MEDICAL PRACTICE

Hospital Topics

Nervous System Involvement in Ankylosing Spondylitis

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Summary

In a review of 45 patients with ankylosing spondylitis 10 had neurological symptoms and signs and three of them had two separate neuropathological disorders. The neurological profiles fell into five main categories—multiple sclerosis, the cauda equina syndrome, focal epilepsy, vertebrobasilar insufficiency, and peripheral nerve lesions. An association between ankylosing spondylitis and multiple sclerosis is suggested, possibly due to an immunological defect or to one being a complication of the other.

Introduction

Ankylosing spondylitis is usually considered to be a disease of the spine with few extra-articular manifestations except iritis and aortitis. The nervous system has been thought to be affected only rarely as a result of atlantoaxial subluxation or spinal fractures. More recently a syndrome suggestive of compression of the cauda equina has aroused much interest. Though it has been described in only a few patients,^{1,2} this probably does not represent its true incidence. This study was started because neurological complications seemed to be common in a group of patients with ankylosing spondylitis who were attending a general medical outpatient clinic.

Patients

All 45 patients with ankylosing spondylitis who were referred to a general medical clinic during the previous four years were reviewed. In each case the clinical diagnosis was confirmed by the radiological appearances of the spine and sacroiliac joints. All were men aged 24 to 72 years (mean 49 years) and had had symptoms for 1 to 30 years (mean 19.7 years). Altogether 38 had received spinal radiotherapy. All underwent thorough clinical examination and further laboratory and radiological investigation where indicated. The haematological and biochemical data on most of these patients were reported previously.³

Of the 45 patients 10 had neurological symptoms and signs. Their cases are reported below.

Multiple Sclerosis

Two patients were found to have both ankylosing spondylitis and a syndrome resembling multiple sclerosis.

Case 1.—A 25-year-old commercial traveller started to have symptoms of ankylosing spondylitis eight years previously, and two years later suffered an episode of diplopia associated with cerebellar dysfunction which lasted several weeks. Three years after this he had several right-sided focal fits with clonic movements starting in his leg and spreading to arm and face, though none became generalized. Initially they occurred frequently but after a few days gradually ceased. There was no recurrence of fits and he was never given anticonvulsants. More recently he had variable weakness and incoordination of his limbs, with numbness of both legs. Examination showed gross nystagmus on lateral gaze, with cerebellar ataxia in all four limbs; both legs were spastic, with extensor plantars and a sensory level at T6. He was just able to walk unaided. Cerebrospinal fluid examination showed a normal cell count but the protein was raised at 60 mg/100 ml and the gamma-globulin fraction was also raised at 20%. No E.E.G. was recorded during a fit but the interictal E.E.G.s were normal.

Case 2.—A 26-year-old man who had had ankylosing spondylitis for nine years was found to have diminished visual acuity in the left eye, with pallor of the optic disc. There was horizontal jerky nystagmus in all directions of gaze, slight ataxia, weakness and hypertonicity of his limbs with brisk reflexes, bilateral ankle

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clonus, and absent abdominal reflexes. No cerebrospinal fluid results were available.

Cauda Equina Syndrome

Only one patient had the fully developed cauda equina syndrome but a second had symptoms and signs, which suggested that he might have been in the early stages of its development.

Case 3.—A 65-year-old man had suffered from low back pains for 40 years and for 10 years had had difficulty controlling bladder and bowel. The latter necessitated his removing faeces manually through his lax anus. Five years previously his urinary symptoms had been treated by a transurethral prostatic resection which removed the retention component but resulted in a constant dribbling incontinence for which he wore a clamp on his numb penis. At examination his buttocks were wasted, ankle jerks were absent, and sensation was lost in all the sacral dermatomes. The only abnormality on myelography was a posterior diverticulum in the lumbar theca. Laminectomy was not performed.

Case 4.—A man aged 50 who had had ankylosing spondylitis for 18 years presented with a short history of urgency and urge incontinence of urine. There was obvious wasting of one buttock but no sensory abnormality. During the two-year period of observation his symptoms did not progress.

Focal Epilepsy

Two patients had focal epilepsy; one of these, case 1, with multiple sclerosis, has already been described. The other was case 5.

