OSTEITIS DEFORMANS WITH SPINAL CORD COMPRESSION

REPORT OF THREE CASES

F. R. LATIMER, M.D., J. E. WEBSTER, M.D., AND E. S. GURDJIAN, M.D.*

Wayne University Neurosurgical Service, Grace Hospital, Detroit, Michigan

(Received for publication May 5, 1953)

APPEARENTLY, osteitis deformans occurred in antiquity. Paleontologic evidence exists which, it is claimed, reveals manifestation of the disease in fossil apes. Some observers have felt that preserved specimens of prehistoric man show the characteristic alterations. It remained, however, for Paget to describe the disease as an entity.

Justifiable therapeutic pessimism usually attends the diagnosis of osteitis deformans. Specific therapy is not available and palliative treatment is unrewarding. However, a syndrome occasionally accompanies Paget’s disease that offers some hope of symptomatic relief for variable periods of time. This entity is spinal cord compression secondary to vertebral osteitis deformans. Three patients with the disease, manifesting signs of progressive spinal cord involvement, have been under our care and surgical decompression was accomplished in each by laminectomy.

REPORT OF CASES

Case 1. A 66-year-old white male was admitted to the hospital complaining of progressive weakness of both legs with burning pain of 2 years’ duration.

Examination disclosed a “steppage” gait with atrophy of the musculature of the left leg. There was no definite sensory level although the joint-muscle-tendon sense was lost to the iliac crest. Roentgenograms revealed the characteristic changes of osteitis deformans in the dorsal spine and right hip joint (Fig. 1).

Since Pantopaque® myelography delineated a complete block of the subarachnoid space at the 8th dorsal intervertebral disc, a decompressive laminectomy was performed. The laminae

* Aided by the Kresge Fund.
Case 1. Photomicrograph of dorsal lamina showing typical trabecular mosaic bone with fibrosis of marrow. (Hematoxylin and eosin stain, ×90)

were extremely thick and vascular (Fig. 2). The patient remained asymptomatic for 1 year when the syndrome again appeared. Laminectomy was again performed at the same level and the pathology observed was similar. However, postoperatively, there was little improvement and the patient remains confined to a wheelchair 3 years after surgery.

Case 2. A white male, aged 65 years, had fallen from a stepladder 1 year prior to admission. Since that time he had noted numbness of both feet. Gradually, weakness of both legs appeared and on admission he was paraplegic with loss of all sensory modalities below the 7th dorsal dermatome.

After demonstration of an obstruction opposite the 5th dorsal vertebral body by myelography, decompression was undertaken. The laminae were 5 cm. in thickness and bled profusely.

Convalescence was remarkable in that within 6 months he was playing golf regularly. Five years later, however, pain and weakness of the legs reappeared. Radiographs revealed extensive osteitis deformans of the skull (Fig. 3) and the entire vertebral column. Myelographic block was relieved by decompression and the pathology was similar to that previously observed (Fig. 4). The patient is again walking unaided.

Case 3. A 34-year-old white male had sudden development of progressive weakness of both legs 6 months prior to hospital entry.

Examination disclosed a left “foot drop,” weakness of all motions of both lower extremities, hypoactive patellar and Achilles reflexes, and a level of sensory impair-
Fig. 3. Case 2. Characteristic “cotton wool” appearance of skull.

Fig. 4. Case 2. Photomicrograph of dorsal lamina showing heavy trabeculae with fibrotic marrow. (Hematoxylin and eosin stain, X$90$)
ment from the 7th dorsal dermatome downward. Extensive osteitis deformans was revealed by skeletal survey x-rays (Fig. 5). Introduction of contrast medium via the cisterna magna outlined a complete obstruction at the 7th dorsal segment.

Surgical exposure of the area disclosed thick, vascular laminae which were removed. A diagnosis of osteitis deformans was made on the specimens of bone submitted.

Results of surgery were disappointing and the patient remains a custodial problem 3 years later, confined to a wheelchair by weakness of the extremities.

GENERAL CONSIDERATIONS

Osteitis deformans with spinal cord compression is a relatively rare syndrome. While Paget's disease and disintegration in lumbar spine, has been found in 3 per cent of autopsies on persons over 40 years of age there have been only about 50 cases of spinal cord compression secondary to the disease reported in the literature.

Since it has been hypothesized that mechanisms other than direct compression may produce spinal cord involvement in osteitis deformans, i.e. ischemia of the cord caused by either arteriosclerosis of spinal arteries or foraminal narrowing with resultant circulatory embarrassment, it cannot be assumed that all patients with osteitis deformans and signs of spinal cord disease suffer from myelic compression. Therefore, perusal of literature pertaining to the entity led us to establish certain criteria for the diagnosis and to exclude those cases that did not fulfill these requirements. We considered it absolute evidence that there was encroachment on the spinal cord if a myelographic block was demonstrated or if the pathologic process was verified by surgery or autopsy. It was regarded presumptive evidence if there was a subarachnoid block as demonstrated by lumbar puncture with jugular compression or if the protein of the cerebrospinal fluid was elevated. It is probable that in some of the cases not included there was, in fact, encroachment but the proof was not indisputable.

We wish to consider, then, only 22 of the cases included in these papers.

