing spondylitis (Edeiken et al., 1964; Hart, 1968; Meijers et al., 1968; Kanefield et al., 1969; Calabro and Maltz, 1970). Subluxation at this joint has occasionally occurred in association with trauma (Bell, 1969; Evarts, 1970) or on a congenital basis (Nicholson, 1968). In the case reported by Bell, the anterior margin of the foramen magnum was displaced posteriorly on the atlas after an accident. There was resultant excessive mobility at this joint associated with the symptoms of an intermittent basilar artery insufficiency (Bell, 1969). These symptoms were relieved by a posterior fusion. None of the four patients recorded by Nicholson with anomalies of the occipitovertebral articulation had an associated neurological deficit (Nicholson, 1968).

Ankylosing spondylitis is commonly associated with neurological deficits primarily due to disease affecting the lumbosacral spine. These complications have been attributed to meningeal and epidural changes including arachnoiditis, arachnoidal cysts, and hypertrophy of the ligamentum flavum. The syndromes reported have included monoradiculopathies, elevated spinal fluid protein, pseudo-tabes, spastic paraparesis, and, most commonly, a cauda equina syndrome associated with arachnoiditis and arachnoidal diverticulae (Matthews, 1968). Secondary neurological deficits have also occurred from associated destructive bone and disc disease.

We have been able to find only two instances in which the atlanto-occipital joint has been subluxated in ankylosing spondylitis. There have been no reported cases to date of atlanto-occipital subluxation in rheumatoid arthritis. Martel’s radiographic review briefly recorded one instance of partial atlanto-occipital subluxation in a 46 year old man which was not apparently associated with a neurological syndrome (Martel, 1961). The case report detailed by Coste and co-authors, writing more than a decade ago (Coste et al., 1960), was in many respects similar to the patient described here (Table). Their patient was a 58 year old man with a 38 year history of ankylosing spondylitis. In 1956, after an automobile ride, he experienced the sudden onset of aphonia. This was associated with emesis and lasted for 30 minutes. These episodes recurred and were initially associated with driving or riding in a car. Within a four year period, however, the attacks had increased in severity and frequency so that the patient was experiencing
three of these episodes daily. These attacks were preceded by an aura of a sensation of cold about the face and larynx. The attack was relieved within five to 10 minutes by lying down. His neurological findings when he was not undergoing an attack included glossal fibrillation and a right Babinski sign. While on prolonged bed rest he remained attack-free, but continued to have intermittent oesophageal spasm, hoarseness, difficulty with swallowing and pronouncing words. The diagnosis of atlanto-occipital subluxation was made by tomography. The patient was treated by occipitovertebral fusion and a hyperextension plaster jacket. He became symptom-free, but continued to demonstrate the same signs on physical examination. Arteriography was not performed and there is no note recorded on the degree of cervical mobility present. The patient’s symptoms were felt to be due to direct medullary compression and that both osteoporosis and dorsal kyphosis were ultimately responsible for inducing the atlanto-occipital subluxation. With anterior projection of the head and neck, they considered that there would be increased stress on the superior portion of the the cervical spine. When added to the effort of holding the head extended while sitting or standing they felt that a superior cervical hyperlordosis would be produced with stretching of the ligaments involved in stabilizing the atlanto-occipital juncture.

Of the various aetiological theories which might be entertained in our patient, one might consider either vascular insufficiency (involving the short and intermediate median perforating branches of the basilar artery, anterior median spinal artery and anterior spinal rami which supply the nucleus ambiguus, the hypoglossal nucleus, and the rootlets of cranial nerves X, XI, and XII) or direct compression of the vagus and hypoglossal nerves. In both instances, the stress of the erect position with attempted cervical hyperextension might result in a shift of the central nervous system or an alteration of the bony and ligamentous relationships in the region of the foramen magnum. A vascular compressive hypothesis was suggested in our patient by the arteriograms which demonstrated definite involvement of both vertebral arteries. The narrowed segment of vertebral artery was ipsilateral to the side of glossal paresis, and one might expect that displacement at this level would affect the fine median perforating branches early on and not the parent trunk. A transient focal ischaemia at this site due to end-vascular compression could conceivably interfere with the stem nuclei or nerve rootlets involved with deglutition and phonation and with glossal movement. The absence of symptoms of basilar insufficiency can be readily explained by the perfusion of the remainder of the posterior system by the anterior circulation and by the residual single compromised vertebral artery. No comment can be made about this possible aetiological mechanism in the other reported case as arteriography was not carried out.

The possibility of mechanical compression of the vagus and hypoglossal nerves can also be considered. However, we were unable to demonstrate any movement at any of the joints involved and the only evidence that such might be possible was the audible ‘click’ that the patient associated with his attacks. Without this, one would have to postulate that all of the movement was done by the brain-stem and cranial nerves, rather than by the musculo-skeletal system. In addition, the presence of a complete aphony rather than hoarseness, associated with a unilateral glossal paresis
perhaps makes a hypothesis of ‘direct compression’ less tenable than one suggesting focal end-vessel vascular insufficiency.

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REFERENCES