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TETHERED CORD SYNDROME: INSTITUTIONAL EXPERIENCE OF 40 CASES.

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ABSTRACT Tethered cord syndrome (TCS) encompasses a spectrum of congenital conditions that result in progressive neurological deterioration due to tension on the spinal cord and nerve root[1]. Patients with TCS present with a wide spectrum of symptoms hence detailed physical examination is vital in patients presenting with TCS. Although TCS is a well-known entity, it continues to pose challenges regarding diagnosis and management. Surgical untethering may either ameliorate these issues or halt their worsening if they have been long-standing for many years.

KEYWORDS : tethered cord syndrome

INTRODUCTION

Tethered cord syndrome (TCS) encompasses a spectrum of congenital conditions that result in progressive neurological deterioration due to tension on the spinal cord and nerve root[1]. Tethered cord is a neurologic disorder caused by tissue attachments that limit the movement of the spinal cord within the spinal column, this can be secondary to a heterogeneous group of disorders, such as spinal lipomas, lipomatous-filum, split cord malformations, and meningomyelocele. It typically occurs in children, and it is rare in adults [2].

The term filum terminale syndrome was first used by Garceau in 1953 in describing three patients.[3] Two decades later, in 1976, Hoffman coined the term "tethered spinal cord" in patients with a low-lying Conus medullaris with a thickened filum[4]. More recently, there have been descriptions of TCS in which patients are described to have the conus medullaris in a normal position on imaging but presenting with signs and symptoms consistent with TCS [4]

Adult-onset cases are rare compared to that in children.[5] The risks include folic acid deficiency [6], mothers very young or very old in age, toxic medications, obesity, multiple gestations, anti-epileptic medications, zinc deficiency, and ingestion of excessive tea in the first trimester of pregnancy [7].

The abnormally low position of the conus-medullaris may lead to neurological, musculoskeletal, urological, orthopaedic, or gastrointestinal abnormalities[7]. Patients with TCS present with a wide spectrum of symptoms these are mainly dependent on the age of presentation and the underlying cause hence detailed physical examination is vital in patients presenting with TCS. Although TCS is a well-known entity, it continues to pose challenges regarding diagnosis and management [8]

The congenital tethered cervical spinal cord is a very rare entity and is usually due to a dermal sinus tract stalk entering the subarachnoid space and attaching to neural elements [9]. Eller and colleagues reported a taut fibro-neural band which tethered the dorsally cleft cord in cervical myelomeningocele with the dural dorsal surface. Patients of TCS usually have complaints of radicular pain in both upper limbs along with stiffness, restriction of neck movements, Lhermitte sign and occasionally bowel bladder involvement is also present.

Our objective is to look at the various clinical, radiological, pathological presentations and surgical outcomes in patients with tethered cord.

MATERIALAND METHODS

1. Place and Period of Study: The prospective study was carried out at the Department of Neurosurgery, J.A. Group of Hospitals, G.R. Medical College, Gwalior, M.P, over 25 months from December 2018 to December 2020.

2. Study Design: Prospective Study

3. Study population:
Consecutive patients of tethered cord syndrome were admitted to the department of Neurosurgery, J.A. Group of Hospitals, G.R. Medical College, Gwalior, M.P. over 24 months of period. The present study was conducted on over 40 patients admitted during the above-mentioned duration.

4. Inclusion Criteria:
Patients with clinical and radiological(CT/ MRI Spine) findings consistent with tethered cord syndrome.

5. Exclusion criteria:
Patients with severe co-morbid illness, non-cooperative, and not willing for surgery were excluded from the study

6. Study procedure:
All patients with clinical and radiological diagnostic features of tethered cord syndrome and satisfying the inclusion criteria were considered for the study.

7. Data collection:
Approval for the study was obtained from Ethical Committee (Medical). Data of all the admitted patients were collected as per the proforma.

