CAUDA EQUINA AND SPINAL CORD COMPRESSION
IN PAGET'S DISEASE

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Paget's disease of the spine seldom causes symptoms. Backache is uncommon, and the
complication of cauda equina or spinal cord compression, as in the three cases to be described,
is rare. Indeed it has been recorded in only forty-eight patients in the literature up to 1964.

Sir James Paget first described the condition of osteitis deformans in 1877. In the advanced
stages of the disease he noted that "the spine, whether by yielding to the weight of the overgrown
skull, or by changes in its own structures, may sink and seem to shorten with greatly increased
dorsal and lumbar curves." Wyllie (1923) first recorded two patients with Paget's disease
causing spinal cord compression. In the same year a similar case was reported by Ardin-Delteil,
Azoulay and Lagrot (1923). Comprehensive reviews with the addition of further examples
were published by Schwarz and Reback (1939) and Turner (1940). Cases were added by
Hillman (1941), Gross (1942), Whalley (1946), Godlewski (1948), Colclough (1949), Fairburn
(1951), Teng, Gross and Newman (1951), Latimer, Webster and Gurdjian (1953), and
Ramamurthi and Visvanathan (1957). Schreiber and Richardson (1963) reported the only
example so far recorded in which compression of the spinal cord was produced by disease of a
single vertebra. Surprisingly, none of these reports was in a journal primarily concerned with
orthopaedic surgery, and therefore this condition may not be well known to orthopaedic
surgeons.

During the past sixteen years three patients with Paget's disease of the vertebral column
and neurological involvement have been seen at the Middlesex Hospital. One of these (Case 3)
has already been reported (Fairburn 1951). In two patients the lumbar spine was involved,
and in one there was extensive thoracic and upper lumbar spine disease. In all three patients
laminectomy produced improvement, as it did in over half of those previously reported.

CASE REPORTS

Case 1—A man aged fifty-seven had been troubled by backache for about ten years and by
left-sided sciatica for five years. He was referred to the orthopaedic clinic because of weakness
of the legs and unsteadiness.

On examination he was thin and looked ill. The spine showed loss of the normal lumbar
curve with a slight kyphos at the level of the third lumbar vertebra. All movements were
markedly restricted, particularly flexion and extension. All the muscles of the lower limbs were
wasted, the quadriceps most of all. Straight-leg raising was permitted on the right to 90 degrees
and on the left to 70 degrees. There was no sensory disturbance. There was outward bowing of
both tibiae and all movements of the left hip were restricted.

Investigations—Radiographs showed the typical changes of Paget's disease in the first and
second lumbar vertebrae (Fig. 1). The body of the second lumbar vertebra was enlarged in
its antero-posterior diameter and its height was reduced. The rest of the spine showed no
evidence of Paget's disease, but the whole of the left side of the pelvis and the proximal half
of the left femur were affected.

The serum calcium and phosphorus levels were normal. The serum alkaline phosphatase
was 59 King-Armstrong units.
Lumbar puncture was difficult but produced clear and colourless cerebrospinal fluid. The pressure was 200 millimetres; bilateral jugular compression did not produce any effect. The protein level was 700 milligrams per 100 millilitres.

At myelography the contrast medium could only be made to travel as far as the upper border of the third lumbar vertebra where there was complete obstruction (Fig. 2).

It was concluded that there was cauda equina compression at the level of this vertebra. Operation (Mr P. H. Newman)—In 1962 the laminae of the first, second and third lumbar vertebrae were removed. When first exposed the dura mater did not pulsate, but by the end of the operation pulsation had returned.

Progress—The patient was immediately relieved of all pain and the weakness of the legs soon disappeared. However, within two months some of the old pain returned. After further admission to hospital and trials of various analgesic drugs he was finally able to resume work as a joiner six months later.

Two and a quarter years after the operation the patient was readmitted for further laminectomy as pain in the lower lumbar region and left buttock was becoming intolerable. Examination of the nervous system was normal except for an exaggerated left knee jerk.

