Tethering of the conus medullaris within the sacrum

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Included among various forms of craniovertebral congenital defects, known generally as spinal dysraphism or myelodysplasia, is a little described condition produced by the conus medullaris being tethered within the confines of the sacrum, usually by a lipomatous growth. Diagnosis may be difficult and is often not considered by clinicians in spite of characteristic clinical and radiographic appearances. Previous writers on anomalies of the central nervous system have seldom considered this entity separately although treatment differs from that in all other lumbosacral myelodysplasias. Three successive cases are now presented, the relevant embryological and clinical literature is reviewed, and emphasis is placed on positive contrast diagnosis and proper definitive therapy.

CASE REPORTS

CASE 1 This 6-year-old white boy was born on 18 August 1958 after a full-term uneventful pregnancy. At birth a lipomatous lumbar swelling was noted but disregarded. He developed normally, walked, and rode a tricycle. In August 1963 he began to develop spasticity, particularly of the thigh adductors, and to fall frequently. Over the next six months he gradually became incontinent. Significant in the family history was a paternal grandfather who was reported to have had rachischisis. The patient was admitted to another hospital in August 1964. Examination revealed a lipomatous mass in the lumbosacral area, spasticity, and wasting of the legs, no sensory loss, and urinary incontinence. On 11 August 1964 the superficial lipoma was removed and a laminectomy of L3, L4, and L5 was performed after which a fatty tumour rose into the wound. The dura was not identified and the procedure was terminated. Postoperatively the neurological loss progressed.

Examination in January 1965 revealed an alert, intelligent, normocephalic boy. The upper part of the body was normal but he had bilateral hip flexion contractures and pes valgus. The only voluntary motion remaining in the lower extremities was adduction of the left thigh. Patellar reflexes were hyperactive bilaterally, but ankle reflexes were absent. Abdominal and cremasteric reflexes were absent. Bilateral extensor plantar responses were shown. Perception of pin and touch were diminished up to the knee bilaterally. The perineum was anaesthetic and vibration sense was lost at the ankles. Micturition was automatic.

Spinal films demonstrated the surgical laminectomy and bifid lumbar and sacral vertebrae (Fig. 1). An electromyogram showed decreased motor unit activity in the legs. No fibrillation potentials were seen. An intravenous pyelogram was normal. Cisternal positive contrast myelography showed a distended, low-lying thecal sac extending deeply into the sacrum and curving to the

FIG. 1. Case 1: plain spine film showing spina bifida and previous partial laminectomy. Superimposed line drawings demonstrate the position of the lipoma, the attachment of the conus, and the level of the thecal sac.
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The spinal cord could not be identified nor any Arnold-Chiari malformation.

On 2 February 1965 an extensive lumbosacral laminectomy was performed, and most of the laminae and the roof of the sacrum were found to be defective. An incomplete dura was opened and nerve roots were seen to be travelling upwards. The empty enlarged thecal sac curved to the right and the conus medullaris extended into a large sacral lipoma, which was both intra- and extradural, and lying on the left (Fig. 1). This mass was dissected and divided as far caudad as possible allowing the cord to rise and tension to be decreased. No attempt was made to close the dura primarily. The postoperative course was uneventful with no immediate improvement. On examination nine months later there was no improvement over the patient's pre-operative condition.

**CASE 2**

This 12-year-old girl was born with a sacral mass, said to have been lipomatous, which was removed at the age of 18 months. As development progressed it was apparent that she had no perineal sensation although she crawled and later walked normally. She had stationary urinary and faecal incontinence up to one year before admission when some rectal sphincter control was gained spontaneously. In March 1964 after she had fallen down several stairs low back pain developed. In July 1964 her left leg became weak but improved within 10 days. Twenty-four hours before admission she again fell and developed weakness of the left leg. A maternal aunt had a low back tumour excised which contained hair and teeth but her symptoms were not so severe. Otherwise there was no significant family history.

On examination on 21 October 1964 the patient was unable to move her left leg. There was no atrophy and muscle tone was normal. She had bilateral pes cavus and a right thoracic scoliosis. The perineum and perianal areas were anaesthetic. Abdominal reflexes were intact and extensor plantar responses were present bilaterally. Tendon reflexes were normally active in the legs.

Spinal films showed bifid laminae of L5 and sacral segments. A cystometrogram was consistent with a spastic reflex bladder. Myelography by the lumbar route demonstrated an enlarged, low-lying thecal sac with upward directed nerve root sleeves (Figs. 2, 3).

