CLINICAL CORRELATES AND SURGICAL MANAGEMENT OF OCCULT SPINAL DYSRAPHISM WITH TETHERED CORD SYNDROME: EXPERIENCE FROM A TERTIARY CARE NEUROSURGERY DEPARTMENT

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ABSTRACT

Background: Open spinal dysraphism is one of the leading cause of congenital central nervous system disorders and contributes to a high proportion of disability and mortality in the younger generations. Occult spinal dysraphism is relatively rare and usually presents clinically once the neurologic sequelae has settled in. The disorder most commonly affects the adolescents and younger children and the most effective treatment is by surgical untethering of the spinal cord.

Methods: The study was conducted from January 2013 to December 2014 at the Neurosurgery Department of Hayatabad Medical Complex Peshawar in a prospective manner involving all patients less than 18 years of age. Clinical characteristics of patients with occult spinal dysraphism presenting with diagnosis of tethered cord syndrome were recorded, surgical management planned and executed and the early postoperative course was followed.

Results: Twenty three patients were admitted during the study period with 12 (52.2%) females and 11 (47.8%) males with a mean age of 9.3 years (SD ± 4.3). The most common presenting complaint was pain in lower limbs (60.9%) and back (34.8%); followed by urinary symptoms (30.4%), lower limb weakness (26.1%) & claudication (26.1%) in that order. The clinical findings included back swelling (65.2%), faun's hairy tail (30.4%), feet ulcers (21.7%), skin dimpling (13%) and kyphoscoliosis (8.7%). Lipomyelomeningocele (34.8%) was the commonest diagnosis, followed by meningocele & diplomyelia (21.7%), dermal sinus & tethering after myelomeningocele repair (8.7%) and one case (4.3%) of filum terminale thickening. The most common operative complication was CSF leak (26.1%) while wound infection occurred in 3 (13%) cases.

Conclusion: Tethered cord syndrome due to occult spinal dysraphism is a rare disease with significant neurologic consequences for the younger generation. Clinical vigilance and good operative treatment could prevent the development of these complications. Further studies are required in order to better delineate the clinical characteristics of this disorder and its operative management.

Keywords: Spinal Dysraphism, Spina bifida Occulta, Tethered Cord Syndrome

INTRODUCTION

Tethered spinal cord was first described by Hoffman in a landmark paper which described the associated characteristics of the condition and its response to surgical release of the tethering.[1] Occult spinal dysraphism is a well-known form of neural tube defects with a relatively rare occurrence and late clinical presentation. It was described by Virchow in 1875 with a recorded first instance of surgical correction performed by Jones in 1891.[2] Tethered spinal cord was described as characterised by low lying conus and thickening of the filum terminale. Similarly tethered cord syndrome was described as lower spinal cord dysfunction due to a constant traction upon the conus medullaris.[3]

A multifactorial aetiological relationship has been described in the development of all neural tube defects which include genetic factors, dietary deficiencies, socioeconomic, environmental, and maternal factors with a variety of unknown contributing aetiologies. Open spinal dysraphism accounts for up to 95% of all neural tube defects while occult spinal dysraphism account for the remaining 5% of the lesions. The apparent paucity of manifest clinical signs at birth, delayed onset neurologic sequelae and low antenatal obstetric coverage of pregnant women in our society has rendered this condition and its diagnosis a very rare clinical entity.[4,5,6]

A diverse variety of occult spinal dysraphism lesions have been described in the paediatric and adolescent population which include lipomyelomeningocele (LMMC), meningocele, diplomyelia or diastematomyelia, thickening of filum terminale, intradural adhesions and adherence of the placode following my-

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elomeningocele repair.[7] Almost all of the lesions require open untethering of the cord in order for the neurologic manifestations to resolve or stop progression.[8,9]

Over the last two decades significant advancement has been achieved regarding the prevention, earlier diagnosis and treatment of this disorder, so as to stop the progression of neurologic deterioration. Antenatal dietary supplementation with folic acid has reduced the incidence globally, frequent clinical visits to the obstetric clinics and increasing expertise in antenatal ultrasound, availability of high frequency ultrasound, antenatal and early post-natal MRI, dynamic or phase contrast MRI has helped in earlier diagnosis, and improvement in micro-neurosurgical techniques has led to improved outcome for these patients.[10,11,12,13,14]

