OCCULT LUMBOSACRAL MENINGOCELE.

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THE mildest form of the congenital anomalies classed under the generic term of spina bifida is the occult variety, in connexion with which nothing may be visible externally beyond a small median fold or dimple; a tuft of hair is sometimes present, sometimes a small lipoma. The palpating finger may discover evidence of ununited laminæ and spinous processes. Through the gap no section of cord or membranes protrudes, but from the posterior aspect of the theca a fibrous band—the membrana reuniens—extends between the muscles to the under-surface of the skin at the dimple. Occasionally the back is entirely normal in appearance.

Should spinal meninges project through the laminal opening at all they are generally considered to reach the skin invariably, bulging it outwards to form a rounded sac constituting a meningocele. This of course is immediately visible, of varying size, sessile or pedunculated, translucent, and in part perhaps replaceable. Its contents comprise cerebrospinal fluid within a lining of arachnoid dura, but no neural tissue except possibly the incidental herniation of a spinal root.

The remarkable numerical preponderance of lower spine cases amid the total of all levels is apparent by a glance at the appended Table, derived from figures compiled by Stockmeyer¹.

TABLE.
LOCAL INCIDENCE OF SPINA BIFIDA.

Region.	Number of cases.
Occipito-cervical	21
Thoracic	7
Lumbar	83
Lumbosacral	42
Sacral	60
	213

No fewer than 185 out of 213 cases involved the lumbar or sacral region, the percentage being just under 87.

In general, the occult lumbosacral variety gives rise to but few symptoms or signs; they are often unobtrusive, even latent; loss of knee- or ankle-jerks will escape notice unless looked for, and the solitary indication may possibly consist of a tendency to urinary incontinence. The condition is sometimes discovered by accident, as in the case of a patient of 40 who first complained of backache after a fall of a few feet.

The case here reported presents a series of unusual features, which may be summarized as follows: (1) complete absence of any exterior deformity or anomaly; (2) complete absence of symptoms till the age of 16; (3) excessive radicular and local pain when at length the condition manifested itself; (4) a pathological basis of what may fairly be termed occult meningocele, inasmuch as the cyst protruded beyond the laminal opening and burrowed under the erector spinæ muscles on each side.

PERSONAL CASE.

A. B., female, age 18, was admitted to hospital towards the close of 1930, with a complaint of pains and loss of power in the lower limbs.

The girl had always been healthy and active; and during both infancy and childhood no abnormality of lower limbs or sphincters was noticed. Some two years prior to her coming under observation a moderate attack of what was diagnosed as sciatica (on the right side) drew attention for the first time to that part of her body. After treatment prosecuted for some weeks the pain subsided. About a year later the pain returned, and thereafter did not disappear though its intensity fluctuated. Gradually during the last months before admission it became more and more severe, and at the same time the legs became weak, especially the right, and were considered by her mother to be getting thinner. Trouble with bladder and rectal control also made its appearance.

On admission, the girl was ascertained to be incapable of walking; her legs were drawn up at knee and hip, and were undoubtedly both wasted and flabby. Complete abolition of the deep reflexes in the lower limbs was noted. The plantar responses were practically nil. Incontinence or retention was present in regard to the bladder and rectum.

By far the most striking symptom, however, was the pain. The patient had become hysterical with suffering, and objective examination was conducted under wellnigh incredible difficulties. She screamed as soon as the bedclothes were removed for purposes of investigation; handling or palpating her spine and back was a frank impossibility. The pain was declared to be excruciating; it started over the sacrum or a little higher and radiated down both legs, especially the right. It was practically continuous, with paroxysmal exacerbations on movement, and in particular on coughing or sneezing. Objective sensory testing became farcical; yet so far as could be ascertained there was either diminution or loss to pin-prick over the lower sacral roots (S 2, 3, 4, 5) on both sides.