On examination on 2 December 1964 the findings were unchanged, and laminectomy was performed on 7...
December. The sacrum was entirely open. The dura was opened, found to be defective at its caudal end, and merging into a lipomatous growth. The spinal cord and conus medullaris extended into the large sacral lipoma and all sacral roots found their exit cephalad. The lipoma was dissected from its attachment which was at the skin of the lower end of the scar remaining from the previous excision in infancy. Tension on the spinal cord was markedly reduced. Post-operatively movement of the left leg improved but sensation was unchanged. On 2 March 1965 she was able to walk normally, but still had urinary incontinence and sacral anaesthesia. Faecal incontinence had subjectively improved even further. The extensor planter response was absent on the left and present on the right. Tendon reflexes were hypoactive in the knees but normally active at the ankles. In May 1965 the patient was readmitted with weakness of both legs and bilateral extensor planter responses. Over a three-week period of bed rest the weakness partially disappeared. In the ensuing six months the patient remained ambulatory on crutches. A suprapubic cystostomy has been required.

CASE 3 This 14-year-old white boy had a two-year history of unexpected episodes of spontaneous micturition, which had been increasing in frequency. For many years the patient had urinary urgency and was noted to stumble often. At the age of 6 months a draining sinus consistent with a pilonidal cyst of the midline coccygeal area was first noted by physicians. The patient had a similar draining dermal sinus in the midline at L5 since birth. Lumbosacral radiographs revealed spina bifida occulta. At 17 months the patient had *E. coli* meningitis which was treated successfully. On 2 March 1952, at 18 months, a lumbar laminectomy was performed, the lumbar sinus tract resected, followed into its communication with the dura, and the dura closed. The pilonidal cyst was not removed. Subsequent development was normal including all sensory and motor function.

On examination a lumbar scar was visible and the pilonidal cyst had healed, its only trace being a coccygeal skin dimple. Rectal tone was poor and many scars were visible on the legs. Positive neurological findings were restricted to the lower extremities where tone was increased. Muscle strength was good, however, and the patient had a normal gait. Bilateral unsustained ankle clonus was present and deep tendon reflexes in the legs were hyperactive. Extensor toe signs were absent. Abdominal and cremasteric reflexes were retained, and all sensory modalities were intact, including those in the perineal area.

A cystometrogram was consistent with a spastic bladder. An intravenous pyelogram was within normal limits. An electromyogram showed fibrillation potentials scattered throughout the lower extremities.

On lumbosacral radiographs the extensive spina bifida occulta was visible. On 4 September 1965 a cisternal myelogram revealed a large thecal sac extending deeply into the sacro-coccygeal area. Although nerve roots could not be seen the negative shadow of the spinal cord was visualized in the sacral area so that the diagnosis of a tethered conus was suspected. A laminectomy of lumbar, sacral, and coccygeal areas was performed. The conus medullaris was attached, within a lipoma, to the coccygeal skin dimple by means of fibrous tissue strands. Nerve roots found an exit upwards and dense adhesions were present. Although the lipoma was freed and the cord was seen to travel cephalad after tension was relieved, a radical operation could not be performed safely for fear of damage to vital nerve roots. Postoperatively the patient did well and experienced no neurological loss as a result of surgery. After a seven-month follow up, the same symptoms and signs were present as pre-operatively, but they had not progressed.

DISCUSSION

The tethered conus medullaris is part of the larger group of midline embryological defects collectively known as myelodysplasia or spinal dysraphism (Lichtenstein, 1940). The many variants comprising the larger groups have been discussed by Fisher, Uihlein, and Keith (1952), and Cohn and Hamby (1953). These defects are thought to occur in early gestation by a process of abnormal or arrested maturation and involve mesodermal structures, skin, or neural elements in combination or alone. As a result of such embryonal dysplasia various clinical conditions, including defects of the vertebral arch, dural and neural tube fusion anomalies, cutaneous cysts and sinuses, ectopic ganglia, and other defects may result (Lichtenstein, 1940). The embryological aspects have been extensively reviewed by Saunders (1943). Defects under general discussion here are restricted to those which have prevented normal cephalad migration of the conus medullaris, namely, sacral tumours (dermoids and lipomata), fibrous traction bands, and tight filum terminale (Jones and Love, 1956) because of clinical examination they may be indistinguishable. These can occur as occult neural defects but are usually associated with a lumbosacral spina bifida (Lichtenstein, 1942).