However, the rarity of this clinical condition warrant repeat clinical research in order to improve upon the already achieved developments and as a reminder for the neurosurgical and spine surgical community to remain vigilant regarding earliest possible diagnosis and good surgical management. In a developing countries like Pakistan, the high prevalence of these disorders, low availability of obstetric, advanced diagnostic and tertiary care neurosurgical facilities and decreased awareness among the primary health care physicians warrants widely available current research about this entity of disorders.[4,5,15]

METHODS

This is a prospective observational study conducted at the Department of Neurological Surgery of Hayatabad Medical Complex Peshawar from 1st January 2013 to 31st December 2014. During the study period all patients male or female of age range from 0 to 18 years with a diagnosis of tethered cord syndrome due to spinal dysraphism were included. The data was collected on a predesigned pro forma. Informed consent was taken from parents of all the children regarding their inclusion in the study. The institutes' ethical committee for research evaluation was approached for approval of the proposed study.

All patients who presented to the outpatient department were diagnosed using clinical methods and diagnostic studies such as MRI & CT scan of the spine were performed.

Data was collected about the demographics, presenting symptoms, duration of symptoms, developmental history, parity, maternal education at birth of the child, maternal education, maternal folic acid use, antenatal clinical visits and socioeconomic status of the family.

Clinical examination was recorded for the presence/absence of back swelling, dermal stigmata, scars, lower limb function, sensory deficits, reflexes, presence/absence of kyphoscoliosis or limb deformities and presence/absence of neuropathic ulcers.

Radiologic findings were recorded for diagnosis, the level of spinal tethering, presence/absence of syrinx and spine deformities.

During the surgical operation the diagnosis of the lesion was confirmed and interventional procedure recorded.

Postoperatively the patients were followed up to the date of discharge. During the postoperative course development of complications, any additional procedures performed for complication management and the total length of stay was recorded.

The data was entered and analysed using SPSS version 20.0 and is presented using charts, tables and graphs.

RESULTS

Twenty three patients with 12 (52.2%) females and 11 (47.8%) males were included over the two years period. Mean age of the patients was 9.3 years (SD ± 4.3). The majority of patients belonged to the neighbouring Afghanistan (26%), followed by the districts of Peshawar (21%), Khyber Agency (13%), Sawabi (13%), Charsadda (8%) and other districts (8%) of the province of Khyber Pakhtunkhwa. (table 1 & 2)

The most common presenting complaints were lower limbs or hips pain (60.9%), backache (34.8%), symptoms of urinary tract dysfunction (incontinence, retention, difficulty voiding) (30.4%) and weakness of the lower limbs with gait problems (26.1%). A majority of patients also complained of the presence of back swelling which was present since birth. The mean duration of symptom presence was 4.3 months (SD ± 2.8). (table 2)

On clinical examination the findings included back swelling (65.2%), wasting of the lower limbs (30.4%), ulceration of the feet (21.7%), faun's hairy tail (21.7%), skin dimpling over the lumbosacral area or over the swelling (13%) and decreased power grade of the lower limbs in these patients. Kyphoscoliosis was present in only two (8.7%) patients. The clinical diagnoses included Lipomyelomeningocele (LMMC) (34.8%), Diplomyelia (21.7%), Meningocele (21.7%), Filum terminale thickening (4.3%) and congenital dermal sinus (8.7%). (table 3 & 4) (Figure 1 & 2)

Diagnostic work up also showed the presence of dorsal or dorso-lumbar region syrinx in two of the patients. All patients underwent surgical untethering of the cord with lipoma resection in 8 patients and resection of a bony spur in 5 patients with repair of the dural sac. The level of the lesion was confirmed during the open surgery and it was found to be lumbar in 9 (39.1%) patients, lumbosacral in 9 (39.1%) patients, and occurrence of a bony spur at L1 level in 3 (13%) cases while at L2 level in 2 (8.7%) patients.

