Original Article

Surgical Treatment of Split Cord Malformations (SCM) In Children

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ABSTRACT

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Background: Split cord malformation (SCM) is a term used for diastematomyelia and diplomyelia. Its incidence is decreasing in the world literature. Objective: This study aims at evaluating the outcome of surgery for SCM in eleven children operated upon at the Neurosurgery Department of Benha University Hospitals and at the Benha Specialized Children Hospital between March 2013 and June 2015. Patients and Methods: This prospective clinical cohort study includes eleven patients, five boys and six girls, with age ranging from one day to twelve years ((38.2 months ±6.7 SD) and magnetic resonance imaging (MRI) -proven SCM. Results: all six patients with back/leg pain recovered completely as regard leg pain and improved as regard the back pain. The two patients with numbness/parathesias also improved greatly. Two of the three patients with leg weakness were neurologically stable while one patient, operated upon at the age of 11 days for SCM associated with meningiomyelocele, returned with retethering, proved on re-exploration to be due to arachnoid scarring. All six patients with no neurologic deficit showed no neurological deterioration. In six patients pre- and postoperative urodynamic studies were performed and showed no great change. Conclusion: We recommend early surgery for SCM to avoid or minimize the suspected neurologic deficit. Outcome of the patients with SCM is generally gratifying. All the tethering elements must be taken care of to release the spinal cord.

INTRODUCTION

Split cord malformation (SCM) is a term used for diastematomyelia and diplomyelia. Its incidence is decreasing in the world literature. Pang & colleagues classified SCM into type I and type II. Furthermore, SCM can be complex SCM. Surgical treatment is indicated in all patients diagnosed with SCM.

PATIENTS AND METHODS

Study design:
This is a prospective clinical cohort study of eleven patients operated upon for SCM with two years follow up period at the Neurosurgery Department of Benha University Hospitals and Benha Specialized Children Hospital between March 2013 and June 2015.

Patient population:
Eleven patients, five boys and six girls (table 1), with age ranging from one day to twelve years (38.2 months ±6.7 SD) with MRI –proven SCM.

Preoperative work-up:
All patients underwent routine physical examination for neurocutaneous stigmata and/or skeletal deformity; they also received a thorough neurological evaluation. Neuroimaging studies of the spine consisted of plain radiography both in antero-posterior and lateral projections, computerized tomography (CT) and Magnetic Resonance imaging (MRI). Urodynamic studies were done in six patients (older than six years).

Operative note:
Laminotomy was carried out in all the patients. With type I SCM, bony spur was excised and two dural tubes were converted to one dural tube. In type II SCM, after dura was opened, all the extradural and intradural bands were excised to untether the cord. In all patients, the filum terminale was also excised to release the traction on the conus.

Postoperative follow up:
All patients were examined immediately postoperative, in the early postoperative period for 2-3 days before discharge and frequently every week for the first 3 months and every 3-month period thereafter. Examination included general physical and neurological examination. Plain radiograms and computed tomograms were made immediately postoperatively and every 3 month period. Urodynamic studies were done in six patients 3 months postoperatively.
RESULTS

In this study, there were 5 boys and 6 girls with a mean age of 38.2 years (Table 1). Back and leg pain was the most common symptom (54.5%) (Table 2). In clinical examination, no deficit was in 6 patients while leg weakness and numbness/parathesias were in 3 and 2 patients respectively.

<table>
<thead>
<tr>
<th>Table 1: Summarizes the criteria of patients</th>
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<tr>
<td><strong>Value</strong></td>
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<tr>
<td>Sex</td>
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<td>Boys</td>
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<tr>
<td>Girls</td>
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<tr>
<td>Age</td>
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<td>N= number of patients</td>
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There were 7 cases of SCM type I and 4 cases of SCM type II (Table 3). One of our eleven patients had SCM type II associated with meningiomyelocele. In SCM type I, location of bony spur was in distal part between two hemicords except one patient proximal.

<table>
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<th>Table 3: Summarizes the site and type of SCM in our patients</th>
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<td><strong>Site of SCM</strong></td>
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<tr>
<td>Lumbar</td>
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<td>Lumbosacral</td>
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<tr>
<td>Dorsal</td>
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<tr>
<td><strong>Type of SCM</strong></td>
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<tr>
<td>SCM type I</td>
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<tr>
<td>SCM type II</td>
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Laminotomy was carried out in all the patients using Midas Rex high-speed drill or by using fine Kerrison ronguer.

In all patients with type I SCM (Figure 1), bony spur was excised by using fine Kerrison ronguer (1mm) or by using diamond drills. In case of type I SCM, two dural tubes were converted to one dural tube. The anterior dura might left open or closed and the posterior one was closed after excising intradural bands and median nerve roots to untether the spinal cord.

In case of type II SCM (Figure 1), after the laminotomy, the dura was opened. All the extradural and intradural bands were excised to untether the cord.

In all patients, the filum terminale was excised to release the traction on the conus.

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**Fig. 1a-f:** a.b.c: Intraoperative views of SCM type 1 and d.e.f: SCM type 2. a: dissection of incomplete lamina. b: bony spur exposed. c: two separate hemicords with its meninges. d: opened one dura and separatdhemicords with fibrocartilagenous band. e: after band excision and f: watertight dural repair.
Only one of our patients had early postoperative CSF leak that improved on conservative treatment with prone position, acetazolamide therapy and no further re-surgery was required.