Streeter (1919) has shown that in normal foetal development before the 11-15 mm. stage, the cord extends to the lower end of the sacrum, and it is at this stage that the conus medullaris becomes fixed within the sacrum. Normally the dorsal nerve roots become attached to the cord before the conus medullaris ascends and thereafter are useful for marking various stages of ascent. At this time the nerve roots are horizontal but after the 30 mm. stage, the vertebral column lengthens caudad more rapidly than the spinal cord and the roots then take a downward course (Streeter, 1919). During this time the filum terminale is formed by atrophy of distal segments of the cord leaving a residual ependymal strand. The formation of the filum terminale may also account for further ascent of the conus. This
showed that in cases in which the conus is trapped the distance between nerve segments in the lumbosacral area is greater than normal which suggests a stretching process rather than active growth. Gardner (1964) has explained the spinal dysraphic syndromes by postulating that inadequate permeability of the rhombic roof causes hydromyelia, distension of the central neural canal, and finally fusion of germ layers either cephalad or caudal as in the cases under discussion. It has been postulated that lumbar diastomyelia and myelomeningocele occur in this manner. If this theory can be applied to sacral tethering one must postulate that germ layers became fused in the sacral rather than in the lumbar area. Traction due to primary lumbosacral fusion has been invoked as a cause of the low-lying conus (Lichtenstein, 1940). Evidence for this is seen at the operating table when tethering is relieved and the cord rises immediately. Not one of our patients had a detectable Arnold-Chiari malformation which conceivably might result from such traction from below.

Clinical symptoms produced by the stretched cord from whatever cause have already been described (Brickner, 1918; Ingraham and Lowrey, 1943; Garceau, 1953; Jones and Love, 1956; James and Lassman, 1960; Love, Daly, and Harris, 1961). Such varied symptoms and signs as enuresis, foot deformities, trophic pedal ulcers, scoliosis, and progressive neurological deficits in the lower extremities occur. Typically, the symptoms appear or become worse during two periods of life. First, when the child assumes the erect posture and second, during the period of rapid growth of early adolescence (Jones and Love, 1956). The common presentation is generally an unexplained progressive spasticity during these two periods of life superimposed on a background of urinary difficulties. Differentiation from certain other myelodysplasias, such as the Arnold-Chiari malformation (Peach, 1965a and b), may be difficult. In each of our cases a lumbosacral scar from previous surgery was present. The presence of a dermal defect is not a prerequisite of the condition but in almost all cord traction syndromes a spina bifida is present.

To explain the prominent signs of an upper motor neurone lesion Katzenstein (1901) formulated the theory that caudal traction on nervous tissue may result in proximal degeneration. However, Barry, Patten, and Stewart (1957), by measuring distances between nerve segments, showed that those distances immediately above the point of tethering are greater than normal, and stated that this effect of traction from below is dissipated within five nerve segments. Distant effects of stretching may, however, cause more proximal microscopic or physio-
logical changes. The stretching may affect the large diameter nerve fibres of the corticospinal tracts by the same mechanism by which Kahn (1947) postulated that the spinal cord is fixed by the dentate ligaments in cervical spondylosis. These larger fibres are more vulnerable to metabolic and physical disturbances than smaller fibres (Kahn, 1947). In some cases there may be concomitant distant neuro-anatomical defects in the posterior fossa or along the thoracic cord which might explain the spasticity which is often seen (Lichtenstein, 1942; McKenzie and DeWar, 1949; Shorey, 1955). In our cases myelographic and clinical evidence of other neuro-anatomical defects was absent.

Diagnosis of a conus medullaris tethered within the sacrum may be established by positive contrast myelography. In any child presenting with progressive neurological deficiencies in the lower extremities against a background of urinary difficulties the diagnosis of a conus traction syndrome must be suspected. Even in the presence of a normal vertebral column a low-lying conus cannot be excluded. A lower than normal dilated thecal sac is suggestive. Swedberg (1963) has advocated the use of negative contrast gas myelography in cases of myelomeningocele. He was able to identify the level of the conus and to show abnormal fixation of the cord to the vertebral column. He was, however, unable to demonstrate the direction of nerve roots. Nerve roots which are seen, on contrast study, to find an exit upwards are pathognomonic of fixing the spinal cord within the sacrum. Gryspeerdt (1963) presented the myelographic findings in James and Lassman’s (1962) cases of occult spinal dysraphism. Fifty-one cases had positive contrast myelographic studies. The value of identifying the level of the conus medullaris in the supine position or indirectly determining the level by seeing a low-lying anterior spinal artery in the prone position was stressed. We have been unable to define the level of the conus by the anterior spinal artery at this level. Taveras and Wood (1964), on the other hand, regard the prone position as superior in defining the level of the conus medullaris. Gryspeerdt (1963) was able to demonstrate wide fila, fibrous bands, and ossified and unossified septa. The direction of nerve roots was not visualized. Bleasel (1961) has delineated the cauda equina and conus medullaris by utilizing an emulsion of Myodil and spinal fluid. This technique renders the material being examined more radiolucent and so provides a better outline of the spinal cord and nerve roots. If the shadow of the spinal cord can be seen in the lumbosacral area a tethered conus medullaris is suggested.