The previous operation wound was reopened. The dura mater was found to be kinked at the level of the lower border of the third lumbar vertebra and laminectomy of the fourth vertebra was carried out. This removed the constriction.

The patient again made excellent progress and was relieved of all the pain in the left lower limb. He was discharged from hospital wearing a lumbo-sacral corset and four months after the second operation was free of symptoms, though he has become increasingly incapacitated by pain in his left hip.

Case 2—A man aged fifty-seven had been troubled for some years by upper thoracic backache which had gradually become worse over the past two years. Walking or lying flat made the pain more severe, and recently coughing had begun to cause pain in both lower limbs. Bending and stooping aggravated the pain and he had been unable to work as a labourer for five months.
On examination he was a small, wiry man with no obvious clinical evidence of Paget's disease. There was a kyphos in the mid-lumbar region and the range of lumbar spine movement was about half normal. There was tenderness over the spine of the sixth thoracic vertebra. The central nervous system appeared normal.

InVESTIGATIONS—Radiographs showed the third lumbar vertebra to be extensively involved by Paget's disease, with flattening of the body and an increase in its antero-posterior diameter (Fig. 3). No other vertebrae were affected. A skeletal survey failed to reveal Paget's disease elsewhere. The calcium and phosphorus and serum alkaline phosphatase levels were within normal limits.

At lumbar puncture the cerebrospinal fluid was clear and the pressure was 140 millimetres. Bilateral jugular venous compression caused a free rise and fall of the pressure. The protein content was 60 milligrams per 100 millilitres.

Myelography revealed a block of the cerebrospinal fluid column at the level of the deformed third lumbar vertebra (Fig. 4). It was concluded that there was compression of the cauda equina at the level of the third lumbar vertebra, and that tension of the dura mater over the greatest convexity of the thoracic kyphosis was responsible for the upper thoracic pain.

Operation (Mr P. H. Newman)—When the lamina of the third lumbar vertebra was removed there was no pulsation of the dura mater. However, when the laminae of the adjacent second and fourth vertebrae had been nibbled away, the dura mater lay free from constriction and pulsation returned.

Progress—After operation the pain was completely relieved and after twelve months he was still free from symptoms.

Case 3—A man aged fifty-nine first noticed bowing of his back in 1945. The following year he found that his size in hats had increased. He remained well until June 1948 when he noticed difficulty in walking, with a sensation of numbness in both legs from his knees to his toes. Shortly before his admission to hospital in 1948 the numbness spread up to the umbilicus, and soon afterwards he began to have difficulty in starting micturation.
Examination showed typical appearances of Paget's disease in several bones. Neurological examination revealed a moderate symmetrical spastic weakness of the lower limbs with grossly exaggerated tendon reflexes and bilateral Babinski responses. Below the level of the umbilicus there was impaired sensibility to pinprick, cotton wool and temperature.

Investigations—Radiographs showed typical Paget's disease of the skull, femora, tibiae, clavicles and one radius. There was also involvement of the lower seven thoracic vertebrae and the first lumbar vertebra, the affected vertebrae showing varying degrees of compression (Fig. 5). Lumbar puncture showed a pressure of 75 millimetres with no rise or fall on jugular compression. The protein content was 120 milligrams per 100 millilitres. Myelography revealed a complete block at the level of the second lumbar vertebra.

Operation (Miss D. J. Beck)—In August 1948 the spines and laminae of the fourth thoracic to the second lumbar vertebra were removed. The bone was soft and cut very easily with the laminctomy forceps. No localised block was demonstrated, but as the bone was removed the dura mater bulged freely over the whole length of the exposure. It was not until eleven spines and laminae had been removed that free pulsation was observed over the whole of the exposed dura mater.

