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Cisternostomy In Lipomyelomeningocele Without Hydrocephalus : A Case Report

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Abstract

Introduction: Spinal dysraphisms can be classified as either open or closed dysraphisms. Closed spinal dysraphisms such as lipomyelomeningocele, diastematomyelia, and spina bifida occulta have no exposed neural tissue and are accompanied by cutaneous markers in 43%–95% of cases, and include lesions such as subcutaneous masses, capillary hemangioma, dimples, and hairy nevus. These cutaneous markers can be used to recognize cases in an asymptomatic neonate. LMMC can be associated with additional pathologies, including Chiari malformation type 1 (13%), spina bifida (14.4%), split cord malformations (3.1%), associated dermal sinuses (3.1%), dermoid or epidermoid cysts (3.1%), diastematomyelia (3.1%), terminal hydromyelia (3.1%), anal stenosis (1.0%), and Down syndrome (1.0%).

Case Report: A year old girl, came to our department with chief complaint of lump on the back since the patient was born. No history of increased intracranial pressure was complained. On physical examination, there is no maceration on the lump, and there is no sign of ruptured lump on the back. Patient also has active motoric on all extremity. We diagnosed the patient with spina bifida. We did CT scan of the head to rule out hydrocephalus and MRI for the confirmation of spina bifida. Excision of cele was done. Postoperatively, there is an increased of csf leak of the patient from the excised cele and we decided to do cisternostomy. After cisternotomy, there is no increased of leakage and patient also have good motoric function. Patient was treated in our outpatient clinic and has good recovery after surgery.

Discussion: Lipomyelomeningocele is a rare but complicated defect, lying in the spectrum of occult neural tube defects. It is actually a form of occult spinal dysraphism in which a subcutaneous fibrofatty mass traverses the lumbodorsal fascia, causes a spinal laminar defect, displaces the dura, and infiltrates and tethers the spinal cord. Spinal lipomas and LMMCs are frequently associated with cutaneous and musculoskeletal abnormalities in addition to sensorimotor deficits and urological dysfunction. Cutaneous lesions include subcutaneous lipomas, capillary hemangiomas, complex dimples, and hypertrichosis, whereas complex malformations, such as dermal appendages, are rare. Magnetic resonance imaging is useful in demonstrating the presence of a fatty mass and cord tethering. Surgical objectives in a lipomyelomeningocele repair include removal of the adipose mass, identification of the defect in the lumbosacral fascia for release of the tether, possible release of the filum terminale, preservation of neural elements, and prevention of retethering of the spinal cord. After operation for lipomyelomeningocele, the cord may not be completely untethered, or after a short period may retether

Conclusion: Cisternostomy may be done in cases of spina bifida without hydrocephalus in our experience

Keyword: Spina Bifida, Cisternostomy, Lipomyelomeningocele

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Introduction

Spinal dysraphisms can be classified as either open or closed dysraphisms. Closed spinal dysraphisms such as lipomyelomeningocele, diastematomyelia, and spina bifida occulta have no exposed neural tissue and are accompanied by cutaneous markers in 43%–95% of cases, and include lesions such as subcutaneous masses, capillary hemangioma, dimples, and hairy nevus. These cutaneous markers can be used to recognize cases in an asymptomatic neonate.[1]

The prevalence of lipomyelomeningocele and lipomeningocele has been found to range between 0.3 and 0.6 per 10,000 live births.[2] 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.[3]

LMMC can be associated with additional pathologies, including Chiari malformation type 1 (13%), spina bifida (14.4%), split cord malformations (3.1%), associated dermal sinuses (3.1%), dermoid or epidermoid cysts (3.1%), diastematomyelia (3.1%), terminal hydromyelia (3.1%), anal stenosis (1.0%), and Down syndrome (1.0%).[1]

As the subcutaneous lipoma is restricted by the defect in the lumbodorsal fascia, the upward movement of the conus medullaris during axial growth may be limited and thus may lead to progressive neurological and urinary deficits, the sequelae of a tethered cord. 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.[1,4]

The subarachnoid cisterns are spaces within the subarachnoid space where the pia mater and arachnoid membrane are not in close approximation. These spaces are filled with the cerebrospinal fluid. Opening of the basal cisterna for evacuation of the cerebrospinal fluid is a well-recognized and effective method for brain relaxation in microneurosurgical approaches to the skull base especially in microvascular surgery.[5]

Case Report

A year old girl, came to our department with chief complaint of lump on the back since the patient was born. No history of increased intracranial pressure was complained. On physical examination, there is no maceration on the lump, and there is no sign of ruptured lump on the back. Patient also has active motoric on all extremity. We diagnosed the patient with spina bifida. We did CT scan of the head to rule out hydrocephalus and MRI for the confirmation of spina bifida. Excision of cele was done