Case 5.—A 57-year-old man had a 32-year history of ankylosing spondylitis. In 1971 over two or three days he had several fits, starting with clonic movements of his left hand, spreading to the arm and face, and becoming generalized. There were no abnormal neurological signs except hypoaesthesia in the distribution of C2 and C3 on the left, where he had suffered paraesthesia for some months. The skull x-ray picture and a brain scan were normal. E.E.G.s showed bilateral slow waves but no focus. No further symptoms or signs appeared.

Spastic Paraparesis

Case 6.—A 52-year-old man with ankylosing spondylitis for 22 years noticed increasing weakness of the legs. When aged 32 he had received radiotherapy in a dose of 1,500 rads to the whole spine. On examination his spine was found to be completely ankylosed, with an appreciable dorsal kyphosis and loss of cervical lordosis, so that he was unable to look up. The only neurological abnormality was a spastic paraparesis with extensor plantar responses. There was no sensory level. All abdominal reflexes were absent. The arms were normal. A slow spontaneous improvement occurred, and six months later he was able to walk unaided though his signs remained. Unfortunately he also had the bilateral upper lobe fibrosis sometimes found in ankylosing spondylitis and died of bronchopneumonia shortly afterwards. Permission for necropsy was not obtained.

Vertebrobasilar Insufficiency

The considerable bony involvement of the cervical spine in most cases of ankylosing spondylitis might be expected to produce symptoms of vertebrobasilar insufficiency in some patients. In both our patients with this problem cervical ankylosis was not complete.

Case 7.—A 56-year-old painter and decorator had been able to work throughout his 28 years with ankylosing spondylitis until the onset of giddy attacks. He had had these for two years. They were usually precipitated by neck movements and were associated with true vertigo. No nystagmus was present at rest but both his symptoms and the nystagmus could be produced by rotating his neck in extension. The only other abnormal neurological signs were weakness of plantar flexion of the left foot with an absent ankle jerk. Cervical spine radiographs showed changes of ankylosing spondylitis with gross bony involvement at the articular facets at

several levels. Vertebral arteriography was not attempted but it is reasonable to assume that both vertebral arteries were kinked forwards by this new bone formation.

Case 8.—A 58-year-old motor mechanic presented similarly with inability to continue working because of attacks of vertigo and visual disturbance. Nystagmus was present with his neck in the neutral position and he showed gross intention tremor and ataxia of gait in between attacks of giddiness. His symptoms were controlled to some extent by a collar.

Peripheral Nerve Involvement

Four men had peripheral nerve signs. Two of these were mentioned above—case 5 with focal fits and C2 and C3 distribution of hypoaesthesia, and case 7 with vertebrobasilar insufficiency and an absent ankle jerk with weak plantar flexion. The other two were as follows.

Case 9.—A 37-year-old man had weakness of the small muscles of the left hand and hypoaesthesia of C5 distribution on the right. He had not received radiotherapy to his cervical region. Radiographs of his neck showed changes of ankylosing spondylitis but no bony encroachment on the intervertebral foramina. His symptoms developed over several weeks but over the subsequent two-year period of observation there was no further deterioration.

Case 10.—A 49-year-old man who had intermittent low back pain throughout his 2-year history developed an annoying numbness on the dorsum of the right foot. Examination showed diminished sensation in the L4 and L5 dermatomes with an absent right ankle jerk.

Three further patients had considerable muscle wasting, two of the buttock and thigh and one of the shoulder girdle. They had all, however, experienced some pain in the neighbouring joint, and radiographs showed loss of cartilage and other arthritic changes.

Discussion

The occurrence of multiple sclerosis in two patients in this series may seem coincidental. If, however, the incidence of multiple sclerosis is taken to be 5 per 10,000⁴ the chance of any group of 50 people containing one case of multiple sclerosis is 1 in 40, and for two cases it is 1 in 1,600 ($P < 0.001$). In addition the only patient with ankylosing spondylitis admitted to the Midland Centre for Neurosurgery and Neurology in the past three years had multiple sclerosis. In a series of 59 cases of ankylosing spondylitis reviewed for neurological abnormalities² one definite and two probable cases of multiple sclerosis were identified. Since then one of the probable cases (case 1) has developed new symptoms and signs, making the diagnosis unequivocal.⁵ We conclude that multiple sclerosis and ankylosing spondylitis are related, perhaps by some common immunological defect, or that a multiple-sclerosis-like illness is a complication of ankylosing spondylitis. It is of interest that an association between a syndrome resembling multiple sclerosis and systemic lupus erythematosus has been reported.⁶

The cerebrospinal fluid protein level is sometimes raised in the acute stages of ankylosing spondylitis^{7, 8} but not in the chronic case.² It is not known whether the cerebrospinal fluid gammaglobulin is also raised. In the one patient in our series with both ankylosing spondylitis and multiple sclerosis who had a lumbar puncture and in the man seen at the Midland Centre for Neurosurgery and Neurology the cerebrospinal fluid protein and the gammaglobulin fraction were raised.

