REVIEW ARTICLE

Split Cord Malformation (Occult Spina Bifida) : An Indian Scenario

Raj Kumar

Introduction

Split cord malformations (SCM) is a new term used for diastematomyelia and diplomyelia. It is a form of occult spinal dysraphism(1). Occult spinal dysraphism is defined as skin covered lesion that have no exposed neural tissue and no visible cystic mass (2), barring few cutaneous anomalies, which are indicators of underlying spinal pathology (3). SCM constitutes about one third cases of spinal dysraphism, where spinal cord is divided over a portion of it's length into two equal or unequal halves(3).

SCM are of two types, type 1 consists of two hemicords, each contained within its own dural sheathed tube and separated by a median bony spur and type 2 consists of two hemicord housed in a single dural tube separated by a fibrous median septum (4) (Fig. 1 & 2).

SCM and complex Spina Bifida:

Off late, with slight modification of Pang's classification by Kumar R *et al*, it is now obvious that SCM is not merely a entity of occult dysraphism, it may coexist with open spinal dysraphism, like meningomyelocele (complex spina bifida) in significant number of cases (5) (Fig. 3). It is surprising that about 40.8% cases of all SCM present with meningomyelocele, suggesting that open and closed form of spinal dysraphism may coexist (3).

Clinical Profile: The cases of SCM manifest with variable cutaneous markers, neurological deficits and neuro-orthopedic syndrome (Fig. 4). Many associated craniospinal anomalies are also encountered in these cases (6). Various cutaneous features include, a tuft of hair (hypertrichosis) over spine, cutaneous hemangioma,

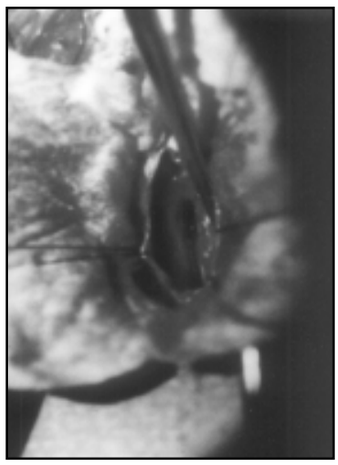

Fig. 1 Intraoperative prints showing type II SCM, the two cords can be seen separately following surgical detethering.

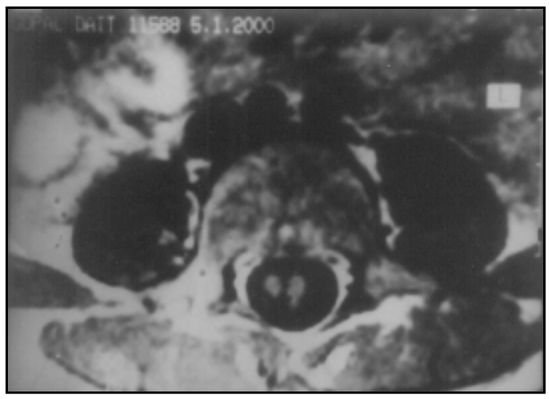

Fig. 2 MRI, axial cuts showing two cord separated by fibrocollagenous spur.

From The Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow - 226014, U.P (India).
Correspondence to: Dr. Raj Kumar, Associate Prof. Deptt. of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow.

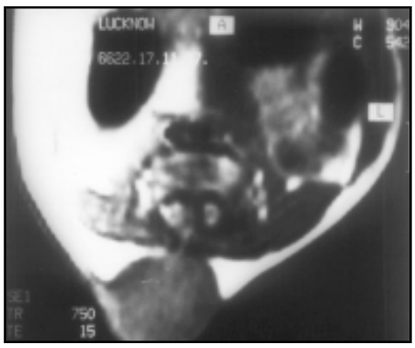

Fig. 3 MRI, axial cuts showing two hemicords with meningomyelocele sac at same level.

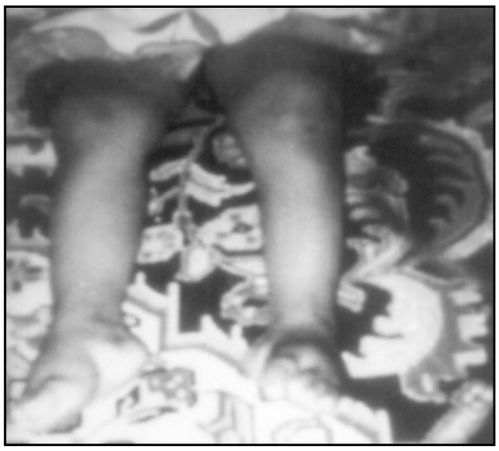

Fig. 4 Child of SCM showing deformity of his feet (Neuro-orthopedic syndrome).