A decision was accordingly reached that further studies should be conducted under an anæsthetic. In this way the X-ray photographs were taken, and they disclosed the existence of a large and almost circular defect of bone-formation on the back of the sacrum, involving the lowest lumbar and the first sacral vertebræ (fig. 1). Its median position and symmetrical character were very striking and informative, pointing as they did to a spina bifida occulta lumbosacralis.

In view of this development in the case, the precise nature of which had not till then been apparent (the skin over the back was smooth and without blemish, though minute palpation of the spine had been rendered impossible owing to the patient's reactions), an operation was proposed and accepted.

FIG. 1 Skiagram of lumbosacral region, showing the circular defect in the last lumbar and first sacral vertebræ. There is complete absence of the laminae and spinous processes.

Operation.—Under intratracheal ether and with the patient lying prone the lower lumbar and upper sacral region was exposed. The muscular masses of the erectores spinæ were retracted on either side of the spine. There was no sign of either laminae or spinous processes of the last lumbar vertebra, and the posterior neural arch of the first sacral vertebra (segment) was absent. Through the bony defect the dura mater bulged so tensely as to appear as though it might burst; it extended beyond the osseous margins and burrowed on each side

under the erector spinæ (fig. 2). It thus constituted in point of fact an occult meningocele.

On incision into the bulging cyst a large quantity of spinal fluid escaped. The edges of the dura mater were retracted, and the roots of the cauda equina could be seen flattened against the lateral walls of the meningocele (fig. 3). There was no obstruction above or below the meningocele, which was carefully examined. Except for this flattening of roots no abnormality otherwise was

FIG. 2. Drawing, showing the tense and bulging meningoceleic dura mater after the erector spinæ muscles had been retracted.

FIG. 3. Drawing, illustrating the marked flattening of the roots of the cauda equina. They were pressed up against the lateral walls of the bulging dural sac.

discovered. The redundant dura mater was cut away, and the edges of the membrane approximated except at the lower part: here the meninx was stitched to the erector spinæ muscle on each side in order to form a drain for any excess of spinal fluid. The latter muscles were stitched together in the midline and the skin wound closed with interrupted silkworm sutures. A small drain was inserted under the skin and removed after 24 hours.

The patient was nursed prone for the first week after the operation, and then alternatively on her side. The wound healed soundly, and the stitches were removed on the ninth day.

The relief from pain was immense; bladder control became more nearly normal, and further examination was made easy. This substantiated the hypalgesia or analgesia of the lower sacral root segments.

Treatment to the wasted legs became practicable, and the improvement was so rapid that the patient left hospital able to walk, some two months after her admission.

COMMENTARY.

Had the girl not been so hysterical over her pain it would no doubt have been possible to detect the ununited laminæ in the lumbosacral region, which seemed to be somewhat fuller and more rounded than usual. Yet the complete absence of any local malformation or congenital anomaly was at once manifest to the eye.

Occult lumbosacral meningocele of this kind is of interest more especially from the standpoint of its late evolution when all symptoms are in abeyance during the earlier years of life. To what factors this not infrequently observed succession of events is due has been much debated. The theory of Katzenstein² and others is that vertebral elongation during normal growth combines with anchorage of the hernia at the site of the lesion to cause progressive stretching and degeneration of spinal or radicular tissues, but it is not applicable to all cases—perhaps only to a minority. In this particular case the problem is to account for the gradual development in the late 'teens of a meningocele hidden under muscles, through a congenital aperture in the lumbosacral spine. No history of trauma was forthcoming; nor was there the slightest indication of any state of internal hydrocephalus such as might conceivably have hastened the bulging at the other end of the fluid system. The pathogenesis remains obscure, and no light is thrown thereon by reference to other cases of spina bifida occulta in which onset of symptoms occurred late. In one recorded by Hassin³ they commenced at 36, in Marie and Léri's⁴ at 46, in Bassoe's⁵ at 52.

The occurrence of severe radicular neuralgia in the same condition is also distinctly rare; it is mentioned, however, by Margulis⁶, Roederer and Lagrot⁷, Beck⁸, and others.

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