1. Split spinal cord malformations (congenital)

Ersahin et al. ([i]) reviewed 74 cases. They noted spinal arachnoiditis "caused by the contrast material used in myelography" in two patients, who had paraparesis.

More recently, Iskandar et al. ([ii]) looked at 20 myelomeningocele patients who had a split cord malformation (who comprised about 6% of all their myelomeningocele patients).

The authors noted that all 15 patients who had a delayed diagnosis (mean age 4.4 years) "as expected" had arachnoiditis secondary to prior myelomeningocele repair.

There was also evidence of tethering at the level of the split cord malformation.

Myelomeningocele (spina bifida) arises as a result of embryonic failure of neural tube closure, during the fourth week of gestation.

This causes a protrusion of the meninges through a midline bony defect of the spine, forming a sac containing CSF. Usually MMC is associated with other malformations such as Chiari II, of the hindbrain.

Before neurosurgical techniques came into use, there was extremely high mortality due to hydrocephalus or meningitis etc.

Surgical repair of the defect aims to prevent CSF leak, infection and subsequent spinal cord tethering, as well as preserving neurological function

Wagner et al. ([iii]) described the consequences of primary myelomeningocele closure.

They noted:

"As primary MMC repair is inevitably followed by the development of arachnoiditis, fibrosis and adhesions between intraspinal structures, MMC closure predisposes to secondary tethering of the cord."

They suggested that about a third of patients with MMC develop a symptomatic spinal cord tethering, which tends to present clinically as a progressive scoliosis, gait changes, spasticity or pain and sometimes with weakness, lower limb contractures and bladder dysfunction.

Wagner and his colleagues noted that upper limb symptoms might also arise due to traction on the cervical cord.

Whilst the majority of patients may gain some remission of symptoms by undergoing surgery, later recurrence is likely.

Wagner et al. also remarked that

"The subarachnoid spaces around the spinal cord regularly develop marked arachnoid adhesions after primary MMC closure."

This may lead to arachnoid cyst formation in both children and adults.

2. Idiopathic spinal cord herniation: Berbel et al. ([iv]) described a case of a man who underwent laminectomy for spinal canal stenosis, which did not successfully remove his symptoms.

His presentation mimicked that of Brown-Sequard syndrome. MRI demonstrated ventral displacement of the thoracic spinal cord associated with an arachnoid cyst. At operation, the cyst was removed but attempts to free the spinal cord were unsuccessful due to severe spinal arachnoiditis.

The authors suggested that whilst herniation of the cord is rare, that patients tend to present in a similar manner, with a gait disorder resembling that in Brown-Sequard syndrome.

3. Arachnoid telangiectasis: Buxton et al, ([v]) described a case of syringomyelia secondary to arachnoiditis arising as a result of arachnoid telangiectasia. The patient had no other signs of hereditary telangiectasia.

4. Spinal dermal sinuses. In September 2003, Ackerman and Menezes ([vi]) published a paper on 28 cases of spinal congenital dermal sinuses, in which they reported 22 tethered cords, 14 inclusion tumors, and 6 patients with evidence of arachnoiditis.

The authors remarked:

"Although most patients were referred for cutaneous stigmata evaluation, >50% had neurologic deficit, intradural tumors, or tethered cords."

Hide Bound Cord

This phenomenon was described by Moquin and others in the United States, who have also coined the term thoracic cord syndrome.

Hide Bound Cord is a transient severe stretching of the spinal cord over a defect in the thoracic spine when the patient bends (even at rest if severe).

It is associated with ischaemia or vasogenic congestion and microscopic injury. There may also be a syrinx.

The defect is typically at the thoracic kyphus (T6-8) or within 2 levels above or below.

Radiographically there may be an exaggerated kyphosis and/or small disc herniations /spondylotic bars at the apex of the kyphosis. The spinal cord is in an anterior, fixed position resulting in draping across the defect.

Moquin et al. postulated ([vii]) that there is a relative tethering of the cord leading to a "Hide bound" appearance on MRI, with thinned/deformed cord, loss of signal anterior to the cord as the cord is closely applied to the kyphosis.

There may be syrinx above or below this area and abnormal cord movement on cord motility studies.

Patients tend to present with local thoracic pain, radiating band-like chest pain (uni- or bilateral), neurogenic claudication, constipation, bladder dysfunction, abnormal sensory perception (not necessarily within dermatomal limits), clumsiness/tripping, abnormal balance (especially falling in the dark etc.).

Maliszewski et al. ([viii]) described tethered cord syndrome in adults. They presented 3 cases that developed adhesions of the spinal cord at a late stage after previous spinal interventions (trauma/surgery); symptoms appeared to have been precipitated by sudden stretching of the spinal cord due to a fall or strenuous exercise.

In all 3, the posterior surface of the cord adhered to the dura at the site of injury. MR findings

were confirmed at operation. The authors remarked:

"The tethering of the cord by the scar was the cause of a non-physiological stretching of the spinal cord on flexion of the body and head. It led to spinal circulation disorders and symptoms of myelopathy."

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