lipoma and dermal sinus etc. Forty seven percent cases of SCM present with one or other cutaneous stigmata (7). However the frequency of individual spin marker is shown in Table 1 A. The incidence and clinical profile of children horboring SCM are tabulated as below:

Table 1 A. Cutaneous markers in patients with split cord malformation

	Cutaneous Manifestations	% of Patients
1.	Hypertrichosis	32.7
2.	Cutaneous hemangioma	10.2
3.	Subcutaneous lipoma	8.2
4.	Dermal sinus	8.2
5.	Multiple cutaneous neurofibroma	2

Table 1 B. Showing the incidence of Neuro-orthopedic syndrome in SCM cases

	Neuro-Orthopedic Syndrome	% of Patients
1.	Scoliosis	32.7
2.	Congenital talipes equino varus	22.4
3.	Leg length discrepancy	10.2
4.	Congenital dislocation of hip	4.1
5.	High arch foot	2
6.	Valgus foot deformity	2

Table 1 C. Neurological features in SCM

	Neurological Examination	% of Patients
1.	Motor weakness of lower limbs	49
2.	Graded sensory loss	40.8
3.	Sphincteric involvement	28.6
4.	Trophic ulcer on feet	8.2
5.	Autamputation of toes	4.1
6.	Backache	4.1

Radiological Finding: Since the spinal dysraphisms are the anomalies of entire central nervous system, hence following associations are found on craniospinal imaging of the children harboring SCM. It is realized that the screening of entire CNS is important, while investigating the children of spinal dysraphism for a complete evaluation of associated anomalies for a successful treatment and good outcome (3, 7, 8).

Table 2

	Image Findings	% of Patients
1.	Meningomyelocele Sac (MMC)	40.8
2.	Low lying Cord	57.1
	- Pure SCM	38.8
	- Complex SCM	18.4
3.	Hydrosyrinx	22.4
4.	Intraspinal lipoma	22.4
5.	Hydrocephalus	18.4
6.	Thick/Fatty filum	14.3
7.	Dermoid	8.2
8.	Arachnoid cyst	4.1
9.	Neurenteric cyst	4.1
10.	Chiari malformation	4.1

Treatment

Because the cord is tethered and fixed in SCM by either a bony spur or fibrous septums, hence the purpose of the surgery is to untether the cord by resecting the osseous, spur or fibrous septum and removing the other tethering lesion(s), to restore the distorted anatomy as far as possible.

Level of Split Cord Malformation

Dorsal cord is the most common site for SCM in 38.8% patients, followed by lumbar region in 28.6%, dorsolumbar in 22.4% lumbosacral in 4.1% and cervicodorsal region in 6.1% cases (3). In 28.6% patients SCM is found at 2 to 3 level higher than the associated MMC sac, whereas in 12.2% children SCM and MMC are encountered at the same level (3).

Postoperative complications

Following surgical intervention, operative site CSF leak is seen in 24.5% of patients, 50% of these may require re-exploration of wound and duraplasty, while rest improve on conservative treatment with prone position and acetazolamide therapy, 16.3% children may develop pseudomeningocele to require resurgery. 12.2% children develop meningitis and most of them respond to antibiotic therapy. Mortality is rare in these children (2%) if managed properly at well equipped centers while 8.2% of patients may develop superficial stitch line infection and all of them respond to antibiotics (3).

Surgical outcome

Various degree of improvement in neurological status is noted in the follow up (average 3.5 year) of these patients. Motor weakness improves to variable extent in 20.4% of patients, 28.6% of cases have no change in motor power compared to preoperative grade, 26.5% children show graded improvement in their sensory loss, whereas about 73% patients remain same as before operation. About 20.4% of patients improve to become able to control voiding of urine to variable extent, but no alteration in bladder function is observed in 8.2% cases. Trophic ulcer of foot completely heals in all 8.2% children. The children with back pain are completely relieved of their pain following surgery. All patients with neruo-orthopedic deformities neither improve nor deteriorate, following a successful surgery. The patient of SCM associated meningomyelocele have relatively poor outcome in comparison to pure SCM cases following surgery (9) (Table 3).

Table 3. Outcome following surgery in children with split cord malformation.

	Improved %	Static %
Motor Weakness	20.4	28.6
Sensory Dysfunction	26.5	14.3
Sphincteric Function	20.4	8.2
Backache	4.1	
Trophic Ulcer	8.2	

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