PARAPLEGIA DUE TO PAGET'S DISEASE (OSTEITIS DEFORMANS)

BY

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Osteitis deformans, or Paget's disease, may affect any part of the skeleton in those of older years. While involvement of the spinal column is frequent, it is unusual for compression of the spinal cord to be produced. It is probable that this complication is commoner than the literature would indicate, as it occurs in an age group in which accurate assessments of the nervous system are rarely made. Two cases are described and their relationship to the usual pattern of the disease discussed.

Case 1

A woman aged 64 was admitted to Dunedin Hospital on January 21, 1952. Three and a half years ago she had begun to notice a nagging pain in the small of her back. This was followed by increasing pain in the hips and knees. About a year ago she had had burning pains in both legs, and her feet had felt numb and lifeless. Walking had become difficult, and eventually she had had to use two sticks. For a year her balance had been bad and she had had numerous falls, in spite of the sticks. She had not noticed that her legs were really weak until six months ago, when she found that she could not lift them up to get into a motor-car. Her left leg had had spasms, when it would cross over the right. She had had increasing trouble with her micturition for six months. Sometimes her water ran away without control and at other times she had great difficulty in passing it. At the time of admission she was getting about the house with difficulty. She had consulted Sir Robert Jones for bow legs at the age of 13.

On examination she was seen to be a short woman with marked bow legs. Her heart was moderately enlarged; her blood pressure was 130/65 mm. Hg. The jugular veins were not engorged. There were crepitations and rhonchi throughout both lung fields. Her ankles were not swollen, the liver was not palpable, and there were no clinical manifestations of any neoplasm. She had a moderate lower thoracic kyphosis with a slight scoliosis to the left. Apart from slight deafness the cranial nerves were normal. Both legs were very spastic, with adductor and flexor spasms that were severest in the right leg. All movements were present in the lower limbs, but the power was very poor. There was a pronounced level of hypalgesia to all forms of sensation at L1. This faded off and there was hypalgesia at T8 on the right and at T11 on the left. Vibration was absent over the spinous processes up to T10. The lower abdominal responses were absent but the upper ones were present. The tendon reflexes in the legs were exaggerated, but no clonus was obtained. Both plantar responses were extensor.

The blood picture was normal. Serum alkaline phosphatase was 12.8 units. Lumbar puncture showed a pressure of 120 mm. H₂O with a slight rise to jugular compression. The fluid contained 10 polymorphs per c.mm. and 80 mg. of protein per 100 ml. X-ray examination showed: fine osteolytic stippling of the skull; minor congestive changes in the chest; increased size of heart as in pulmonary heart disease; a mixture of sclerosis and osteoporosis of T9 and 10 with slight collapse, and similar changes at L3 (? osteitis deformans, ? secondary carcinoma); normal lower limbs. A myelogram from below showed a complete block at T11.

Operation. This was performed on January 24 (anaesthetist, Dr. J. Ritchie). To supplement the general anaesthesia, hexamethonium bromide was used as a hypotensive

agent. A low thoracic incision was centred over T10 and 11. In separating the muscles there were numerous small blood vessels passing from the muscles to the bone that had to be controlled. The laminae and spines of T9 and 10 were massive. The laminae of T9 and 10 were completely removed and part of those of T8 and 11. The thick bone was soft and very vascular. When the bone had been removed it was obvious that there was a small kyphus with its apex at T10. The spinal theca was opened and appeared commodious. There was no evidence of any tumour, and it was possible to pass a fine rubber catheter for 8 in. (20 cm.) upwards and downwards without any difficulty. The dura mater was sutured and the wound closed in layers. It was not necessary to give a blood transfusion. The pathological report (Dr. R. Rodda) on the removed bone showed the typical changes of osteitis deformans.

After the operation there was slow improvement. When discharged home on April 2 the patient was able to walk but still used two sticks; bladder function was normal; tone of both legs was normal and the power slightly reduced; pain and temperature were felt everywhere; cotton-wool sensation was diminished in both feet; vibration was absent up to the anterior superior spines; position sense was very deficient in the ankles and toes; knee- and ankle-jerks were increased; the abdominal responses were all present; the plantar responses were extensor and there was knee and ankle clonus in the left leg.

When re-examined on June 5 she was walking well and used only one stick. Movements of the knees were still a little weak, the left more than the right. There was a trace of ataxia in the left leg. Vibration was absent in both lower limbs, but all other forms of sensation were normal. The right plantar response was flexor and the left extensor. Moist sounds were heard all over the chest.