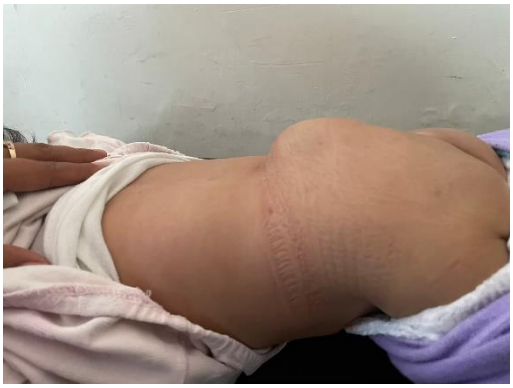

Figure 1. Cele of the patient

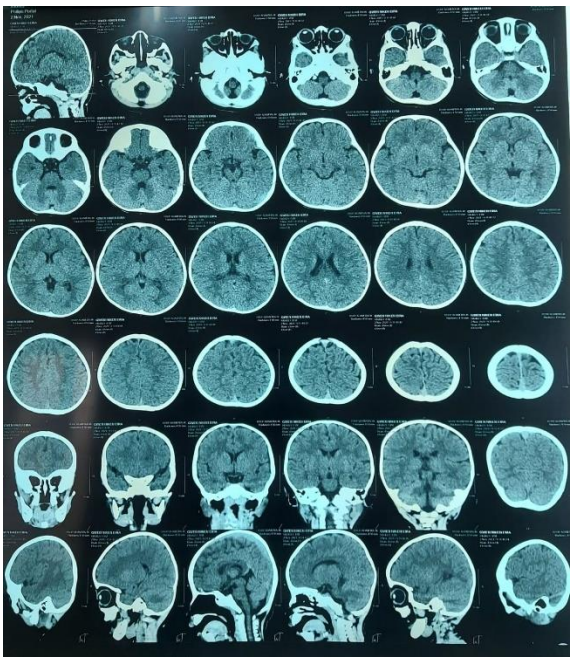

Figure 2. Head CT scan of the patient

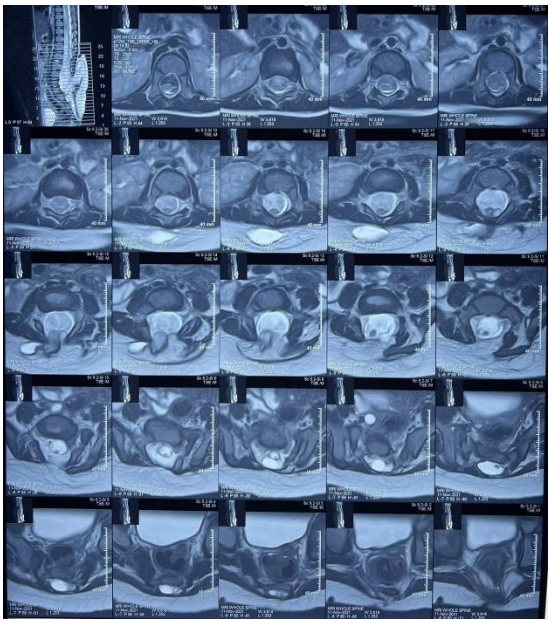

Figure 3. MRI of the Spinal

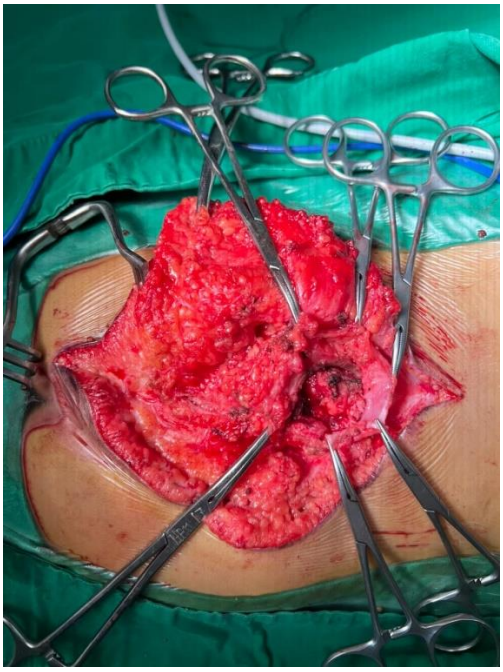

Figure 4. Excision of Cele

Postoperatively, there is an increased of csf leak of the patient from the excised cele and we decided to do cisternostomy. After cisternotomy, there is no increased of leakage and patient also have good motoric function. Patient was treated in our outpatient clinic and has good recovery after surgery