It is possible but not likely that the paraparesis in case 6 was due to radiation myelopathy. This is a well-recognized complication of spinal irradiation but symptoms typically develop after about a year, though they may present from one month to five years after treatment and then progress. In our patient symptoms developed after 20 years and then improved. Demyelination may have been responsible as it was in Matthews's third case,² in which the patient had a spastic paraparesis and at necropsy was found to have demyelination of the thoracic cord.

Two of our patients had focal epilepsy, and in each a series of attacks occurred over a short period of time and did not recur. We have no explanation for this. Focal attacks are a complication of multiple sclerosis, and one of the patients also had this disease.

Posteriorly situated diverticula in the lumbar theca must now be regarded as a characteristic myelographic appearance in the cauda equina syndrome of ankylosing spondylitis. They were present in both cases described by Matthews² and in the myelogram of McGill's case.⁹ In one of Matthews's patients necropsy showed the diverticula extending backwards and encroaching extensively on the laminae and spinous processes. The arachnoid cysts that are being recognized increasingly with more frequent screening of iophendylate in the supine position do not involve bone.¹⁰

The finding of nerve root lesions in the presence of extensive bony abnormalities of the spine may not be surprising at first sight. It becomes so when it is realized that bony ankylosis does not encroach on the intervertebral foramina.¹¹ We have confirmed this.

Direct nerve compression by abnormal bony structures does not seem to be the major cause of the spinal cord and peripheral nerve signs seen in ankylosing spondylitis. In fact, there are probably several different mechanisms, including associated disease—for example, demyelination; interference with blood supply, perhaps from radiation or possibly from arteritis; and areas of arachnoiditis, occurring earlier in the disease and resulting in atrophy. The last is likely because patients usually

develop their symptoms over a few months and then do not progress further. The cerebrospinal fluid protein level is raised in some acute but not in chronic phases, and the laminectomy and necropsy findings in the cauda equina syndrome reviewed by Matthews² suggest arachnoiditis followed by atrophy. More operative and necropsy observations are necessary to clarify the aetiology.

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Medicine in Old Age

Gastrointestinal Problems in the Old—II

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Acute Abdomen

The acute abdomen often causes considerable problems in elderly patients. Its presentation may be atypical, its course more stormy with later diagnosis and a much greater likelihood of complications—especially at and after surgery.

Old people are often less able to cope with their illness than are younger patients, so that early diagnosis becomes of major importance; yet these are often the very patients who are diagnosed late because of difficulties in communication, few or atypical symptoms, absence of "classical" physical signs, and difficulty in physical examination due to associated degenerative or disease processes. Pain may be absent or

unimpressive and the description of its type and site is often vague. Abdominal rigidity, and even guarding, may be absent where it would certainly have been present in earlier years, and in such patients the presence or absence of gut sounds can become a most important physical sign, the situation being in many ways analogous to the acute abdomen developing in a patient being treated with steroids.

Complications occur frequently: thus, complications of acute appendicitis are found at operation five times more often in the old than in younger patients. Peritonitis from whatever cause is less effectively kept localized in elderly patients. The mortality of major operations is increased several-fold, especially if the operation is performed as an emergency. The mortality rate increases in the elderly as age advances.

It is a good general rule to admit to hospital elderly people with severe abdominal pain, even if there are no physical signs. It must be remembered that many medical causes of acute abdominal pain may occur in the elderly, including acute pancreatitis, diabetic ketosis, tabes dorsalis, Addison's disease, hypercalcaemia, haemochromatosis, or porphyria. Furthermore, abdominal pain may be due to extra-abdominal disease—for example, pleurisy, spondylosis, zoster, etc.—and these are common in old people. For an excellent review of the acute abdomen in the elderly, see Charlesworth.¹

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