Charcot Joint of the Lumbar Spine

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Historically, the relationship between spinal cord lesions and certain joint diseases is well recognized, having been pointed out by Mitchell in 1831. Later, neurogenic arthropathies were observed in hemiplegia by Scott and Allison in 1843, in syringomyelia by Blasius in 1848, in acute myelitis by Magnier in 1859, and in relation to lesions of peripheral nerves by Packard in 1861 (described by Delano).

Following Charcot's description in 1868 of joint changes in tabetics, neurogenic arthropathies became known as "Charcot joints." Jordan in 1936 first described the condition in association with diabetes mellitus.

The first reported case of spinal neuroarthropathy is attributed to the German physician Kronig in 1884 (described by Holland). Zucker and Marder in 1952 described the first reported case of Charcot spine occurring with diabetes mellitus.

Etiology

Eloesser in experiments with cats in 1917 contributed important information on the etiology of Charcot joints. In one group of animals, he divided a number of posterior roots supplying a limb and found that some of the animals developed Charcot joints in the affected limb. In a second group of cats, he injured joints and then divided posterior roots to the involved extremity. He found that the cutting of the posterior roots had no effect on the existent joint deformity. In a third group of animals, he divided posterior roots and then injured the joint; within 3 weeks every cat thus treated developed a Charcot joint. He concluded "this, then, was not a slow aggravation of a pre-existing deforming arthritis; it was a sudden response of an anesthetic joint to the acute trauma of operation, a rapid reaction to bone injury by the production of a typical Charcot joint."

Foster and Bassett stated that the following conditions may contribute to the formation of Charcot joint: 1) major trauma or repeated small traumas associated with continued movement of a diseased limb; 2) intact motor power to the affected joint; 3) impairment of pain impulses; 4) impairment of proprioceptive impulses which normally inhibit hypermobility of joints; 5) chronicity of the underlying nervous disorder; and 6) metabolic disturbances.

In several large series of Charcot joints, the condition occurred most commonly in tabes dorsalis, with syringomyelia being the second most common cause. Diabetes mellitus and other neurologic disorders are infrequent etiologic conditions.

Pathology

The pathological changes typical of Charcot joints have been described by several authors. The process is believed to be essentially the same in all involved joints. Early in the process, joint cartilage is lost in places and may become covered by a vascular connective tissue. In the deeper layers of the joint, there is an abnormal proliferation of cartilage cells. Destruction of cartilage is accompanied by destruction of underlying bone, which becomes irregular in structure. The spaces between the lamellae are filled with connective tissue. Resumption of enchondral ossification occurs, and is often so great and so diverse that by simultaneous absorption and proliferation an enormous incongruity of the joint surfaces may result. Either the atrophic or hypertrophic changes may predominate.

In the vertebral column, there is initially a narrowing of the intervertebral disc space and a relaxation of ligaments. Degenerative changes lead to erosions and osteoporosis of the vertebrae. Proliferative changes result in sclerosis of the vertebral bodies, as well as bony growth and osteophytes. Tiny
fractures occur, and detached bony fragments are found. Late in the process bony proliferation springs from the articular margins and tends to strengthen the disorganized joint by producing some degree of ankylosis. Compression of the vertebral body develops at times with displacement posteriorly and laterally. Usually, the process is localized in one to three vertebrae, often with a sharp kyphosis and lateral rotation.

The gross changes described above are evident radiographically, although early signs of fragmentation and fine debris formation are usually less evident in radiographs of the spine than in the joints of extremities because of overlying soft tissues and gas shadows.7,21

Clinical Data

Most patients are in the fifth and sixth decades of life. The time interval in tabes dorsalis of the syphilitic infection and the development of the arthropathy varies from 5 to 40 years. Among tabetics, approximately 4% to 10% develop Charcot joints, most often in the lower extremities. The knee is the joint most often involved, with hip and feet (tarsal joints and phalanges) being next in frequency. Spine involvement comprises from 6% to 21% of Charcot joints depending on the series quoted.5,20

Ankle, shoulder, hands, elbows, and other joints are less often involved.

Several authors have described the general features of Charcot joint of the spine.7,10,11,13,14,21,22 The site of predilection is the lumbar spine, but cases have been reported involving the thoracic spine,13,14 the cervical spine7,21 and the sacrum.10 The great majority of cases occur in tabetic patients. Only a minority of patients have associated neurogenic arthropathies of other joints.

Usually the signs and symptoms characteristic of tabes dorsalis are well developed. A kyphosis or scoliosis of the lumbar or thoracolumbar spine is often present. The irregularity of the spine is sometimes palpable. There is no local tenderness and the spinal movements are excessively free, but the affected segment of the spine may remain rigid due to ankylosis.

The differential diagnosis7,10,11,13,21 includes osteoarthritis, suppurative osteomyelitis, spinal tuberculosis, typhoid and Brucella spondylitis, Paget's disease and other bone maladies, and old traumatic conditions.