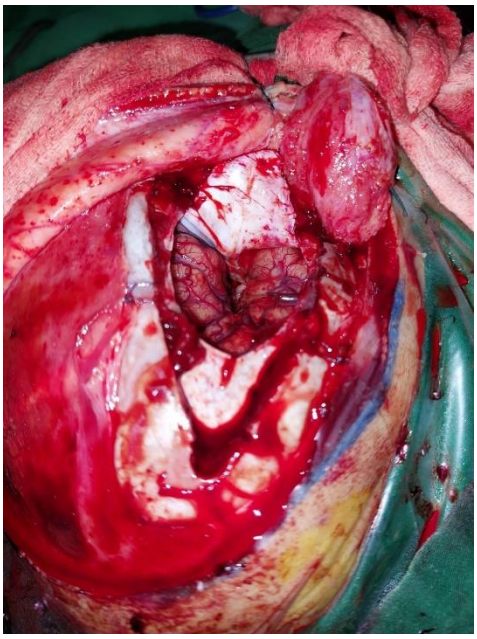

Figure 5. Cisternotomy of the Patient

Discussion

Closed SDs with subcutaneous mass include lipomas with dural defect, lipomyelocele, lipomyelomeningocele, terminal myelocystocele and meningocele. Closed SDs without subcutaneous mass include intradural lipoma, filum lipoma, tight filum terminale, persistent

terminal ventricle and dermal sinus tract.[6,7] Lipomyelomeningocele is a rare but complicated defect, lying in the spectrum of occult neural tube defects. It is actually a form of occult spinal dysraphism in which a subcutaneous fibrofatty mass traverses the lumbodorsal fascia, causes a spinal laminar defect, displaces the dura, and infiltrates and tethers the spinal cord.[8]

Three categories of lipomyelomeningocele 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.[1]

Spinal lipomas and LMMCs are frequently associated with cutaneous and musculoskeletal abnormalities in addition to sensorimotor deficits and urological dysfunction. Cutaneous lesions include subcutaneous lipomas, capillary hemangiomas, complex dimples, and hypertrichosis, whereas complex malformations, such as dermal appendages, are rare. Musculoskeletal findings include scoliosis, unilateral or bilateral foot deformities, such as pes cavus, club feet, or abnormal rotation, or asymmetry of the foot or leg. Any of these findings should prompt consideration of an underlying embryomorphic etiology. Urological dysfunction, such as incontinence, frequency, urgency, and urinary tract infections, are also commonly associated. Neurological symptoms frequently correspond to those expected of a tethered cord syndrome, such as back or leg pain at rest that worsens with activity, in addition to weakness, sensory disturbances, or gait abnormalities.[2]

Prenatal diagnosis of lipomyelomeningocele can be very challenging. A detailed examination of the fetal spine requires diligent scanning in various planes, with results that are very dependent on the position of the fetus.It may be difficult to detect lipomyelomeningocele by ultrasonography if the spine lies adjacent to the uterus, resulting in limited visualization of the subcutaneous mass. Magnetic resonance imaging is useful in demonstrating the presence of a fatty mass and cord tethering.[1]

Postnatally, MRI has aided in both the diagnosis and treatment of lipomyelomeningoceles. Lipomyelomeningocele features can vary substantially depending on the relative size of the lipoma and meningocele, along with the orientation of the neural placode. Characteristically, imaging of lipomyelomeningocele reveals expansion of the spinal canal and subarachnoid space. The cord and the dura extend dorsally through the spinal dysraphism. Most cases present with a deformed and stretched neural placode that is rotated toward the lipoma on 1 side. The meninges herniate on the opposite side.[1]

Surgical objectives in a lipomyelomeningocele repair include removal of the adipose mass, identification of the defect in the lumbosacral fascia for release of the tether, possible release of the filum terminale, preservation of neural elements, and prevention of retethering of the spinal cord. After operation for lipomyelomeningocele, the cord may not be completely untethered, or after a short period may retether. The overall complication rate of surgery is between 10% and 30%. In a series of 120 patients, worse neurological function was found in 5.8% of patients after primary surgery. The incidence of spinal cord retethering following lipomyelomeningocele resection has been found to be between 10% and 20%. Retethering may present first with back pain and deterioration of lower-extremity function followed by worsening urological and bowel function, generally occurring 3–8 years after the initial surgery.[1]

Cisternostomy has been recently proposed in the setting of severe TBI as an adjuvant surgical technique that may have a potential for effectively improving ICP control and outcomes. The procedure consists of the opening of the cisternal spaces and draining this compartment for a period of approximately 1 week. The rationale of the procedure lies in the recognition of the important contribution of the paravascular Virchow-Robin spaces to CSF circulation.[6]

The indications for decompressive hemicraniectomy have been used for this new procedure for cisternostomy which this procedure could be a more elegant and better procedure than decompressive hemicraniectomy. A dural opening is first done, parallel to the supraorbital ridge. About 5 cm of the dura is opened in this manner and the subdural hematoma if present is drained. After this, a large handheld brain spatula and suction are used to get into the interoptic cistern. Once the interoptic cisterns are opened, the microscope is brought in and the opticocarotid cistern is opened in a sharp fashion. The lateral carotid cistern between the carotid and the third nerve is also opened. Through either one of these cisterns, the membrane of Liliequist is opened and the basilar artery, bilateral posterior cerebral arteries (PCAs), superior cerebellar arteries, and the third nerve are visualized. Constant irrigation is performed and the subarachnoid blood is washed out.[4] By using cisternostomy technique, we can decrease morbidities associated with removal of the bone flap in decompressive craniectomy and also the need for doing cranioplasty after that.[5]

Conclusion

Cisternostomy may be done in cases of spina bifida without hydrocephalus in our experience

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