Postoperative complications included cerebrospi-

Table 1:

Clinical Variable	Mean	Standard Deviation
Patient's age (years)	9.3 years	± 4.3
Maternal age at birth (years)	31.7 years	± 7.3
Symptoms duration (months)	4.35 months	± 2.8
Length of stay (days)	5.7 days	± 3.0

Table 2: Clinical Presentation

Variable	Frequency	Percent
Gender		
Males	11	47.8%
Females	12	52.2%
Backache	8	34.8%
Lower limbs pain	14	60.9%
Lower limbs Weakness	6	26.1%
Urinary symptoms	7	30.4%

Table 3: Physical Findings

Variable	Frequency	Percent
Lower Limbs Power		
0	1	4.3%
1	1	4.3%
4	5	21.7%
5	16	69.6%
Feet ulcers	5	21.7%
Wasting	7	30.4%
Faun's tail	5	21.7%
Skin Dimpling	3	13.0%
Swelling	15	65.2%
Kyphoscoliosis	2	8.7%

Table 4: Physical findings

Diagnosis	Frequency	Percentage
Dermal Sinus	2	8.7%
Diplomyelia	5	21.7%
Filum Terminale fibrosis	1	4.3%
Lipomyelomeningo-cele	8	34.8%
Meningocele	5	21.7%
post MMC repair	2	8.7%
Syrinx	2	8.7%
Tethered spine level		
L1	3	13.0%
L2	2	8.7%
Lumbar	9	39.1%
Lumbosacral	9	39.1%

Table 5: Complications & their management

Postop complications	Frequency	Percentage
CSF leak	6	26.1%
Wound Infection	3	13.0%
Additional procedures		
Lumbar Drainage	2	8.7%
Reoperation/ dura repair	2	8.7%
post MMC repair	2	8.7%

Table 6: Maternal factors

Variable	Frequency	Percent
Maternal Education		
None	20	87.0%
Primary	3	13.0%
Parity		
1	7	30.4%
2	1	4.3%
3	5	21.7%
4	5	21.7%
5	2	8.7%
6	2	8.7%
7	1	4.3%
Folate use		
Yes	0	0%
No	23	100%
Antenatal visits		
Yes	3	13%
No	20	87%
Socioeconomic status		
Low Income	19	82.6%
Middle Income	4	17.4%

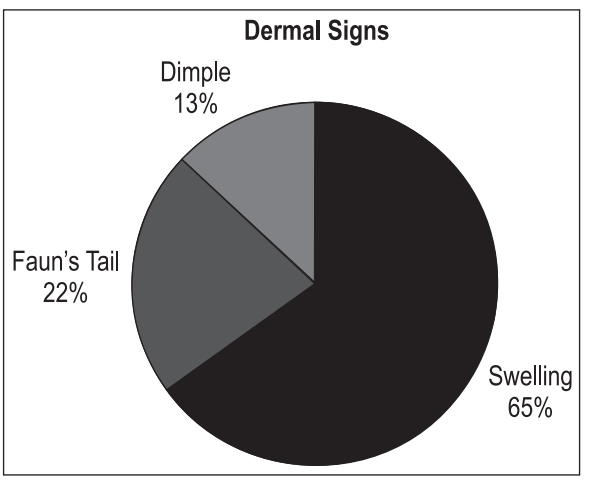

Figure 1:

