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Original Article

Analytical Study of Occult Spinal Dysraphism - Its Varied Presentations, Management and Outcome in South Indian Population

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Abstract

Introduction: Occult spinal dysraphism (OSD) represents a subset of spinal dysraphism in which the congenital defects are covered by intact skin. The risk of neurologic deterioration exists at all ages and increases with time and is frequently progressive. The detection and early treatment of such a subtle cutaneous anomaly in a child may be crucial to future neurologic, urologic, and orthopedic development.

Aim: To know demographics, presenting symptoms, clinical features, imaging, and surgical findings of the patients presented with OSD and to assess the outcome and to establish the necessity of surgery before the development of neurological deficits.

Methods: This study was conducted at Department of Neurosurgery, GRH, Madurai, during 2007-2011. A total of 50 patients presented with OSD were evaluated. Age, sex, presenting complaint, past history, cutaneous findings, neurological deficits, imaging findings, associated abnormalities, and outcome of surgery were assessed.

Results: Congenital dermal sinus was presented in 29 out of 50 patients (58%). The most common location of congenital dermal sinus tract in this study was lumbar region. The least common location of the sinus was at cervical region. 18 patients (36%) presented with neurological deficit. All patients had undergone surgical management with detethering. All patients were followed up at regular interval for 2-30 months. One patient developed recurrence of dermoid and another one showed deterioration of motor and autonomic deficits with the presence of pus in the tract. There were no symptoms of worsening or evidence of retethering in all other patients.

Conclusion: The presence of cutaneous stigmata over the midline neural axis should not be considered benign and must initiate prompt radiologic evaluation and neurosurgical referral. All patients with OSD should be offered aggressive surgical treatment.

Key words: Congenital dermal sinus, Dermoid, Detethering, Occult spinal dysraphism

INTRODUCTION

Occult spinal dysraphism (OSD) represents a subset of spinal dysraphism in which the congenital defects are covered by intact skin. Such anomalies united by similar embryological causes, common forms of presentation, and a propensity for multiple expressions within a single patient. These anomalies are numerous and include lipomyelomeningocele, hypertrophied lipomatous filum terminale, anterior and posterior meningoceles, myelocystoceles, split cord malformation, neuroenteric cysts, dermal inclusions, and terminal syringomyelia in addition to the ubiquitous sacral dimples.

The natural history of such abnormalities is variable and often unpredictable. Although some individuals remain asymptomatic throughout adulthood, others may develop progressive dysfunction of the lower limbs and bladder. The insidious fashion in which such complication develops may lead to irreversible damage to any symptomatic manifestation. The risk of neurologic deterioration exists at all ages and increases with time and is frequently progressive.

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These syndromes are important for all physicians to recognize because early diagnosis and treatment can prevent sequelae of progressive neurological deficit. In this era of magnetic resonance imaging (MRI), asymptomatic OSD is recognized very early and presents a dilemma in the management. Prophylactic surgery has a high likelihood of changing the natural history in which a gradual loss of function may be replaced by clinical stability or improvement. The onset of signs and symptoms may be so gradual that a patient's initial presentation to a neurosurgeon may not be until adulthood. The detection of such a subtle cutaneous anomaly in a child may be crucial to future neurologic, urologic, and orthopedic development. It is for all of these reasons that it behooves the neurosurgical community to be knowledgeable about this complex and fascinating group of problems.

Aims and Objectives

To know the demographics, presenting symptoms, clinical features, imaging, and surgical findings of the patients presented with OSD and to assess the outcome and to establish the necessity of early surgery before the development of neurological deficits.

MATERIALS AND METHODS

This study was conducted at the Department of Neurosurgery, GRH, Madurai, during 2007-2011. A total of 50 patients presented with OSD were evaluated. Age, sex, presenting complaint, past history, cutaneous findings, neurological deficits, imaging findings, associated abnormalities, and outcome of surgery were assessed. All patients had undergone surgical management with exploration, excision of cutaneous lesions such as skin tags, DST, dimples, intradural exploration of sinus tracts, dethering of the tethering, removal of associated abnormalities, and subjected to histopathological examination. Pus when present was sent for culture and sensitivity. Patients were followed up at regular intervals according to their post-operative status with particular care to the symptoms of retethering. Duration of follow-up was 2-30 months.

