Lesson of the Week

Misleading symptoms and signs of cervical spine subluxation in rheumatoid arthritis

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Radiological evidence of cervical spine subluxation is found in 30 to 40% of patients in hospital with severe deforming rheumatoid arthritis, and of these, 2 to 5% have typical symptoms and signs of cord compression.^{1,2} During the past 10 years we have seen four cases in whom the presenting neurological features were atypical and misleading.

Case reports

Case 1—In January 1972, a 47-year-old woman with rheumatoid arthritis complained of three weeks of tingling down the right side, weakness of the left arm and leg, constipation, and decreased frequency of micturition. Examination confirmed weakness and exaggerated deep tendon reflexes and Babinski response on the left side. Proprioception and vibration sense were normal. Pain and temperature sense were absent over the right arm and leg, the right side of the trunk, and the neck, jaw and occiput, consistent with the diagnosis of incomplete Brown-Séquard syndrome (figure). X-ray films showed 14 mm of atlantoaxial subluxation in flexion. Myelography showed a complete block at C1/2. Atlantoaxial fusion was performed, which resulted in improved bowel and bladder function. Signs of the Brown-Séquard syndrome persisted, however.

Case 2—In October 1980, a 62-year-old man with rheumatoid arthritis complained of three to four weeks of progressive numbness, weakness, and clumsiness in both arms and legs. On examination his arms and legs were weak and hypertonic, with exaggerated deep tendon reflexes, bilateral ankle clonus, and Babinski responses. Sensation to light touch and pin-prick was diminished over the arms and legs distally in a glove-and-stocking distribution (figure). X-ray examination showed subluxation of C3 on C4. Myelography showed a complete block at C4. Anterior fusion of C3-5 was performed and sensation returned to normal. Power increased somewhat, though mild ankle clonus and upgoing plantar reflexes persisted.

Case 3—In September 1980, a 70-year-old woman with rheumatoid arthritis complained of dyspnoea beginning a few days earlier. On direct questioning she had had occipitocervical pain and tingling in both hands for three weeks. Movement of the cervical spine was limited in all directions by pain. Her arms and legs were generally weak and hyperreflexic. Plantar reflexes were normal. A sensory level for light touch and pin-prick was present at T6 (figure). X-ray examination showed appreciable atlantoaxial subluxation. Fusion of C1-2 was performed. After operation she lost all voluntary movements and developed up-

Considering cervical spine subluxation as the cause of neurological deficit is important in patients with rheumatoid arthritis

going plantar reflexes with bilateral ankle clonus. Traction was applied and she was given dexamethasone to no avail. She died eight weeks later.

Case 4—A 67-year-old woman with rheumatoid arthritis was admitted in March 1978 having had three or four "drop attacks."

The major presenting symptoms and neurological signs in four cases of cervical spine subluxation in patients with severe deforming rheumatoid arthritis.

There were no abnormal neurological signs. X-ray examination showed subluxation of C1 on C2 and C2 on C3. Fusion of C1-C3 was performed, though the subluxation was not reduced. Immediately after operation she developed acute respiratory failure requiring intubation. She fully recovered by that evening, but during the next three weeks she had recurrent episodes of loss of consciousness associated with apnoea and tachycardia. She died four weeks later in cardiorespiratory arrest.

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Comment

These four patients show the wide range of syndromes with which cervical subluxation may present and emphasise the importance of considering the neck as a cause of neurological deficit in patients with rheumatoid arthritis. The Brown-Séquard syndrome has been reported only once before in association with atlantoaxial subluxation due to rheumatoid arthritis.³ Proprioception and vibration sense were retained in the first patient, reflecting asymmetric anterior compression of the spinal cord by the subluxed odontoid peg. Sensory deficit in a glove and stocking distribution in the patient in case 2 could have been confused with the peripheral neuropathy of rheumatoid arthritis, but it cleared with cervical spine traction and fusion, suggesting that it was due to cord compression. A similar case was reported by Marks and Sharp.⁴ The presenting symptom of dyspnoea was most unusual; only on further questioning and physical examination did the more usual features of cervical cord compression become evident. "Drop attacks" in association with cervical subluxation are ominous and have been described before in association with atlantoaxial subluxation; our fourth case serves as a reminder. The attacks are usually attributed to brainstem ischaemia secondary to vertebral artery compression,

which may also explain the respiratory and cardiovascular disturbances in these patients.² Myelography was not performed in the patients in cases 3 and 4. Myelography is advisable in all such cases because the rheumatoid changes may not be confined to the atlantoaxial joint, and the major source of compression may be at a lower level.⁵

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References

- ¹ Nakano KK. Neurologic complications of rheumatoid arthritis. *Orthop Clin North Am* 1975;**6**:861-80.
- ² Stevens JC, Cartledge NEF, Saunders M, Appleby A, Hall M, Shaw DA. Atlanto-axial subluxation and cervical myelopathy in rheumatoid arthritis. *Q J Med* 1971;**64**:391-408.
- ³ Boyle AC. The rheumatoid neck. *Proc R Soc Med* 1971;**64**:1161-5.
- ⁴ Marks JS, Sharp J. Rheumatoid cervical myelopathy. *Q J Med* 1981;**50**:307-19.
- ⁵ Anonymous. Rheumatoid atlantoaxial subluxation. *Br Med J* 1976;**ii**:200-1.

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For Debate . . .

Impact of maternal serum alpha-fetoprotein screening on antenatal diagnosis

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Abstract

An analysis has been made of indications for amniocentesis in the Edinburgh area from 1979 to 1981. About 5% of all mothers underwent the procedure. Among 2137 amniocenteses, 37% were performed on mothers 35 years old or more, and 30% on patients with raised serum α -fetoprotein. The total number of amniocenteses and the categories have been stable for the past three years. As a result of amniocentesis 104 pregnancies were terminated, 66 of which (63%) followed a raised maternal serum α -fetoprotein indication, while only 10 (9.6%) were in mothers aged 35 or more. There were a further 12 terminations based on raised serum α -fetoprotein but where no amniocentesis had been thought necessary. Even when figures for anencephaly are excluded from the analysis, maternal serum α -fetoprotein screening was responsible for detecting 35 out of 63 (56%) abnormal fetuses. This constitutes a strong case for the continuation of α -fetoprotein screening programmes.

Introduction

One of the aims of early antenatal diagnosis is the reassurance of parents who have an increased risk of bearing a child with a serious birth abnormality. The other aim is to detect fetal disorders early enough to allow the possibility of termination of pregnancy. Though reassurance is important in counselling and obstetric management, the effectiveness of antenatal diagnosis must in the end be judged by its ability to reduce the birth prevalence of serious disorders. One way of judging effectiveness is to examine current indications for amniocentesis and the frequency with which they allow abnormal fetuses to be detected. In this report I have analysed experience in one part of Scotland over the past three years.

Materials and methods

Antenatal diagnosis in Edinburgh is carried out in five separate laboratories. All amniotic fluids are referred to my laboratory for α -fetoprotein analysis, and it has therefore been possible to construct and maintain a central register of indications for amniocentesis and outcomes of pregnancy for the whole region. I have analysed the indications for amniocentesis and the terminations of pregnancy resulting from laboratory determinations for the years 1979-81. The three most recent years are chosen since the increasing demands on diagnostic services seen in the early and mid-1970s appear to have reached a plateau. The analysis is restricted to samples taken in south-

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