Case 2

A man aged 65 was seen in consultation on October 16, 1951. In 1941 he had had an attack of congestive cardiac failure. At that time it had been noticed that he had an advanced osteitis deformans of the skull and spine. During 1945 he was admitted to hospital again, complaining of difficulty in walking. It was noted that both legs were weak and ataxic. All forms of sensation were poorly appreciated in the legs. The knee-jerks were absent and the plantar responses were flexor. By 1947 the weakness of his legs had increased and there was bowing of the right femur. X-ray films of both thighs showed severe osteitis deformans. He was fitted with a spinal brace. In 1949 he was admitted to hospital with retention of urine. The prostate was normal to palpation. Cystoscopy showed a fibrous bladder-neck and marked sacculcation of the bladder. His bladder function improved and no active treatment was given.

He was again admitted to hospital in 1950 with urinary retention, and a transurethral resection of the prostate was done. A microscopical examination of the fragments showed senile hyperplasia. In spite of the resection he continued to have difficulty in passing his water. In the previous six months his leg weakness had rapidly increased and he had become bedridden. He had frequent attacks of breathlessness.

On examination he had a noticeably enlarged head and a very big right femur. His spine was rigid and there was an upper thoracic kyphosis. He had paraplegia in extension and was not capable of any voluntary movement in the legs. There was a marked hypalgesia to all forms of sensation to T11. The abdominal responses were absent. The plantar responses were extensor and there was bilateral ankle clonus. His bladder was palpable at the umbilicus. The blood pressure was 200/100 mm. Hg, and the apex beat of the heart was felt 1 in. (2.5 cm.) outside the nipple line. There was considerable oedema of both ankles and he had severe dyspnoea on slight exertion. X-ray examination at that time showed gross osteitis deformans of the right scapula, pelvis, femurs, lumbar spine, thoracic spine,

and tarsal bones. There was left ventricular enlargement. A diagnosis of paraplegia due to osteitis deformans of the spine was made. In view of his poor condition he was regarded as unfit for laminectomy, and a gloomy prognosis was given.

He was seen next on August 28, 1952. There had been some improvement in his general health. He no longer had attacks of breathlessness, although his ankles swelled intermittently. He had not walked for about eighteen months. He was able to pass water, but it often leaked away, and there had been attacks of cystitis. Both legs were painful and his buttocks were uncomfortable. He had noticed that his head was still getting bigger and that his dentures no longer fitted. He was a permanent inmate of a "chronic" hospital.

On examination his blood pressure was 220/120 mm. Hg, and the apex beat was felt $1\frac{1}{2}$ in. (3.8 cm.) outside the nipple line. There was no evidence of any congestive cardiac failure. His bladder was palpable at the umbilical level. He had a marked upper thoracic kyphosis. His head was big and the right femur was massive. There was a complete paraplegia in extension. There was considerable hypalgesia for all forms of sensation to T7 on the right and to T11 on the left. The abdominal responses were absent and the plantar responses were extensor. Fresh x-ray films showed that the disease had progressed and that the lumbar spine was ankylosed. The cervical spine was normal. Further, many ribs and the tibiae and fibulae were affected.

The paraplegia was of such long standing that little could now be expected from decompression of the spine. The cardiovascular reserves were small and so no operative treatment was advised.

Literature

In 1877 Sir James Paget described the bone disease that bears his name. Thereafter sporadic cases were recorded in which neurological symptoms had complicated osteitis deformans. Wyllie (1923) gave the first verified account of spinal-cord compression due to osteitis deformans of the spine. Kay, Simpson, and Riddoch (1934), besides having had two cases of paraplegia in 34 cases of osteitis deformans, had seven other cases with less severe neurological signs. Schwarz and Reback (1939), Turner (1940), and Teng, Gross, and Newman (1951) have surveyed the literature. The total number of published cases was 37. Further cases have been published, and Petit-Dutaillis (1951) gave details of three cases treated operatively. Fairburn (1951) had a case in which symptoms recurred after successful laminectomy and which was improved by further operation. Godlewski (1948) had a case of cervical spinal-cord compression successfully treated by head traction with skull callipers. With the above two instances, the total number of published cases of compression of the spinal cord due to osteitis deformans is 44.