8. Pre-operative clinical assessment:
All included patients were thoroughly evaluated with preoperative history, detailed clinical examination, radiological assessment using CT scan, MRI of the whole spine, and USG for pre and post-void residual volume and associated congenital anomaly. MRI is now the investigation of choice in patients presenting with TCS, as it helps in delineating structural information which is useful for identification and classification of the underlying pathologies and enables the surgeon to plan further management of these patients. MR imaging is used primarily to identify the level of the conus and the nature of the filum terminale. Patients with thick/normal filum and conus level below the L1-L2 disk were included in the study. Filum was considered to be thick, if the diameter was more than 2 mm, at L5–S1 on magnetic resonance imaging (MRI). CT scans were done in patients with MRI finding suggestive of split cord malformation CT scan of the spine was done.

Spinal ultrasonography has been advocated as the ideal screening tool

for occult spinal dysraphism in young infants (less than 6 months old), given its availability, portability, low cost, and ease of obtaining high-quality images.

Post-void residual volume (PVR) is the amount of urine retained in the bladder after a voluntary void and functions as a diagnostic tool. Post void residue is estimated either by USG bladder or by doing a bladder scan with a foley catheter in situ. Post-void residual evaluation is by measuring the remaining urine in the bladder shortly after a voluntary void. Urinary catheterization is the gold standard for measuring the post-void residual. Measurement of post-void residual immediately after voiding is crucial for accurate measurement, with delays of as little as 10 minutes from bladder emptying to post-void residual measurement potentially causing clinically significant overestimation of post-void residual [10]. USG is used to visualize the bladder both transabdominal and transvaginal. Transvaginal ultrasound appears especially accurate for measuring low bladder volumes[11]. Adults[12] having less than 50mL PVR is adequate bladder emptying, Children[13,14,15]having more than 20mL PVR is considered abnormal, Elderly[16,17,18]patients having 50-100 ml PVR is considered normal.

PVR is assessed within five minutes of voiding. For children aged <6 years, a single PVR >30 ml or >21% BC, or repetitive PVR >20 ml or >10% BC can be regarded as elevated. For children aged >7 years, a single PVR>20 ml or 15% BC, or repetitive PVR >10 ml or 6% BC can be redefined as elevated[19]. By expert opinion, there is no universally accepted definition of a significant residual urine volume. For clinical practice, PVR <30 ml can be considered insignificant, while residual volumes persistently >50 ml could be regarded as important[20].

8. Follow up:
Patients were followed up in Neurosurgery OPD after discharge from the neurosurgery department at an interval of 3 months, 6 months, and then annually. Patients in whom surgery-related complications developed were reviewed more frequently.

RESULT

1. Age-wise distribution(TABLE-1):
Among 40 patients, 24 patients (60%) were in the age group of 0-10 years, and 8 patients (20%) were in the age group of 11-20 years. 3 patients (7.5%) were presented in the 21-30 years of age group.4 patients (10%) were in the age group of 31-40.

S.No	AGE (Yrs.)	No. Of patients	Percentage
1.	0-10	24	60
2.	11-20	8	20
3.	21-30	3	7.5
4.	31-40	4	10
5.	41-50	1	2.5
	TOTAL	40	100

2:Gender-wise distribution:
Male outnumber female in our study, male was present in N=26(65%) while female in N=14(35%). Male to female ratio is 1.85:1. (Table 2)

Table 2: Gender-wise distribution

S.NO	Gender	No. of patient	Percentage%
1.	MALE	26	65
2.	FEMALE	14	35
		40	100

3: Cutaneous stigmata:
Most common cutaneous stigmata were lipoma found in N=13(46.52%) patients followed by dermal sinus in N=5 (17.85%) patients. (Table 3)

Table 3: Cutaneous stigmata in patients with tethered cord

Cutaneous stigmata	No. of patients	Percentage
Lipoma	13	46.42
Dermal sinus	5	17.85
Cervical meningomyelocele	4	14.28
Lumbar meningomyelocele	3	10.71
Tuft of hair	1	3.57
Skin dimple	1	3.57
Lumbar meningocele	1	3.57
Total	28	100

4. Type of spinal dysraphism associated with tethered Cord:
Lumbar lipomyelomeningocele was most commonly found in N= 15(37.5%) patients, N= 5(12.5%) patients had dermal sinus, cervical meningomyelocele in N=4(10%) and lumbar meningomyelocele in N=3(7.5%), thickened filum terminale was found in N=2(7.5%) patients and N= 1(2.5%) patients had diastematomyelia (Table 4).

