HYPERTROPHIC SPINAL PACHYMENINGITIS
WITH SPECIAL REFERENCE TO APPROPRIATE SURGICAL TREATMENT*

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Charcot and his pupil Joffroy studied and delineated the clinical picture of hypertrophic pachymeningitis of the cervical spinal canal so accurately that little has been added to our knowledge since then. In 1869 they described a case in the Archives de Physiologie and in 1871 Pierret reported another example from the service of Charcot. In 1873 Joffroy published his classical thesis on the subject under Charcot’s direction. They were able to find only 4 cases of this condition that had previously been recognized and reported. The first example was recorded by Abercrombie and the second by Ollivier d’Angers. William Gull in 1858 published the third case, with illustrations of the appearance of the cross-section of the thickened dura mater and the compressed cervical spinal cord. They credited Köhler with a fourth case but review of Köhler’s original description reveals that it was the pia and arachnoid that were grown together forming a thick, leather-like covering to the spinal cord and that the dura mater was not involved in this process.

Charcot and Joffroy pointed out that the symptomatology is divisible into three periods. (1) The painful period. The onset is with pain in the neck and back of the head, which soon begins to radiate into the upper extremities. These pains are caused by involvement of the cervical meninges and, in turn, of the posterior cervical spinal roots. At first the pain is remittent but after 2 to 5 months it becomes continuous. It is often associated with stiffness of the neck and is aggravated by movement of the neck. Finally, tingling, like needles and pins, develops in the hands and fingers, and sensitivity in the upper extremities is gradually impaired. (2) Atrophic paralysis. The painful period and that of atrophic paralysis in the upper extremities may be combined or clearly separated. In any event the pain occurs first and after a variable interval atrophy and weakness appear in the upper extremities. The distribution of the atrophy is variable. It may appear in one or both upper extremities. The small muscles of the hands are usually involved, but the muscles of the arms are by no means spared. (3) Spastic paralysis. As the disease progresses a spastic paralysis of the lower extremities, with the usual changes in reflexes, appears. Respiratory movements may be interfered with. The bladder and bowels are usually affected and sensory

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changes of variable nature and extent develop. Eventually a severe paraplegia supervenes and the patient becomes bedridden with paroxysmal flexor-defense reflexes.

Charcot and Joffroy recognized that the dura mater posterior to the spinal cord was most severely involved and that the disease usually occurred in the cervical region. They were also aware that it might occur in other parts of the spinal canal and might even involve the intracranial dura mater, particularly that in the posterior fossa. In the latter location the disease might give rise to paralysis of various cranial nerves. They described the thickened dura mater as composed of concentric layers of fibrous connective tissue, similar to the cornea, the vessels increased in number and their walls thickened. They believed that the disease initially is confined to the dura mater but that gradually the spinal roots become inflamed and compressed, and that the involvement tends to extend gradually to the leptomeninges and even to the spinal cord. They noted that the leptomeninges often become thickened and that the dura mater may become adherent to them and to the spinal cord. In addition to evidence of inflammation the spinal cord may develop areas of softening and cavitation secondary to the prolonged and increasing compression.

They clearly distinguished between hypertrophic pachymeningitis and various other diseases of the dura mater. They were well aware of the difference between this condition and hemorrhagic pachymeningitis interna (subdural hematoma), which they frequently found intracranially but seldom intraspinally; between it and pachymeningitis externa which was seen particularly in association with Pott's tuberculosis of the vertebrae; and between it and purulent affections of the internal surface of the dura mater. They also stated clearly that they had been unable to find the cause for hypertrophic pachymeningitis, and that no specific or satisfactory form of treatment had been found.

Since the 1870's little progress has been made and much confusion has been added. The distinct entity that Charcot and Joffroy so clearly described has been largely lost sight of in a welter of confusion with epidural and subdural disorders, particularly epidural granulomas and intradural inflammatory processes secondary to the acute meningitides. Probably the most serious misunderstanding has come from the now nearly universally accepted belief that the disease is almost invariably of luetic origin. This is simply not true. The disease has also occasionally been erroneously attributed to tuberculosis. It would not be possible to state that these infections had never given rise to hypertrophic spinal pachymeningitis but when one reviews the cases so diagnosed he finds that they either occurred before the introduction of the Wassermann reaction (1906) and the discovery of the Treponema pallidum (1905) or rested solely upon the microscopic evidence of a chronic inflammatory process in the dura mater without any positive evidence as to the etiology. In fact in many cases the disease has been attributed to syphilis in spite of negative serological tests on the blood and