Hypertrophic Spinal Pachymeningitis

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Hypertrophic cervical pachymeningitis is a clinical syndrome characterized by compression of the spinal roots and spinal cord caused by a marked inflammatory hypertrophy of the cervical dura mater. The pathological process can spread to the leptomeninges and give rise, because of the progressive compression, to areas of softening of the spinal cord, where it is also possible to find cavities similar to those in syringomyelia.\textsuperscript{26,29,40} The cause of the disease is still unknown.\textsuperscript{15,30}

The clinical picture is still that first described by Charcot in 1869\textsuperscript{5} and Joffroy in 1873,\textsuperscript{18} who divided the evolution of the disease into three stages. The first stage is characterized by intermittent radicular pains in the neck, shoulders, and arms, which gradually become continuous. In the second stage, muscular weakness and atrophy develop in the arms. Finally, spastic paralysis of the legs, loss of various modalities of sensation, paresis of bowel and bladder, and respiratory disturbances characterize a third stage.

The disease has been described in the cervico-thoracic,\textsuperscript{6,8,29,31,27} thoracic,\textsuperscript{1,14,21,25,28,39} and lumbar\textsuperscript{12,11} regions, or along the entire extension of the spinal column,\textsuperscript{6,24,27} with symptoms varying according to the level of the lesion.

We are reporting five cases of hypertrophic spinal pachymeningitis operated upon during 1955 to 1965. We have excluded from this series 14 cases of chronic epidural inflammatory processes and 30 cases of spinal arachnoiditis operated on in the same period, even though some of these showed a moderate thickening of the spinal dura mater.

Case Reports

Case 1. S.S., a 32-year-old male, entered the clinic on January 18, 1955, suffering from continuous pains in the buttocks, and numbness and weakness in both legs, of 3 weeks duration. He had no sphincteric disturbances or fever. His past medical history was noncontributory except for cervical lymphoglandular tuberculosis during infancy. One year before, he had complained of sharp pains in the right buttock radiating to the posterior side of the thigh, and weakness in the right leg, both of which disappeared spontaneously within 1 month.

Examination. The patient was found to have a bilateral spastic gait and weakness of the legs, more marked on the right; Babinski's signs; increased tendon reflexes and transient ankle clonus; and absent abdominal reflexes. The sensory level was D-10.

Laboratory data. Blood count, urinalysis, syphilis tests, and x-rays of the thoracic spine were normal. Lumbar puncture revealed total block and a xanthochromic cerebrospinal fluid containing 18 white blood cells per cc; protein content was 400 mg\%; Pandy 3+, Nonne-Apelt 3+, Weichbrodt negative; the colloidal gold curve was 11220.0. Lumbar myelography demonstrated a posterior filling defect from T-9 to T-7 where the flow of contrast medium was completely blocked (Fig. 1).

Operation. At laminectomy, palpation revealed a hard thickened dura which when opened was about 1 cm thick at the T-7 level. The spinal cord appeared pale and soft. There were some adhesions going from the dura to the arachnoid; these were cut. The dura was then removed in two parallel strips between the midline and the emergence of the posterior spinal roots. The wound was closed.

The pathological material was composed of many layers of connective fibers liberally infiltrated with lymphocytes and plasma cells (Fig. 2). Special stains revealed no organisms.

Postoperative course. By the 15th postoperative day, muscular power had increased in the legs. At the last follow-up in May, 1966, gait was only slightly spastic and tendon reflexes were hyperactive. Plantar responses were neither flexor nor extensor.
The sphincters were intact, and there were no sensory abnormalities.

**Case 2.** In 1948 this 15-year-old boy had low thoracic radicular pains; 3 years later spastic paraparesis and hypesthesia appeared in the legs. At laminectomy from T-9 to T-11 a thickened dura was removed; total recovery followed this procedure. In 1954, weakness in the legs reappeared and was associated with radicular cervical pains, atrophy of the hands, and paresis of both arms. The following year, both legs became totally paralyzed; the sphincters were poor, and changes in sensation marked. Myelography demonstrated obstruction below C-5. Laminectomy from C-4 to T-3 revealed thickened dura mater (Fig. 3) which was removed. Marked improvement followed and continues 11 years later. The arms are normal, and he walks with a slightly spastic gait. He has no pain but some sensory deficit remains.

**Case 3.** When admitted in 1959, this 65-year-old woman had had weakness in both legs and a bilateral band of paresthesia at the umbilical level for 2 months. There was clinical evidence of spinal cord com-

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**Fig. 1.** Case 1. Myelogram showing complete block of contrast medium at T7-T9.

**Fig. 2.** Case 1. Photomicrograph showing infiltration of dural fibers with lymphocytes and plasma cells. E. & E., X200.

**Fig. 3.** Case 2. Photomicrograph showing areas of necrosis within dural connective tissue surrounded by epithelioid elements and lymphocytes. E. & E., X100.