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Lipomyelomeningocele Lumbosacral: A Case Report

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Abstract: ***Lipomyelomeningocele (LMMC) is a type of congenital occult spinal dysraphism consisting of the presence of lipomatous tissue attached to the dorsal spinal cord, which protrudes though a spinal defect along with the menings or spinal cord to form a posterior mass under the skin. LMMC is the commonest cause of congenital tethering (tethered cord syndrome) and causes neurological deterioration due to the conus medullaris and root ischemia. Here we presented a 6 year old girl with a lump over the back region since birth. The lump was progressively increasing in size. There was history of urinary and fecal incontinence since birth. Local examination of the lumbosacral region revealed a single lump with no redness over it, spherical in shape, 12×10 cm in size, soft consistency, and fixed. CT Scan of lumbosacral spine was done which revealed soft tissue mass at lumbosacral that connected with defect canal sacral from L5 until S2. We diagnosed patient with lipomyelomeningocele. Surgery was performed and there are no post operative complications and now after 1 month of surgery the symptoms were improved. Lipomyelomeningocele is rare case in our Department. Surgical treatment is indicated because the patient had urinary disturbance. There are no post operative complications and now after 1 month of surgery the symptoms were improved.***

Keywords: Lipomyelomeningocele, tethered cord syndrome, occult spinal dsyraphism

1. Introduction

Lipomyelomeningocele (LMMC) is a type of congenital occult spinal dysraphism consisting of the presence of lipomatous tissue attached to the dorsal spinal cord, which protrudes though a spinal defect along with the menings or spinal cord to form a posterior mass under the skin. Neural ectoderm separates from the cutaneous ectoderm and periaxial mesoderm comes in intact with the unfused ventral neural ectoderm. The mesoderm then differentiates into fatty tissue, thus preventing the neural canal and the posterior aspect of the spine from fusing.[1]

LMMC are lipomas that are tightly attached to the dorsal surface of a neural placode and extend dorsally through a spina bifida to be continuous with the subcutaneous fat. The lipoma lies immediately external to the connective tissue in the extradural space. LMMC rate has been estimated to be 2.5 per 10000 births. Patients are typically female and present before the age of six months but occasionally the condition goes undetected into childhood. LMMC usually occur in the lumbosacral region of the cord and tether the cord at the level. In general, lipomas located subcutaneously have benign behavior and cause no problem except cosmetical matter but according to their localization, sometimes may cause pressure symptoms.[2,3,4] Here we presented a case with a lump over the back region since birth. The lump was progressively increasing in size.

2. Case Report

A 6 year old girl weighing 15 kg presented with a lump over the lower back region since birth. The lump was progressively increasing in size. There were history of urinary and fecal incontinence and well recognized at 3 year old of age. There were no complain of back or leg pain. The child developments were normal for the age and have no deformities. A girl was born by spontaneous vaginal delivery at 38 weeks of gestation to a 22-year-old woman, gravida 1. Vaccination history was satisfactory.