The symptoms and signs of nerve root compression occur in a minority of cases. Involvement of posterior spinal roots with resultant sensory disturbances is mentioned by several authors, including Campbell and Doyle.3 Pressure upon nerve roots due to collapsing spine or hypertrophic bone formation may lead to considerable discomfort. These root pains are usually gradual and insidious in onset and of moderate severity. Occasionally, however, they attain considerable intensity and may simulate gastric crises of peptic ulcer pain.

Alergant in 1960 claimed to be the first author to report the occurrence of Charcot spine as a cause of motor symptoms and signs due to pressure upon anterior nerve roots. He reported two patients with tabes dorsalis, both of whom developed bilateral foot drop followed by marked weakness in both legs. In both cases radiographs showed marked destructive changes affecting the lumbosacral vertebrae of a type characteristic of tabetic arthropathy. Storey9 reported a further case of paralysis of the legs from lumbosacral root involvement.

Case Reports

We have observed five cases of Charcot joint of the lumbar spine.

Case 1. A 61-year-old white man was hospitalized in 1961 with the diagnosis of atonic neurogenic bladder and urinary tract infection. The patient gave a history of tabes dorsalis diagnosed in 1941 and treated with several courses of penicillin. A full neurologic examination was not done; however, the patient was noted to have a "tabetic gait," a positive Romberg sign, and absent deep tendon reflexes in the legs. The blood serology (VDRL and Kolmer) tests were positive. A lumbar puncture was not done. Charcot joints of L4–5 and L5–S1 were noted on pyelography, and films available from 1956 and 1961 showed progressive bony changes over the 5-year period (Fig. 1 left). The spine lesion was not treated.

Case 2. A 58-year-old white man was hospitalized in 1963 for evaluation of weight
loss. He gave a history of gonococcal infection treated with “fever therapy” in 1925. In 1947, he developed shooting pains in the legs, and in 1958, the diagnosis of tabes dorsalis was made. He was treated with Bicillin. At that time, both blood and spinal fluid serology were positive. He was seen again in 1961 with continuing attacks of shooting pains in the posterior aspect of both legs. At that time, both blood and spinal fluid serology were negative. He gave a history of a “stroke” in 1959 with a residual right hemiparesis.

In 1963, neurological examination revealed the pupils to be small, regular, and equal, with no reaction to light, adduction absent in the right eye and weak in the left eye, and absence of convergence. There was a mild right hemiparesis, absent deep tendon reflexes in the legs, absent deep pain in the testes and legs, and absent position and vibration sensibility in the legs. Radiographs taken in 1958 and 1963 showed progressive bony changes of L2–3 and L3–4 compatible with the diagnosis of Charcot joints of the lumbar spine (Fig. 1 right). The spine lesion was not treated.

Case 3. A 35-year-old white man was hospitalized in 1959. He had a history of childhood scoliosis, and at age 14 a fusion of the lower thoracic and upper lumbar spine had been performed. At age 17, congenital syphilis was diagnosed, and he was treated with unknown medications. Since age 25, he had had progressive difficulty in walking, and he gave a 10-month history of pain in the low back. On neurologic examination, the patient was thought to have “classic tabes dorsalis,” with small equal pupils capable of accommodation but not reactive to light, a “flapping gait,” positive Romberg sign, hy-
potonia, absent deep tendon reflexes and absent position and vibration sense in the legs. Deep pain was absent in the testicles and the legs. The blood Kolmer test was positive. A lumbar puncture was not done. Radiographs revealed neuropathic changes of L2–3 and L3–4 (Fig. 2 left). Bony bridging, presumably due to prior surgical fusion, was noted from T-8 to L-2. The patient was treated conservatively with a back brace.

Case 4. A 72-year-old white man, hospitalized in 1964, gave a history of lues noted 20 years before admission and treated with an arsenic compound and penicillin. A lumbar puncture in the early 1940’s reportedly revealed a positive serology. In 1959, fleeting lightning pains began in the legs and difficulty with gait and balance developed. The patient’s height had reportedly decreased since 1959.

Physical examination in 1964 showed lumbar kyphoscoliosis. Pupils were round, small, with the right slightly larger than the left; there was no reaction to light and only slight accommodation capability. Strength in the legs was slightly decreased. The gait was broad-based, with slapping of the feet. Deep tendon reflexes were absent in all four extremities. Vibration, position sense, and pin-prick pain were absent in the legs, and deep pain was present in the testicles but absent in the legs. Serologic tests for syphilis were positive in the blood but negative in the spinal fluid. Routine spinal fluid examination was normal. Radiographs revealed L-2 through L-5 arthropathic changes, and there were multiple indentations of the dural sac in the lumbar region at myelography (Fig. 2 right). The patient was given a course of 21.6 million units of penicillin.