Progress—The patient made a good recovery. At the end of three months he returned to work, being then able to walk a mile. He had lost the sensation of numbness in the lower part of his body except for the great toes. The sensory level had disappeared, and apart from slight weakness of dorsiflexion at the right ankle, power was normal throughout. However, in February 1949 he again noticed increasing difficulty in walking with numbness of the feet and legs. Examination revealed spastic weakness of the lower limbs, more profound than at the time of his first admission. Radiographs of the spine showed further collapse of the bodies of the sixth and tenth thoracic vertebrae. It was decided that further decompression was indicated.

At operation the laminectomy wound was reopened. The spines and laminae of the third thoracic and third lumbar vertebrae were removed. At the site of previous operation it was found that a dense layer of tissue resembling soft bone had formed over the posterior surface of the spinal cord covering it completely. After this firm plate had been removed over the whole length of the previous laminectomy the theca again began to pulsate.

After the second operation recovery was slow but steady and the patient was again able to walk a mile after four months. He maintained a moderate level of activity for a man of his age over the next ten years until he died in December 1961 from a Paget's sarcoma of the right femur, which was proved histologically.

DISCUSSION

A study of the distribution of the lesions of Paget's disease shows that, after the sacrum, the vertebrae are the most commonly affected bones (Schmorl and Junghanns 1959). Spinal cord or cauda equina compression is not necessarily associated with gross external deformity of the spine. Compression is the result of a slow progressive narrowing of the vertebral canal from enlargement of the vertebral bodies, which bulge antero-posteriorly and at the same time are compressed in the vertical plane. Bony prominences projecting from the posterior surfaces of the bodies and enlargement of the pedicles and laminae may be contributory factors.
seen from the three patients described, neurological involvement may follow either diffuse changes effecting a considerable length of the vertebral canal or a monostotic lesion. The upper thoracic spine is the commonest site for cord compression to occur because here the vertebral canal is narrowest in relation to the diameter of the cord.

Vascular insufficiency does not appear to be of importance in this syndrome, particularly as the blood supply of the affected bones is markedly increased. Campbell and Whitfield (1943) recorded three patients in whom the compression was due to the development of an osteogenic sarcoma in the affected vertebrae.

The presenting symptoms are often those of neurological involvement rather than of bone disease (Turner 1940). There is usually a gradual progressive impairment of the functions of the spinal cord or cauda equina without remissions. In the early stages, however, the patient may have the same complaints of backache and pain in the legs as patients with disc degeneration. The diagnosis may easily be missed in an elderly patient with multiple areas of discomfort when a careful examination of the central nervous system has not been done (Robinson 1953).

Investigations may be complicated by difficulty in performing lumbar puncture due to bony fusion between adjacent laminae. This was certainly a problem in Cases 1 and 2 and has been noted by Wyllie (1923) and by Turner (1940).

Decompression of the cord or cauda equina is the only effective treatment. When the disease affects a long segment of the spine an extensive laminectomy may be necessary. At operation the laminae are thicker than normal and the bone is soft and vascular. The spinal canal is diminished in size and epidural fat is absent. The dura mater is densely adherent to the adjacent bone. When the dura mater is first exposed there is no pulsation but as the decompression is completed pulsation returns. As with all varieties of neural damage, recovery after operation may be slow. Recurrence of symptoms after relief by operation may be due to advance of the underlying Paget's disease and a second operation may be necessary, as shown here in Cases 1 and 3.

Twenty-eight of the patients recorded in the literature had laminectomies performed and five died following the operation.

SUMMARY

1. Three patients with backache and spinal cord or cauda equina compression due to Paget's disease of the vertebrae are reported; all three were relieved by laminectomy.
2. One case is of particular interest because it is only the second one reported where compression was due to a single affected vertebra.

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REFERENCES

Paget, Sir James (1877): On a Form of Chronic Inflammation of Bones (Osteitis Deformans). Medico-Chirurgical Transactions, 60, 37.
Teng, P., Gross, S. W., and Newman, C. M. (1951): Compression of the Spinal Cord by Osteitis Deformans (Paget's Disease), Giant-Cell Tumor and Polyostotic Fibrous Dysplasia (Albright's Syndrome) of Vertebrae. Journal of Neurosurgery, 8, 482.