Table 4: Type of spinal dysraphism associated with tethered Cord

Type of spinal dysraphism	No. Of Patients	Per cent
Lipomyelomeningocele	15	37.5%
o/c/o MMC	7	17.5%
Dermal sinus	5	12.5%
Cervical meningomyelocele	4	10%
Lumbar Meningomyelocele	3	7.5%
Thickened filum terminal	2	5%
Diastematomyelia	1	2.5%
Dorsal Meningomyelocele	1	2.5
Conus dermoid	1	2.5%
Lumbar meningocele	1	2.5%
Total	40	100 %

5. MRI Findings of the position of the Conus:
Most of the patients had conus at the L3 level (N=22, 55%, while 7.5% (N=3) and 12.5 %(N=5) of patients had conus at L2 and L4 level. 22.5% (N=9) of patients had conus at a normal position.

Position of conus	No. of patients	Percentage
L3	22	55%
L2	3	7.5%
L4	5	12.5%
L5	1	2.5%
Normal conus position	9	22.5%
Total	40	100%

6. Type of operation done:(TABLE-6)

Type of spinal dysraphism	Operation	Percentage	Number
Lipomyelomeningocele	Excision of lipoma with the division of filum terminale	37.5%	15
Thickened filum terminal	Excision of filum terminale	5%	2
Lumbar Meningomyelocele	Excision of meningomyelocele	7.5%	3
Cervical meningomyelocele	Excision of meningomyelocele with the division of adhesion.	10%	4
o/c/o MMC	Adhesion release with excision of filum	17.5%	7
Dermal sinus	Excision of the sinus with the division of adhesion	12.5%	5
Diastematomyelia	Laminectomy with excision of the bony spur with excision of filum	2.5%	1
Dorsal Meningomyelocele	Excision of meningomyelocele with the division of adhesion.	2.5%	1
Conus dermoid	Laminectomy with near total excision with excision of filum terminale.	2.5%	1
Lumbar meningocele	Excision with repair	2.5%	1

7. Summary of intraoperative findings (TABLE-7)

Intraoperative finding	No. Of Patients	Percentage
Lipoma with thickened filum terminale	15	37.5%
Adhesion with Thickened filum terminale,	7	17.5%

Dermal sinus+ adhesion + thickened filum terminale	3	7.5%
Adhesion (4 cervical + 1 dorsal sinus+ dorsal MMC+ lumbar MC)	8	20.0%
Myelomeningocele + thickened filum terminale	3	7.5%
Thickened filum terminale	2	5%
Dermoid with the tight filum	1	2.5%
SCM+ Tight filum terminale + Bony spur.	1	2.5%
Total	40	100%

8. Follow-Up of Signs & Symptoms (TABLE-8)

Sign and symptoms	IMPROVED	STABLE	WORSE	RECURRENCE	UNKNOWN	TOTAL
Urinary dysfunction	13(81.25%)	2(12.5%)	0	0	1(6.25%)	16
Bowel dysfunction	5(62.5%)	2(25%)	0	0	1(12.5%)	8
Pain	5(83.3%)	0	0	0	1(16.6%)	6
Motor deficit	8(90.9%)	1(9.09%)	0	0	1(9.09%)	10
Sensory deficit	6(85.71%)	0	0	0	1(14.28%)	7
Sexual dysfunction	0	0	0	0	1(100%)	1

DISCUSSION

The word "Spinal Dysraphism" was coined by B W Liechtenstein in 1940. Spinal dysraphism involves a spectrum of congenital anomalies resulting in a defective neural arch through which meninges or neural elements are herniated, leading to a variety of clinical manifestations.[21] The anatomic level of the myelomeningocele sac correlates with the patient's neurologic, motor, and sensory deficits.