These cases, on the whole, fall into a clinical pattern. Males predominated and the highest incidence was in the 4th through the 6th decades. Anamnesis usually revealed the steadily progressive course of spinal cord involvement, 1 to 2 years in duration. As stated by Turner, the patient's
complaints are those of neurological impairment and not of the bone disease. In the great majority of the cases the initial complaint was impairment of sensation. Usually paresthesias appeared in the lower extremities; numbness, burning or “cramping” were included. Pain was also a frequent complaint. When present, the discomfort began in the vertebral column later to radiate into the extremities. In 11 of the 22 cases “girdle” sensations appeared but were not of great value in predicting the exact level of the lesion. In some cases, however, the syndrome began with progressive weakness in the involved limbs. As the disease developed, the gait became affected, varying from “shuffling” to definite ataxia. In 3 of the patients once the weakness had appeared the paralysis became rapidly complete. Trauma was not a significant precipitating factor.

Examination revealed that the usual stigmata of Paget’s disease were not a necessary accompaniment. Since in all but 2 of the cases the level of involvement was in the upper dorsal area, hyperactive deep reflexes in the lower extremities with varying degrees of weakness in the legs were the rule. A sensory level corresponding roughly to the vertebral involvement was usually in evidence but the line of demarcation was by no means a sharp one.

Survey of the laboratory studies performed disclosed, as would be anticipated, that the protein content of the spinal fluid was elevated and that while the serum calcium, phosphorus and acid phosphatase remained normal, the alkaline phosphatase level was raised.

From a radiographic standpoint, spinal cord compression did not occur in the monostotic form of osteitis deformans in the cases under consideration. The patients who had received adequate roentgenologic investigation presented alterations in two or more adjacent vertebral segments with or without compression fractures, and demonstrable lesions in other portions of the skeleton. The great majority were classed as advanced involvement.

The operative findings were essentially the same in all cases. When exposed the laminae were extremely thick although easily removed with a rongeur. The bone was very vascular and bled profusely. After complete decompression by laminar excision, the dura mater appeared normal and was not opened in most of the cases. It was usual to observe the dura mater begin to pulsate freely after the constricting bony ring was removed. In all operated patients, save 1, the compression was found in the mid-dorsal area. The explanation for this predilection may lie in the fact that since the interpedicular space is narrowest in the thoracic region it follows that the cross-sectional area of the spinal canal is at its smallest. Therefore, any proliferation of the bony vertebral ring would be more apt to manifest itself by spinal cord embarrassment at this level. This hypothesis has been advanced by Schwarz and Reback.

Of the 22 patients, 16 were operated upon. There were 3 deaths in this group of surgical cases; 12 patients were considered benefited by the procedure. The amount of improvement varied from complete, but temporary,
recovery up to periods of 11 years to partial relief of symptoms for shorter periods of time.

DISCUSSION

In the light of the above presentation our cases conform to the general pattern. All were males and 2 were over 60 years of age. Case 3 is the exception. At 34 years of age he probably represents one of the youngest individuals in the literature of the syndrome.

The symptomatology of the 3 patients was the manifestation of progressive spinal cord involvement, and trauma appeared to be a precipitating factor in 1 case. Upon examination, they revealed varying degrees of incapacity. Case 1 was ambulatory with a "steppage" gait while Case 2 was bedridden with complete paraplegia. The occurrence of this type of gait in Case 1 and hypoactive tendon reflexes in the lower extremities of Case 3 appears paradoxical in the presence of a cord lesion in the thoracic region. However, it is possible that they were the product of a second lesion. Perhaps foraminal overgrowth in the lumbar area produced compression of peripheral nerves at this level with resultant hypoactive deep reflexes and weakness of dorsiflexion of the feet. The clinical picture may then have represented a composite of both pathological processes.

Laboratory studies in these cases merit little additional comment. Suffice it to say that they fell within the limits anticipated in osteitis deformans.

Roentgenograms in our patients demonstrated that involvement in all was of the polyostotic variety. Myelography delineated a complete block of the subarachnoid space in each. The level of obstruction was mid-dorsal in 3 cases. In all respects the radiographic findings paralleled closely those observed by other authors.

The appearance of the thickened laminae and the soft, vascular structure of the bone removed in conjunction with its microscopic appearance leaves little room for doubt that this was osteitis deformans. The difficulties in achieving adequate hemostasis in the 3 patients confirms the observation made by Edholm, Howarth and McMichael who measured the blood flow through bone affected by osteitis deformans and found it increased many times that in normal bone.

The results of surgery were gratifying in 2 of the 3 cases. Case 1 was afforded symptomatic relief by the first procedure for 1 year. Case 2, initially the most severely affected, was able to resume nearly normal activity for a period of 6 years and is again improving after a second decompression. Case 3, however, was benefited only a short time by the operation and is now incapacitated.

CONCLUSIONS

From the information gained from the cases reported by other authors and the personal experience afforded by 3 cases it is our belief that a definite program of management should be adhered to in spinal cord compression secondary to osteitis deformans. All patients with a history suggesting pro-
gressive spinal cord involvement in whom Paget's disease is suspected should be investigated by myelography. Inability to perform lumbar puncture should not be a deterrent since the medium may be injected via the cisternal route. If an obstruction is encountered in the subarachnoid space the area should be adequately decompressed by laminectomy. We do not stand with Adson\textsuperscript{1} who believed that "it is doubtful whether decompression of the spinal cord is indicated, since the relief obtained is but temporary"; nor do we believe that the severity of initial neurological deficit is a reliable guide as to the ultimate benefit from the surgery.

**SUMMARY**

Three cases of osteitis deformans with spinal cord compression are presented; 1 patient was aged 34 years. It is concluded that all patients with osteitis deformans manifesting signs of spinal cord involvement should be subjected to myelography and that a block of the subarachnoid space, if present, should be relieved by adequate laminectomy. Severity of neurological deficit should not be regarded as a reliable guide as to the ultimate benefit from surgery.

**REFERENCES**

2. BUTLAN. Cited by Silcock.\textsuperscript{38}