Discussion

Teng *et al.* (1951) gave the incidence of paraplegia as 1 out of 131 cases of osteitis deformans, on hospital admissions. The true incidence would be much less, as osteitis deformans is diagnosed commonly without recourse to hospital admission. Gutman and Kasabach (1936) found 59 cases of osteitis deformans in the wards and out-patient department of a hospital that treated 200,000 new cases in the same period. Some cases were symptomless and the radiological changes were found in the course of examinations for other conditions. Osteitis deformans occurs equally in the two sexes (Kay *et al.*, 1934; Gutman and Kasabach, 1936). Spinal complications have usually been recorded in males. Case 1 is exceptional in being female, and is the fourth such case. The age spread has been from 36 to 77 years, with a maximum incidence in the sixth and seventh decades. In the spine, Paget's disease is most frequent in the lumbar vertebrae and then in the thoracic vertebrae; affection of the cervical spine is uncommon

(Gutman and Kasabach, 1936). Paraplegia develops mainly in patients with thoracic spine involvement. Wyllie (1923) showed that there was an overall enlargement of all components of the vertebrae with the exception of the spinal canal. This was consistently narrowed. The most frequent site for the paraplegia was in the upper half of the thoracic spinal cord. Schwarz and Reback (1939) indicated that, for the whole spine, the interpeduncular spaces were narrowest between the third and tenth thoracic vertebrae. There is the additional factor of a kyphus due to collapse of the softened body of the vertebra. This is well shown in Case 1, in which the spinal-cord compression occurred at the apex of a small kyphus. The vertebrae affected were the ninth and tenth thoracic, which is lower than usual. The roominess of the lumbar spinal canal is the reason why narrowing in this area rarely causes nervous signs. Case 1 had incidental involvement of the third lumbar vertebrae, but a rubber catheter was able to pass this site without difficulty. It is rare for the whole of the thoracic and lumbar spines to be affected, as in Case 2. The ultimate fusion of the adjacent vertebrae occurs when the intervertebral disk is replaced by the pathological bony process (Turner, 1940).

The usual history is of a slow and persistent interference with the functions of the spinal cord. The onset is insidious, and may be masked by the pains and failing mobility that characterize many cases of osteitis deformans. The duration of symptoms has been a year in most cases. Both of the above cases had long histories; in Case 1 it was three and a half years, and Case 2 had had osteitis deformans for seven years and neurological symptoms for six of them. Otherwise the progression of symptoms was typical in both cases. It was unfortunate that more attention was not paid to the nervous system in Case 2 when the bladder symptoms became troublesome. There are no special features of the spinal-cord compression except for the predilection for the upper thoracic segments.

The radiological features of osteitis deformans are well known. In Case 2 the emphasis was on an osteoporotic expansion of the many bones involved. There was radiological evidence that the whole lumbar spine was ankylosed. It may be difficult to differentiate the localized form of osteitis deformans from secondary deposits due to cancer. In Case 1 there was a combination of a sclerosis and a porosis with expansion of the pedicles, arches, and spinous processes of the affected vertebrae. On radiological grounds a carcinomatosis of the spine could not be excluded.

The condition most likely to cause confusion in the diagnosis is a benign spinal-cord tumour. Infective processes and malignant tumours cause a more rapid disintegration of nervous function. Turner (1940) pointed out that usually it was not the clinically obvious case of osteitis deformans that developed paraplegia. Although there may be evidence of a widespread bony involvement on radiography, the radiological features are typical enough to make the diagnosis reasonably certain. The presence of deafness is an additional diagnostic feature in many cases. All the clinical evidence in Case 1 pointed to a non-tumorous cause, in spite of the radiological uncertainty. While Case 2 has not been confirmed operatively, the long history and the affection of so many bones leave little doubt that the paraplegia is due to osteitis deformans of the spine. Among the recorded cases of paraplegia due to osteitis deformans the association of this condition with a benign spinal-cord tumour does not seem to have been made.

The only satisfactory form of treatment is removal of the affected spinous processes and laminae of the vertebrae. Myelography is desirable as a pre-operative measure, to confirm the level of compression and to exclude another lesion. When many vertebrae are implicated it may not be expedient to do an extensive laminectomy, and myelography would show the site of compression. The laminectomy can be very bloody owing to the vascular nature of the diseased bone. These patients come into an age group in which cardiovascular and respiratory diseases are

common. Osteitis deformans is associated with a special form of heart failure due to high output (Edholm, Howarth, and McMichael, 1945). The results of laminectomy depend on the duration of symptoms, and little improvement will occur when the paraplegia has been present for a long time. Out of the 44 cases in the literature, laminectomy was done 25 times, with three (12%) deaths, and 18 cases have shown improvement after surgery. An important complication is the recurrence of the paraplegia after the initial relief. This has happened in at least three cases.