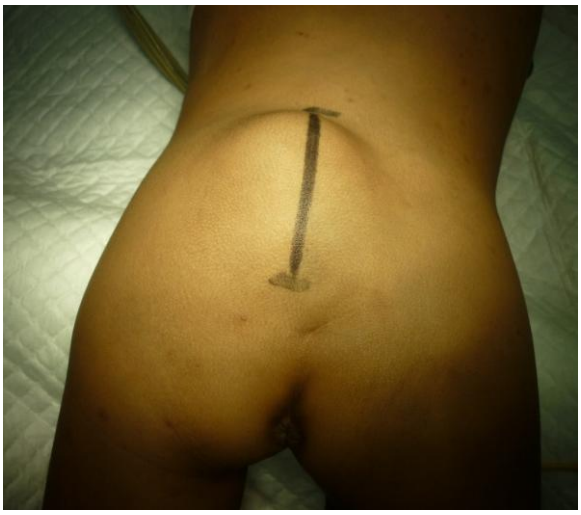

Figure 1: Preoperative lipomyelomeningocele

On physical examination, pulse, blood pressure, repiratory rate were normal. Local examination of the lumbosacral region revealed a single lump with no redness over it on left paramedian, spherical in shape, 12×10 cm in size, soft consistency,and fixed. From motoric and sensory examination were normal. Rectal examination revealed decreased of tonus anal spincther. We also done cough stress test and urine loss was positive when patient cough carefully. From the laboratory findings the blood still within normal limit and the urinary laboratory indicates no urinary tract infections. The patient already brought CT Scan of lumbosacral spine which revealed soft tissue mass at lumbosacral that connected with defect canal sacral from L5 until S2.

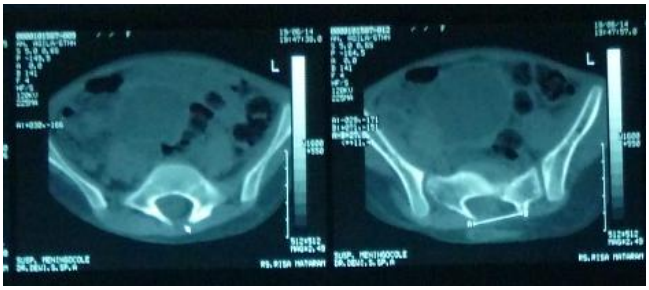

Figure 2: CT Scan of lumbosacral spine show the defect

The patient was operated under general anesthesia in prone position. We done vertical incision straight on the cele and met fatty tissue at L4-S2. We identify the dura and found the spinal cord out of dura. After the fat is cleaned, cele opened so that the spinal cord can be inserted intradura and sutured to close dural defects.