Statistical Methods

The information collected regarding all the selected cases were recorded in a Master Chart. Data analysis was performed with the help of computer using Epidemiological Information Package (EPI 2002). Using this software, range, frequencies, percentages, means, standard deviations, Chi-square and *P* values were calculated. Kruskal–Wallis Chi-square test was used to test the significance of the difference between quantitative variables and Yate's test for qualitative variables. A $P < 0.05$ is taken to denote significant relationship.

RESULTS

A total of 50 patients with OSD who were evaluated and underwent surgical management during a period of 5 years, from 2007 to 2011 were included in this study. A total of 46 patients (92%) were seen in pediatric age group. Only four patients (8%) were seen in adult age group. There were 26 male and 20 female in pediatric age group. In adults, three were male and one was female (Table 1).

The M: F sex ratio in adult was 3:2 and in pediatric age group was 1.4:1. When both age groups were combined, the M: F ratio became 1.4:1. Lowest age in this group was a female child of 10 days old. A 27-year-old female was the one with the highest age, presented with congenital spinal dermal sinus. The median age of the patient with OSD in our study was 3 years.

Most of the patients presented had congenital dermal sinus (CST), and CST was presented in 29 out of 50 patients (58%). The most common location of congenital dermal sinus tract in this study was lumbar region. The least common location of the sinus was at cervical region. Sinus in dorsal location was seen in one adult male, one adult female, and three pediatric male patients.

Sinus in cervical location was seen in one male and three female patients of pediatric age group. Regarding the number of the sinuses, 30 sinuses were seen in 29 patients because one patient in pediatric age presented with two sinuses at different spinal levels. Two male patients in adult age group and eight patients (5 males and 3 females) in pediatric age group were presented with sinus in the lumbar region. Dermal sinus tract in the lumbosacral region was seen in five female and four male patients of pediatric age group.

Twelve patients (48%) in this study came for their cutaneous manifestation only like sinus tract or hairy patch on their back.

Eighteen patients (36%) presented with neurological deficits in this study. Out of them, 17 patients belonged to pediatric age group. One adult who presented with the deficit was a female patient. Three patients presented with

Table 1: Age wise incidence

Age group (in years)	Cases
	n (%)
0-14	46 (92)
14 and above	4 (8)
Total	50 (10)
Range	<1 month - 27 years
Mean	6.2 years

the acute neurological deficit. Regarding the type of deficits, bladder, and bowel disturbances were common and seen in most of the patients presented with neurological deficits. Two patients presented only with autonomic disturbances. Two patients presented with radicular pain in the lower back. One adult male and one pediatric male patient came for pain around the sinus tract. One female patient in pediatric age came for restriction of neck movement only. Four patients came either for local infection or discharge from the sinus. There was no history of the previous surgery or exploration of tracts in any of the patients. Past history of episodes of meningitis was present in one patient (Figure 1).

We have noticed scoliosis in one female and four male in the adult group and one in pediatric group. Spina bifida was seen all except one adult male patient in patients with dermal sinus. Spina bifida was not localized to dermal sinus tract path. It was seen in adjacent levels in all patients. Spina bifida was the most common skeletal abnormality seen in this study. Scoliosis was seen in 5 dysplastic costal elements with hemi vertebrae were seen in three patients. Block vertebra and foot abnormality were not seen. Sacral agenesis of Pang type 3 was seen in one patient with Type 1 SCM (Table 2).

It was possible to demonstrate sinus tract in MRI in all patients. Tethering of spinal cord was seen in all patients.

Therapeutic Approaches

Good surgical planning is very important when evaluating these lesions. In the case of filum terminale dysgenesis, management principally involves division of the filum. The surgical approach generally involves exposure of the filum through a limited laminectomy at L5 to S1 although 15% of filum may fuse above S1 and 11% fuse off the midline.

A more generous exposure may be accompanied by a laminoplasty from L3 to L5. After exposure of the cauda equina, filum and the surrounding nerve roots are often easily identified. The filum may be truncated at a single location or at two separate points.

DST in the lumbar and lumbosacral region was always associated with low-lying conus in this study.

