Letters

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The role of childbirth on the natural history of occult spinal dysraphism

Sir: Occult spinal dysraphism is the term used for a variety of embryological malformations of the spinal neural structures and neighboring tissues which, in contrast with spinal bifida aperta, are less or not obvious externally. The condition may go unrecognized for many years, especially when symptoms start in adult life. We report the case of a woman with a lumbar sacral lipoma associated with a widened dural sac and posterior spina bifida, in whom urinary symptoms developed shortly after the birth of her first child and exacerbated after a second childbirth. This unusual presentation led to an initial incorrect diagnosis of post-partum stress incontinence and resulted in a needless urological operation.

A 36-year-old woman was referred to the neurology clinic because of unexplained urinary sphincter disturbances. Symptoms started at age 30, when after the delivery of her first child, she intermittently noted loss of urine during sexual intercourse. Three years later, she gave birth to a second son, and soon afterwards, she complained of urinary frequency and loss of urine during physical strain. Symptoms were attributed to stress incontinence due to the successive deliveries. Pelvic floor exercises were unsuccessful and her gynaecologist decided to perform an anterior colporrhaphy and urethroplasty. After the intervention, she had urinary retention and overflow incontinence. Since then she has required intermittent self catheterisation. Urodynamic examination revealed a hypotonic bladder with vesicouphincteral dysfunction.

She was then referred to the neurology department. For the first time, she admitted to have noticed minimal faecal incontinence during recent months. Although she only complained of sphincter disturbances, examination showed saddle hypaesthesia (S5 to S3 on the right, and S5 to S2 on the left), absent ankle jerks, a hypotonic anal sphincter, bilateral pes cavus and a small operative scar at the lumbosacral region, from the removal of a small “skin nodule” soon after her birth.

Radiographs of the lumbosacral spine showed an enlarged canal at the lower lumbar spine and sacral levels, with posterior sacral spina bifida. Myelography with metrizamide showed a marked widening of the sacral dural sac which contained a multilobulated mass. Combined computed tomographic scanning revealed that the mass had a density of fat (fig.). There was no evidence of a tethered cord since the conus terminated at the lower edge of L1. Surgical exploration confirmed the radiological findings. The lipoma had no extension outside the dural sac. Because the lipoma was merged with the nerve roots, only a small proportion could be dissected. The thin filum terminale was sectioned, although it was not under traction. Histological examination showed normal adult adipose tissue. Postoperatively, the sphincter disturbances remained unchanged.

Stress incontinence is a common manifestation of a neurogenic bladder. Stress incontinence occurring after childbirth is usually due to weakness of supporting tissues of the urethra and base of the bladder. Our patient suffered stress incontinence from an autonomous neurogenic bladder, but the relation of this symptom to the successive deliveries initially suggested the latter cause to be responsible.

A few other anecdotal cases of occult spinal dysraphism, in which symptoms started after childbirth, have been reported. Pool\textsuperscript{4} described a case which shows some similarities with ours. His patient had an intrasacral meningocoele with a lipoma that compressed the adjacent nerve roots and tethered the spinal cord. Symptoms started at age 22 years, when after her first childbirth, she developed pain and paresthesia in the legs. Eight years later, she developed urinary frequency followed by stress incontinence. She also underwent a needless urological intervention. Joseph and McKenzie\textsuperscript{5} reported the case of a 27-year-old woman with an occult intrasacral meningocoele who developed symptoms shortly after the delivery of her fourth child. Although the spinal cord was tethered by a small terminal lipoma, surgical intervention was restricted to bony decompression. Post-operatively there was rapid regression of symptoms and abnormal signs. They postulated that the intensive straining during childbirth could have caused the arachnoid to herniate through a possible dural defect. In their series of adults with a tethered cord syndrome, Pang and Wilberger\textsuperscript{6} included three female patients in whom symptoms started shortly after childbirth. Injury of the conus, due to traction during the straining associated with childbirth, is the postulated mechanism in these patients.