Case 5. A 52-year-old white man was hos-
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pitalized in 1964. He had contracted syphilis in 1927, and was treated at that time with mercury. In 1947, he developed lightning pains in both legs, and the diagnosis of tabes dorsalis was made. In December, 1960, a left cervical chordotomy was done, and 1 month later, a right thoracic chordotomy. Subsequently, pain sensation was absent below L-1 bilaterally.

In December, 1962, the patient returned complaining of "spasms" and "tingling sensations" in his legs which interfered with his work. Examination showed the left pupil to be 1.0 mm larger than the right, and no pupillary reaction to light or accommodation. Absent knee and ankle reflexes and a flat-footed gait were noted. Pain sensation was absent below the L-1 level bilaterally, and there was decreased graphesthesia, vibration and position sense in the legs, with no weakness present. Spinal fluid Kolmer test was nonreactive. Radiographs revealed L4–5 changes compatible with Charcot joint of the spine; at myelography, there was a block at L3–4.

In January, 1963, bilateral hemilaminectomies of L-3 through S-1 were done with removal of redundant ligamentum flavum. The L4–5 joint was thought to be unstable at surgery, and a posterior bony fusion of L3–4 and 4–5 was done. In July, 1963, the patient developed sphincter symptoms, and his right leg became weak. A further decompressive laminectomy of L4–5 was done along with partial removal of the bone graft. The patient recovered motor and sphincter function except for a persistent right foot drop. He subsequently returned to work as a pharmacist.

In 1964, the patient presented a 4-month history of pain in the medial aspect of the left knee with weight-bearing only, which had become so severe he was unable to work as a pharmacist. At that time, he had a bilateral foot drop with marked weakness and atrophy of the tibialis anticus bilaterally. There was slightly decreased strength in the quadriceps femoris, hamstrings muscle group, and gastrocnemius on each side.

There was no tenderness along the spine and straight leg raising was negative. Sensory and reflex examination was unchanged. Blood serology (VDRL) was nonreactive. Radiographs showed further changes of L3–4 and 4–5 in comparison with previous films, with disintegration of the body of L-4. A myelogram showed an L2–3 block.

In November, 1964, complete laminectomy L3–5 was done with decompression of the left L-3 and L-4 nerve roots. At L-4, a large mass of nucleus pulposus was found anterolateral to the dural sac on the left. Postoperatively, the patient developed a Staphylococcus aureus wound infection and a spinal fluid fistula, which were treated successfully. After the wound healed, the patient walked without further leg pain. He developed further weakening of the right leg despite back and right leg braces, and radiographs (Fig. 3) showed skeletal shortening due to the diseased spine. He was discharged in February, 1965, with back and right leg braces. He has since returned to work as a pharmacist.

Discussion

In the five cases of the present series, all the patients had symptoms and signs characteristic of tabes dorsalis, all had been treated, and all at some time during the course of their illness had a positive serological test for syphilis in the blood or spinal fluid. In the first two cases, there were no symptoms directly referable to the spinal lesion. In the third case, the only case of congenital lues, the patient complained of back and left leg pain, possibly due to involvement of posterior nerve roots. The fourth case showed questionable involvement of motor roots. In the fifth case, there was unequivocal evidence of both sensory and motor root involvement. Also, in this last case, bilateral chordotomies were done 6 years before the development of symptoms due to Charcot spine, and it is interesting to speculate that perhaps the further reduction in pain sensibility occasioned by the chordotomies may have contributed to the development of the neuropathic spinal lesion in this patient.

In regard to therapy, conservative treatment is generally recommended, including bed rest, traction, and braces. Surgical fusion of various involved joints other than the spine occasionally succeeds but usually fails.7,16,19,20 Fusion procedures on the cervical and lumbar spine have been reported, but the efficacy of this form of treatment is
not clear.\textsuperscript{20} In the one case of the present series subjected to surgery (Case 5), progressive bony changes with nerve root involvement occurred subsequent to the fusion attempt.

Few procedures involving surgical decompression of compromised nerve roots have been reported. Relief of retention of urine by decompression of the spinal canal has been reported.\textsuperscript{20} Case 5, with both motor and sensory root involvement subjected to surgical nerve root decompression, symptoms were temporarily relieved by each procedure only to recur when progressive bony involvement led to further neural compromise.

Summary

Five cases of Charcot joint of the lumbar spine secondary to tabes dorsalis have been reported, and the incidence, etiology, pathology, clinical features and therapy of this condition discussed. Conservative treatment is recommended in most cases. The efficacy of surgical spinal fusion is not clear, but in the one patient of the present series (Case 5) who underwent spinal fusion, progressive bony changes occurred following the fusion procedure. In cases in which nerve root compression develops with resultant sensory and motor symptoms and signs, surgical decompression of neural elements may be of benefit.

References

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