Walker (1944) pointed out that often in cases of myelodysplasia the bony vertebral canal is seen to be widened on plain radiographs. He presented one case, similar to ours, with a sacral lipoma and upward directed nerve roots which was treated by unroofing the sacrum. In the case of Alexander, Garvey, and Boyce (1954) the nerve roots were seen to find an exit upwards at the time of surgery but in this instance the conus medullaris was bound down by scar tissue, not lipoma, at L5-S1. Continence of urine was restored after the third sacral roots were crushed bilaterally. Bassett (1950) described lipomas of the cauda equina in nine patients, and some of them were in an abnormally low position. Campbell (1962) discusses good post-operative results in several cases. In our cases beneficial effects of surgical treatment are not proved. There are other available reports in which the conus medullaris was found to be tethered, but generally by tight fila, scar tissue, or neoplasm (Meredith, 1944; Jackson, Thompson, Hooks, and Hoffman, 1956; Jones and Love, 1956).

Gryspeerdt (1963) noted that of 32 consecutive cases of occult spinal dysraphism there were no cases of the Arnold-Chiari malformation. He therefore regarded cisternal puncture as safe. Russell and Donald (1935) agreed that in spina bifida occulta the hind brain is usually normal. However, in 10 consecutive cases of meningomyelocele they showed the presence of the type II Arnold-Chiari malformation and cautioned against cisternal puncture. Daniel and Strich (1958) discussed 26 cases of the Arnold-Chiari malformation all of which had a meningoceleolec. One fatality occurred after cisternal puncture in a patient with meningomyelocele. Campbell (1962) has discontinued pre-operative diagnostic studies because he feels insufficient information is gained to justify the risk of trauma to neural elements, but valuable information was gleaned from the pre-operative positive contrast studies in our cases and we recommend such studies. From this evidence it would appear that a contrast study is desirable but should be performed cautiously in any child suspected of having a cord traction syndrome (List, 1941; Cameron, 1957). If an Arnold-Chiari malformation is present, cisternal puncture for instillation of contrast material carries risk. On the other hand if symptoms are due to a low-lying conus medullaris, lumbar puncture is theoretically hazardous.

The surgical approach varies with the severity of the lesion. In cases of tight fila terminale the conus seldom extends into the sacrum and the nerve roots travel downward. Here a simple lumbar laminectomy and division of the tight filum suffices (Ingraham and Lowry, 1943). In cases of conus medullaris caught below the sacrum the deformity is
more severe and the junction between conus and lipoma is unclear. The conus extends deeply into the sacrum and nerve roots take a cephalad direction.

Generally the conus is bound down by a fibrofatty tumour. In these cases the laminectomy should begin in a relatively normal lumbar area and proceed caudal. If the laminectomy begins in the sacrococcygeal area there is a greater risk of neural damage. The dura must be opened. In a series of 474 cases of spina bifida Ingraham (1943) showed that 45 cases were associated with lipomata. Hauge (1963) described such a case in which the sacral roof was removed and the conus freed from a fatty tumour. Four days after operation the patient controlled micturition for the first time in his 16 years of life. On the other hand Garceau (1953) described a patient who had three laminectomies before the sacrum was unroofed and a short thick filum was divided. In our cases there was initially a failure to recognize the nature of the lesion. Had a more aggressive approach been taken earlier, much neurological function might have been preserved. Beneficial effects of adequate surgery have not been proved in our series although progression of the neurological defects has probably been halted. This is in contradistinction to some of Campbell’s (1962) cases and other cases previously cited. The problem of management of the accompanying urinary tract infection depends on the success or failure of neurosurgical therapy and has been extensively discussed (Mertz and Smith, 1930; Gross, Holcomb, and Swan, 1953; Morales, Deaver, and Hotchkiss, 1956).

These cases are examples of tethering occurring during the first month of gestation usually by a lipomatous growth and therefore differ from cases of tight filum terminale or other traction bands. This distinction can be made pre-operatively by myelographic or at the time of laminectomy by noting the direction of nerve roots and the level of the conus after the dura is opened. Nothing less than unroofing of the sacrum and dissection from below to release the tethered conus from its point of attachment can be considered adequate. In other forms of tethered conus, such as a tight filum terminale or certain fibrous traction bands, an ordinary lumbar laminectomy may be considered adequate.

SUMMARY

A congenital anomaly, tethering of the conus medullaris by a lipomatous tumour within the sacrum, can be diagnosed pre-operatively by myelographic identification of the length of the spinal cord, the caudal extent of the thecal sac, and the direction which the nerve roots take in the caudally displaced thecal sac. Upwards directed nerve roots are diagnostic and indicate that the dissection should be carried into the sacrum below the lipoma for release of traction. In this report the beneficial effect of adequate surgery in three cases is questionable.

REFERENCES