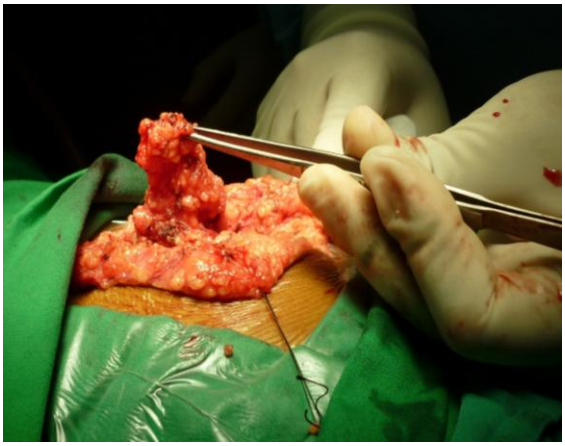

Figure 3: Perioperative photography of lipoma

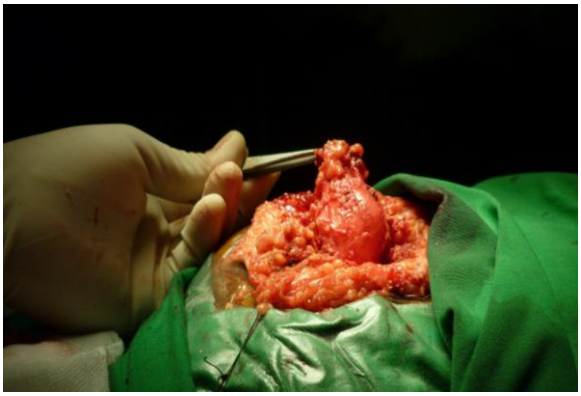

Figure 4: Perioperative photography of cele

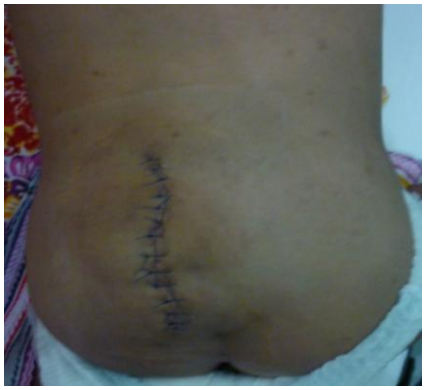

Figure 5: Postopertive photography

3. Discussion

LMMC is the commonest cause of congenital tethering (tethered cord syndrome (TCS)) and causes neurological deterioration due to the conus medullaris and root ischemia.[1,5] The pathogenesis of the clinical syndrome in TCS is believed to arise from traction on the lower end of the spinal cord by a thickenend terminal filum. Traction on the conus medullaris then leads to decreased blood flow and decreased oxidative metabolism, which may eventually cause the clinical symptoms and signs of the TCS. Its clinical features include neurological, musculoskeletal, and urological abnormalities that may be improved after untethering of the filum terminale. There are several corollaries to ischemic hypothesis in TCS. The first is that the mechanical properties of the cauda equine are important, so that a thicker terminal filum or one with fat infiltration of the filum causes higher tension. Second such mechanical properties prevent normal spinal cord migration, which exacerbates this traction.[5]

The most common presenting symptom are fatty mass positioned in the midline or just off the midline in the lumbosacral region. Additionally, the majority present with other skin lesions associated with the lipoma, including a hairy nevus, skin dimples, and cutaneous hemangiomas. The other symptoms mainly related to traction on the lower spinal cord, include, motor deficits, spasticity, sensory disturbance, urodynamic and urorectal dysfunctions, and secondary orthopedic deformities. Symptoms are progressive if untreated.[4,5,6]

A series of 80 patients reported by Kanev et al. demonstrated that bowel and bladder function deteriorates prior to motor function or sensation. Patients in their series demonstrated complete paralysis of bowel and bladder prior to the appearance of motor or sensory loss on physical examination. The disease progression can result in frequent urinary tract infections and neurogenic bladder and bowel incontinence or constipation, as well as leg length discrepancy, foot deformities, gait abnormalities, scoliosis, spasticity, and back and leg pain.[6]

Urinary complaints in these children are secondary to the impaired innervation of the urinary system, either from malformation during embryogenesis, or a tethered cord as a result of the LMMC. Urinary dysfunction can be due to detrusor paresis, external sphincter dysfunction, or most commonly, detrusor sphincter dyssynergy. Urinary dysfunction may cause symptoms such as urinary incontinence, frequent urinary tract infections, urinary urgency, and in severe cases of urological dysfunction, hydronephrosis or pyelonephritis may cause upper urinary tract damage. The initial symptom of a neurogenic bladder is frequently a change in micturition pattern. Urodynamic testing aimed at evaluation of urological dysfunction may help with evaluation of the severity of dysfunction. Additionally, abnormality of bladder function may be the only evidence of neurological compromise in these children.[6]

Three categories of LMMC exist, based on the relative anatomy of the lipoma and neural components: dorsal, transitional, and caudal. The dorsal-type lipomas have an area of attachment to the dorsal spinal cord at the area of myeloschisis in the lower lumbar or lumbosacral levels of the spinal cord and are continuous with the subcutaneous tissue. The lipoma passes through a fascial defect, and may extend into and expand the central canal. A dural defect is present and the placodelipoma interface may lie in the extradural space. Transitional lipomas have an attachment that extends beyond the area of myeloschisis down to the conus, with a less distinct lipoma-cord interface. The lipoma again extends through a dural defect. The caudal-type lipomas arise predominantly from the caudal end of the conus medullaris. These lipomas may extend through a dural defect or may be encased in the dura.[5,6]

Because the natural progression of LMMC is causing neurologic deficits, and/or urological disturbances, the prophylactic surgery is recommended for these lesions by some authors.[2] The aims of surgery are to remove the fibroadipose mass, to relieve the tethering effect on the spinal cord, to preserve neural tissues, and to prevent retethering of the spinal cord. Basic surgical procedures are the same for each type of lipoma, but the degree of difficulty of the surgery is different for each type.[3]

4. Conclussion

LMMC is rare case in our Department. Surgical treatment is indicated because the patient had urinary disturbance. Early surgery can prevent cord infarction and severe urological dysfunction complications From the operation we found that the lipomyelomeningocele was dorsal type. There are no post operative complications and now after 1 month of surgery the symptoms were improved.

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