The follow up period ranges from three months to three years by clinical evaluation and neuroimaging studies (Figure 2) with a mean of 16.8±7.5 month.

**Fig. 2a-c:** Neuroimaging studies of SCM type 1; a: pre-op CT axial view and b: pre-op T1W MRI axial view show bony spur and two separate hemicords. c: post-op 8 months T1W MRI axial view shows no regrowth of spur.

**Surgical outcome:**

All six patients with back/leg pain improved completely as regard the leg pain and greatly as regard to back pain.

The two patients with numbness/parathesias improved.

Two of the three patients with leg weakness were neurologically stable while one patient, operated upon at the age of 11 days for SCM associated with meningiomyelocele, returned with increased leg weakness at the age of 2 years. MRI showed retethering that proved on re-exploration to be due to arachnoid scarring. Detethering of the bands was done. She is neurologically stable after one year of the second operation.

All six patients with no neurologic deficit showed no neurological deterioration.

No change occurred in the six patients when comparing their pre- and postoperative urodynamic studies.

**DISCUSSION**

Split cord malformations (SCM) is a term used for diastematomyelia and diplomyelia. It was considered a form of occult spinal dysraphism.² Off late, with slight modification of Pang’s classification by Kumar et al, it is now obvious that SCM is not merely an entity of occult dysraphism, it may coexist with open spinal dysraphism, like meningomyelocele (complex spina bifida) in significant number of cases.³⁶⁷⁸ Among the MMC, SCM is reported in 10% cases.⁹ Sometimes the MMC arises from one hemicord.⁹¹⁰ One of our eleven patients had SCM associated with meningiomyelocele.

There are several classifications of SCM. Pang et al. classified SCM into type I and type II.³ Furthermore, SCM can be complex SCM.⁸ The complexity is because of associated anomalies like lipoma, teratoma and dermo-epidermoid tumors.¹¹ Another classification is into ventral and dorsal type I spur. SCM could be in single site or in multiple sites. Gupta and Mahapatra classified type I spur into a-d types, depending on the location of spur in between the proximal part and distal part of the split, the space available above and below the spur.¹² This classification has a direct relation to the surgical approach and risk of postoperative deterioration. Thus, the type of SCM I can determine the surgical approach and risk of postoperative deterioration. Thus, the type of SCM I can determine the surgical approach and risk of postoperative deterioration. This classification has a direct relation to the surgical approach and risk of postoperative deterioration.

Skin stigmata are well recognized and reported in occult spinal dysraphism with SCM.¹⁶ Izciet et al., in 2007, highlighted the diagnostic value of skin marker.¹⁷ Mahapatra found that SCM was most common in lumbar area (55%) followed by dorsolumbar area in 23%. In 8% it was present in lumbosacral area. Only in 3%, the SCM was found in cervical region. Long segment of SCM type I was noticed in5% and it was found in multiple sites in 2% patients.¹² In our study, SCM type I in 63.6% while 36.4% was SCM type II. One patient had SCM type II associated with meningiomyelocele. In SCM type I, location of bony spur was in distal part between two hemicords except one patient proximal.

In our study, only two patients had dorsal lesions (18.2%) while nine patients were lumbar (45.5%) and lumbosacral (36.4%).

Kumar et al reported that operative site CSF leak was seen in 24.5% of patients, Meningitis in 12.2% of patients, superficial wound infection in 8.2% of patients and mortality of 2% of patients.¹⁹ Only one of our
patients had early postoperative CSF leak that improved on conservative treatment with prone position and acetazolamide therapy and required no further re-surgery.

Geyik et al. found that postoperative improvement of back pain (61.6%), neurological manifestations (17.2%) and urological problems (11.5%).

In our study, all six patients with back/leg pain improved completely as regard to leg pain and greatly as regard to back pain. The two patients with numbness/paresthesias improved greatly. Two of the three patients with leg weakness were neurologically stable while one patient, operated upon at the age of 11 days for SCM associated with meningiomyelocele, returned with increased leg weakness at the age of 2 years. MRI showed retethering, proved on re-exploration to be due to arachnoid scarring. Detethering of the bands was done. She is neurologically stable after one year of the second operation. All six patients with no neurologic deficit showed no neurological deterioration. All six patients with pre- and postoperative urodynamic studies showed stabilization in urinary function. Proctor and Scott reported a good long-term outcome of SCM surgery with 70-80% improvement or stabilization of neurological function. They also advocated performing urodynamic studies in all patients with SCM.

No regrowth occurred in our patients till the end of follow up period. Kumar et al. reported that approximately 7-10% patients develop retethering over a period of 5 years. One of the causes of the retethering is the regrowth of the spur and development of postoperative arachnoiditis. Gupta et al., reported regrowth of a bony spur in 2010. Thus, regrowth of the spur must be carefully evaluated and excised, in case the patient starts deteriorating in neurological status.

CONCLUSION

We recommend early surgery for SCM to avoid or minimize the suspected neurologic deficit. All the tethering elements must be taken care of to release the spinal cord. Outcome of the patients with SCM is generally gratifying.

REFERENCES