Case 1 had chronic bronchitis and pulmonary heart disease, but she tolerated the operation well. Her recovery from the paraplegia was slow, but at the end of five and a half months she had only slight neurological deficiency. Case 2 was not operated upon when first seen, owing to his congestive cardiac failure and poor general condition. It was surprising to find him alive ten months later. Although his cardiovascular state has improved, he has been bed-ridden for eighteen months. It is extremely unlikely that any improvement in his paraplegia would occur if decompression was done at this date. The general risk is considerable and it has been decided that he should not be submitted to operative intervention. Sarcomatous change is a real contingency that cannot be obviated.

Summary

Two cases of paraplegia due to osteitis deformans of the spine are described.

The literature is brought up to date.

The differences between the two cases and the general pattern of the disorder are discussed.

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The results of the first real test of the Emergency Bed Service's warning system are considered in the report of the service for the year ended March 31, 1953. The year under review includes the great London fog of last December, which resulted in a quite unprecedented rush of applications, and this was followed at the end of January by the influenza epidemic. Throughout this time of crisis, the warning system worked remarkably well, but the report says: "Although an outstanding success, experience has shown, however, that improvement is possible. At present the issue and cancellation of the warnings is solely dependent on the admission rate, and the winter has shown that this tends to result in the warning being put on rather too late and taken off too early. A modification of the scheme is being worked out whereby account is taken not only of the admission rate, but also of the number of applications being received and of the number of admissions made only after referring the case to the medical referees." Applications over the year as a whole increased, and analysis tends to confirm that this was owing to a rise in sickness rather than in the number of doctors using the service. The report also contains numerical details of the applications and admissions during the year.

RECURRENT GALL-STONE ILEUS

BY

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Gall-stone ileus is stated by various authorities to be the cause of 1-2% of all intestinal obstructions (Illingworth and Dick, 1941). Vick (1932) found that in over 6,000 cases of intestinal obstruction 0.68% were due to gall-stones. The high mortality, said to be over 50%, is due to delay in diagnosis and to the advanced age of the patients. Few of the reported cases were diagnosed pre-operatively. Several cases have been reported with multiple stones in the small intestine, but in only a small minority has a second intestinal obstruction occurred. Maurice Lee reported such a case in 1945. The stone usually ulcerates through between the gall-bladder and the duodenum, where it passes down into the small intestine, becoming impacted at the lower ileum. Wakefield, Vickers, and Walters (1939) in 141 post-mortem examinations found the fistulous track in 101 cases.

Case History

A housewife aged 57, 1 para, was seized by violent epigastric pain followed by vomiting two days before admission. These symptoms soon eased but returned on the day of admission, the pain this time being colicky in nature and central in situation. It was associated with severe and persistent vomiting. There was no relevant previous history apart from a dislike of fatty foods.

On examination the patient was dehydrated, with a dry, furred tongue. The abdomen was slightly distended, and on auscultation continuous loud peristaltic sounds were heard. Two enemas were given with negative results. After a diagnosis of acute intestinal obstruction of unknown causation, a laparotomy was performed.

First Operation (J. A. M.).—A distended loop of small bowel presented itself through a right middle paramedian incision. This loop contained a gall-stone which was impacted about 2 ft. (60 cm.) from the ileo-caecal valve. As it was too hard to crush, it was milked up into the distended loop and removed through a longitudinal inci-

Photograph of the two gall-stones.

sion. No other gall-stone could be felt in the small intestine. The gall-bladder area was not examined. Up till the ninth day the post-operative course was uneventful. The patient was taking nourishment and her bowels were working normally. On the tenth day, however, she complained of epigastric pain and vomiting. During the following 10 days she had attacks of incomplete intestinal obstruction, with colic, vomiting, and constipation one day followed by the passage of diarrhoea and flatus the next. It was thought that she had a post-operative obstruction due to local peritonitis and kinking, and attempts to relieve it by intestinal decompression were made. By the twentieth day it was decided that as she was not improving a further laparotomy should be performed.