Age and gender distribution

The present study revealed most patients are in the age group of 0-10 years which is slightly different from previous studies. The present study revealed male predominance with a male-to-female ratio of 1.85:1 which is similar to the previous studies. Kafle et al 2017 in a study of 97 patients found that males to females with a ratio of 1.77:1 [22]. B. J. Iskandar, et al.1998 in their prospective study found that there were 12 men and 22 women[23].

Types of associated spinal dysraphism with cutaneous stigmata

Our Study revealed cutaneous stigmata like lipoma in 13(46.42%) patients and dermal sinus in 5 (17.85%)patients. Ailawadhi et al. 2012 in a study of 34 patients noted abnormality of spine curvature in 61.7%, 29.4% patients presented with different types of cutaneous stigmata like a sacral dimple in 1 (2.9%) patient, subcutaneous lipoma in 1 patient (2.9%) or tuft of hair at the back in 5 patients (14.7%). Parchment skin, dermal sinus and skin appendage were seen in 1 patient each. Urological complaints were present in 32.3% of patients, limb weakness in 55.8%, numbness in 17.6%, leg and foot deformity in 32.3% and back pain in 20.5%[24].

In our study, lipomyelomeningocele was found in 15 (37.5%)patients, 5(12.5%) patients had dermal sinus, cervical meningomyelocele 4(10%) and lumbar meningomyelocele 3(7.5%) and thickened filum terminale were found in 2(7.5%) of patients, 1 (2.5%) patients had Diastematomyelia. S. Rajpal et al 2007 in their prospective study on lipomyelomeningocele (25 patients), tight filum terminal (22 patients), SCMs (15 patients), syringomyelia (7 patients), dermoid cysts (two patients), and a meningocele (one patient)[25].

Position of conus

Ailawadhi et al.2012 the conus was low lying at L3 in 20(58.8%) of the total patients[24]. In the present study 22 (55%)out of 40 patients has conus at the L3 level, L4 in 5 (12.5%) patients, L2 in 3 (7.5%) patients, L5 in 1(2.5%) patients and 9 patients had a normal position of conus.

Intraoperative findings

In the present study, intraoperatively 15(37.5%) had lipoma with tight filum terminale, followed by adhesion 8(20%), adhesion with tight filum terminale 7(17.5%), dermal sinus with adhesion with tight filum terminale 3(7.5%), 3(7.5%) had myelomeningocele thickened filum terminale, 2 (5%) had thickened filum terminale and 1 (2.5%) patient each of dermoid with low lying conus, split cord malformation with fibrous septa and dermal sinus with adhesion with tight filum terminale. G. Y. F. Lee, et al. 2006[26] Lipoma + tight terminal filum 15(25%), tight filum terminale 17(28.3%), myelomeningocele 5(8.3%), tight terminal filum with adhesions 4(6.66%), SCM + tight terminal filum 3(5%), dermoid tumour with adhesions 4(6.66%), adhesions 3(5%) myelomeningocele with tight terminal filum 2(3.33%) lipoma with adhesions 2(3.33%) and one patient each of tight terminal filum with lipoma with adhesions 1(1.66%), myelomeningocele with lipoma SCM with lipoma with tight terminal filum and epidermoid terminal filum with S-2 lipoma.

Outcome

For most of the patients, N=13(81.25%) had improvement in urinary symptoms, N=5(62.5%) patients had improvement in bowel symptoms, N=5(83.3%) patients had improvement in pain symptoms, N=6(75%) had improvement in motor deficit, N=6(85.71%) had improvement in sensory deficit. Through the follow-up of 56 cases of adult TCS patients, Huttmann et al [27] found that the pain relief rate was 86%, which was the most obvious symptom that alleviated, the remission rate of the lower limb spasticity was 71%, and the remission rate of bladder dysfunction and feeling movement dysfunction was 44% and 35%, respectively. G. Y. F. Lee, et al. 2006 in their prospective study found that 21 (50%) patients had improvement in urinary symptoms, 29(64%) had improvement in motor weakness, 36(83%) had improvement in back pain symptoms and 20(50%) patients had improvement in sensory symptoms [26].

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