

We describe a seminal case report of a child with a tethered cord syndrome secondary to the unusual constellation of a split cord malformation, lumbar myelomeningocele, and coexisting neurenteric cyst. A 17-year-old adolescent girl with a several-month history of myelopathy and urinary incontinence was examined whose spinal MRI scan demonstrated a type II split cord malformation with a large bone spur and an intradural neurenteric cyst in addition to lumbar myelomeningocele. Untethering of the spinal cord was achieved via a lumbar laminectomy. Pathological examination confirmed the intradural cyst to be a neurenteric cyst. Postoperatively there was stabilization of the neurological symptoms. Prophylactic surgery with total resection of the neurenteric cyst when feasible and spinal cord un-tethering appears to be associated with excellent outcomes.