In our patient there was no evidence of a tethered conus, but the sacral roots were compressed by the lipoma. Normally, when a traction is applied on the lumbosacral roots, they can to some extent move out of their foramina. During straight leg raising it has been demonstrated that the L5 and S1 roots can move out of their foramina for some millimeters.\textsuperscript{7} In our case, the lower sacral roots were merged with the lipoma, so that outwards movement through their foramina was impossible. During the straining of childbirth, momentary stretching of the tethered sacral roots could have caused permanent injury to them. Another factor to be considered is that the condition of pregnancy itself could have played a role in the growth of the lipoma. It is well

\textsuperscript{4} Pool RH. J Neurol Neurosurg Psychiatry: first published as 10.1136/jnnp.48.7.721 on 1 July 1985. Downloaded from http://jnnp.bmj.com/ by guest. Protected.
known that steroids, especially glucocorticoids, can modify body fat composition. Excessive epidural fat growth causing spinal stenosis has been reported during both high and low dose glucocorticoid treatment.\(^2\) \(^3\) Progesterone also causes enlargement of body fat depots,\(^4\)\(^5\) and levels of this hormone rise steadily during pregnancy. It might be therefore, that the elevated progesterone levels during pregnancy stimulated the growth of the lipoma, leading to further compression of the sacral roots.

Whatever the mechanism, in some forms of occult spinal dysraphism, especially a tethered cord syndrome or lumbosacral lipoma, symptoms can start after childbirth. When the clinical picture is restricted to urinary dysfunction, especially stress incontinence, diagnosis may be delayed and the patient submitted to needless urological interventions.

FIG. Computed tomographic scanning after metrizamide myelography at the upper sacral level. Posterior spina bifida of the sacral bone and a filling defect with fat density.

References


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Paraspinal myoclonus due to spinal root lesion

Sir: Myoclonus is a symptom produced by a wide variety of neurological diseases.\(^1\) Most commonly, myoclonus is generated from cerebral cortex, thalamus, brain stem, or spinal cord. By contrast, localised myoclonus arising elsewhere is less familiar.\(^2\) As an example of unusual localised myoclonus I report a case in which segmental mid-thoracic paraspinal myoclonus occurred as the only sign of an advanced spinal nerve root lesion.

A 38-year-old man was admitted with unilateral involuntary jerking in the dorsal back muscles. In the early 1960s he had had paralytic ileus, operation for which revealed a neurosarcorna. Later, mesenteric and pelvic metastases were found. The patient's general condition had remained good until 4 months prior to admission when he experienced slight superficial pain below the left scapular region for a few weeks. The pain disappeared spontaneously. Thereafter, he was asymptomatic until he experienced jerking in the muscles of the back on the left side. The vigorous but painful jerks were present occasionally, varying from one day to another, sometimes being absent for weeks. On admission, cranial nerve and cerebellar functions were unaffected. Muscle tone and strength and deep tendon reflexes and the abdominal reflexes were normal and symmetric. The plantar responses were flexor. No sensory disturbances could be detected, and in particular, no thoracic segmental abnormality was seen. The only abnormal sign observed was myoclonic jerking in the paraspinal muscles at the level of the 5th thoracic segment on the left side. Extensive laboratory investigations showed normal values. No abnormality was found in the plasma radiographs of skull, chest and spine. Computed tomography (CT) of the brain and EEG were normal. Electromyography (EMG) showed a normal pattern in the muscles of the extremities and in the paravertebral muscles in the thoracic area. Unfortunately, the jerking was not present and could not be elicited at the time of the EMG investigation. Normal values were also obtained in the measurement of nerve conduction velocities and in the somatosensory evoked potentials which were examined using median, ulnar and posterior tibial nerve stimulation. Metrizamide myelography revealed a mass at the level of the 4th thoracic vertebra. CT examination showed destruction of the left pedicle of the