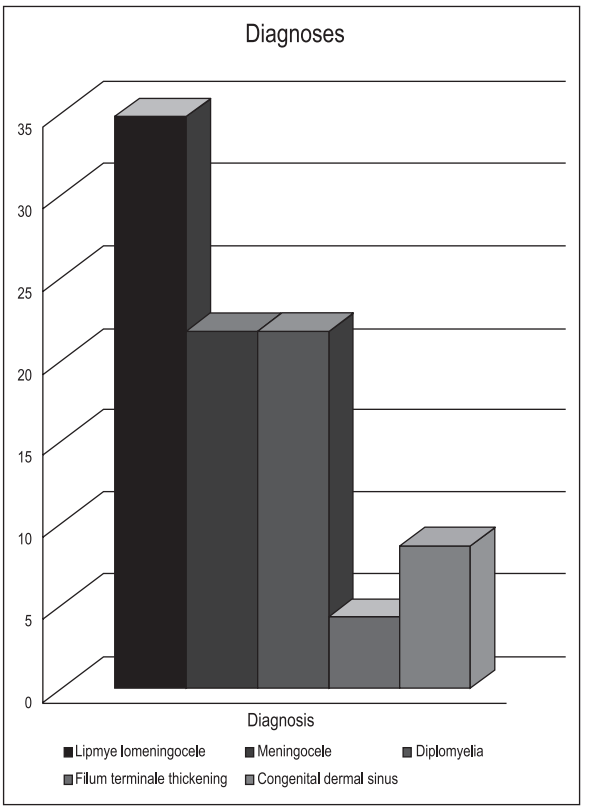

Figure 2:

nal fluid (CSF) leak in 6 (26.1%) patients and infection of the wound in 3 (13%) patients. Only two patients underwent reoperation for dural closure and two patients were treated for CSF leak through insertion of a lumbar drain. Mean length of stay for the patients was found to be 5.7 days (SD ± 3.0). (table 5)

Mean maternal age at the time of birth of the affected child was found to be 31.7 years (SD ± 4.3) (table 1). Out of the 23 mothers, 20 (87%) were found to have not attended a school while only 3 (13%) had formal education up to primary level. None of the mothers used folate during their pregnancy. Only 3 (13%) mothers attended the antenatal clinic, most of them only once during the course of their gestation. Nineteen(82.6%) out of the 23 patients belonged to families with low socioeconomic status while only 4(17.4%) belonged to middle class or fair level of economic status families. (table 6)

DISCUSSION

Spina bifida occulta (SBO) is a rare entity in the spectrum of neural tube malformation which may accompany tethering of the spinal cord causing a myriad array of symptoms and signs. The tethering produce symptoms by various mechanisms, among which ischemic insults to the spinal cord and nerve roots are one of the most favoured explanations. The incidence of tethered cord syndrome (TCS) is reported to be in the range of 0.05 to 0.25 per 1000 births.[16] The clinical features of occult spinal dysraphism and associated TCS are insidious in nature and in some instances they may be entirely absent until the neurological sequelae becomes apparent. The symptoms of the TCS may sometimes appear later in adolescence or adult life and it has been shown that untethering by surgery has very satisfactory results in terms of neurological improvement.[16,17,18] The insidious nature of the tethering of the spinal cord warrants a heightened clinical vigilance for both the general physicians as well as the specialists.[2] Iskandar et al has shown the obvious advantage of surgical intervention for TCS in terms of neurological and functional improvements, despite the wide debate about the feasibility of intervention in adolescence and adult patients.[19]Therefore, it is apparent that surgical intervention especially in the presence of symptomatic TCS has clear benefits over the wait and watch approach.[2,20]

The spectrum of SBO varies in frequency with respect to the individual lesions, however, diplomyelia and lipomyelomeningocele collectively form about 60% of the total.[16,20] Quite similar are the findings in our study where more than 56% of patients comprised of LMMC (34.8%) and diplomyelia (21.7%). The majority of paediatric patients present within an age range of 7 to 12 years (mean 9 years in our study) with complaints of pain occurring in legs, hips or back (95.7% collectively in our study) followed by dysfunction of the bowel or bladder (30.4%), various degrees of motor deficits, neuropathic ulceration of the feet, orthopaedic deformities and dermal stigmata in the form of hair tuft over the lumbosacral area, dimples and swelling caused by the underlying lipomata. These findings have been noted in the studies by Das et al, Sarris et al, Muthukumar et al, Solmaz et al, Koo et al, Phi et al and Iskandar et al.[2,7,11,12,16,17,18,19] Findings in our study as shown in table 2 & 3, concur with the findings of the above studies. These consistencies in the findings of various studies denote that the presenting symptoms of SBO and TCS are widely constant, despite differences in patterns of appearance, and can be relied upon in a clinical perspective.

Magnetic Resonance Imaging has been shown to be the diagnostic tool of choice for defining the specific lesion of SBO and the level of tethering of the spinal cord.[2,16,22] Das et al has also described the efficacy of ultrasonography for defining SBO in very young neonates. However, for older patients MRI has superseded CT scan as it delineates the soft tissue pathology clearly and defines the tethering level.[2] In addition to defining the spinal dysraphism, MRI was used by Erfani et al and Gharedaghi et al for evaluating the presence or absence of syrinx and its resolution after surgery and also for the determination of congenital spinal orthopaedic deformities such as congenital scoliosis.[23,24] MRI was the principle investigation after clinical diagnosis in our study. However, we tried to further confirm the lesion during surgery along with its spinal level which

was shown by MRI during preoperative workup. This strategy helped us additionally in confidently classifying the lesion according to their type as found on open surgery.

Open surgical release of the tethered cord is the standard method of intervention in symptomatic cases of TCS due to SBO. There has been increasing use of release and repair of TCS under magnification and with the use of motor and sensory evoked potentials.[16,22] It has also been shown that these two modalities helps improve the postoperative results in terms of functional improvement and complications avoidance. Four principle surgical methods are recommended by a review by Das et al[2] namely;

- Laminectomy & detethering,
- Lipoma excision,
- Resection of bony spur, and
- Division of filum terminale.

We used resection of the lipoma/spur and release of the dural sac from any adhesions as well as repair under magnification. Motor or sensory evoked potentials were not used. A very limited untethering approach was used for those who had minimal or no motor deficits and only the dural sac was released taking care of the nerve rootlets.

Postoperative course of these patients usually is a dramatic improvement in the symptoms of pain with only a minority of patients experiencing a transient worsening of sensory or motor deficits. We, however, followed the patients for occurrence of early postoperative complications. The two major postoperative complications which affects the early postop course was CSF leak (26.1%) and wound infection (13%). No patient experienced a worsening or appearance of new motor deficit in our study, although it has been reported in 3.9% to 5% patients of untethering surgery.[22]

Wound infections responded quickly by instituting barrier dressing methods and the use of intravenous antibiotics according to the sensitivity patterns. The mean hospital length of stay (LOS) was found to be 5.7 days (table 1). Only two (8.7%) instances of reoperation for repair of the dural defect in cases of CSF leak were found and two (8.7%) cases of CSF leak were relieved with insertion of a lumber drain above the region of the dura repair. Most of the CSF leaks occurred in patients who had a meningocele sac repaired at operation. This signifies the fact that the dura in this subset of patients is very thin and closing sutures usually holds poorly. A meticulous closure technique is therefore advised. Studies by Solmaz et al and Galhom et al employed similar techniques of untethering and repair of the thecal sac with complications of CSF leak and wound infections (20% CSF leak and 10% wound infection rates) similar to our study. The mean LOS was similar to the above studies as well as a study by Shweikeh et al where the mean LOS was 3.5 to 5.1 days.[25]

Studies by Golalipour et al and Grewal et al have shown the significance of association of maternal, socioeconomic and dietary factors on the incidence of neural tube defects.[13,14] These studies have found significant association between maternal folate intake, maternal education level, age at the time of pregnancy, parity and antenatal visitations. Findings of our study are consistent with the above studies and it was noted that a very high incidence of spinal dysraphism is noted in mothers of low socioeconomic status, no folate intake, low education and increasing age at the time of birth of the child as shown in table 6. Although the rural population in our country and the neighbouring Afghanistan has a simple diet, it cannot be adequately said that the diet is sufficient in its contents of various minerals and vitamins. However, the major factor for high rural prevalence of neural tube defects could be the overall non-availability of food due to poor socioeconomic situations. Literacy of the female population is very low and in our study only three of the mothers were educated up to primary level while the rest had no formal education.

CONCLUSION

Early diagnosis and optimal treatment strategy development is the key to successful outcome in patients with TCS due to SBO. Earlier diagnosis is only possible if high degree of clinical suspicion is observed, especially by the primary care physicians and neurosurgeons. Further studies are required to further elucidate the association of various clinical correlates with occult spinal dysraphism and tethered cord syndrome and to establish their diagnostic and prognostic role.

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