In this study, intramedullary dermoid was seen in five patients (10%) out of which four are seen in association with DST. Terminal syrinx was seen in six patients (12%). Neuroenteric cyst, SCM Type 1, anterior sacral meningoceles, and terminal lipoma were seen in one patient each. Brain screening did not demonstrate hydrocephalus in any of the patients.

Regarding cutaneous findings, the presence of simple dermal sinus was the manifestation in 29 patients. Swelling in the lumbosacral region was seen in 16 patients, and gluteal swelling was observed in one patient. Hypertrichosis was seen in 11 patients. Skin tag was seen in four patients. Dimples were seen in two patients. Surgery was done in all patients with intradural exploration. In DST, the presence of pus was demonstrated in three patients, and all of them were presented with acute neurological deficits. One patient presented with evidence of surrounding inflammation around dermal sinus. Intramedullary dermoid was an associated finding in two of these patients. Culture and sensitivity of pus revealed no growth. However, the anaerobic culture was not done in these patients. Near total improvement, partial improvement, and worsening were seen one in each, who presented with the acute deficit.

Those who presented with deficits (8 patients), near total recovery was seen in one, partial recovery was seen in two, and worsening of symptoms was seen in two patients. In three patients, there was no improvement of symptoms postoperatively. Four patients who had dermoid cysts were presented with deficits and one patient who had inclusion dermoid presented with low back and radicular pain. In this group near total recovery was observed in two patients and partial recovery, worsening and no improvement of symptoms was seen in one patient each. One patient developed post-operative CSF leak and wound infection which was treated successfully in a conservative manner. Excised specimens were subjected to histopathological examination, and the presence of stratified squamous epithelium lining the tract was confirmed in all DST.

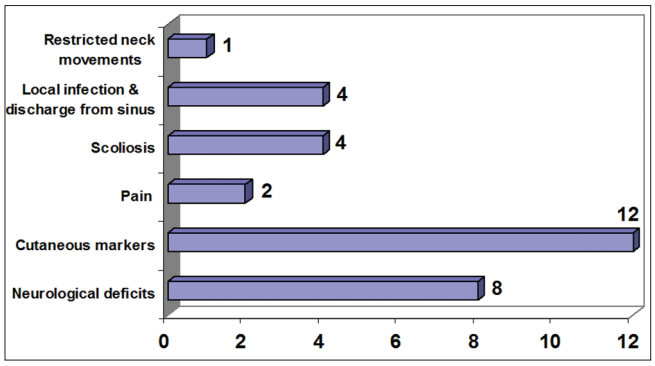

Figure 1: Reason for attending consultation

Table 2: Incidence of skeletal abnormalities

Skeletal abnormalities	Patients
SCM	3 (6)
Spina bifida	27 (54)
Block vertebra	Nil
Scoliosis	5 (10)
Costal dysplasia with hemi vertebrae	3 (6)
Sacral agenesis	1 (4)

All patients were followed up at regular intervals according to their post-operative status (Table 3). One patient developed recurrence of dermoid and presented with paraparesis after improvement of deficits in initial surgery. There was no improvement after re-exploration and the patient lost for follow-up later. Another patient showed deterioration of motor and autonomic deficits who presented with the acute neurological deficit and the presence of pus in the tract and subarachnoid space. There were no symptoms of worsening or evidence of retethering in all other patients till date. The duration of follow-up was 2-30 months.

DISCUSSION

A total of 29 patients with congenital spinal dermal sinus treated in our department during the past 5 years were evaluated, and the factors such as age, sex, location of the sinus, presenting features, neurological deficits, cutaneous markers, MRI findings, intraoperative findings, and operative outcome of the patients were assessed. The findings of this study were compared with the study of Ackerman and Menezes[1,2] which reflects US scenario and with the study of Jindal and Mahapatra which reflects Indian scenario.

In our study, 84% (21/25) of patients were seen in pediatric age group as against 70% (16/23) in Mahapatra's study and 86% (24/28) in Meneze's study. A number of patients presenting in pediatric age group were more in all the three studies.

Regarding the location of the sinus (Table 4), the most common location was at the lumbar region in other two studies. In this study also, the sinus tract was most commonly encountered in the lumbar region. The least common location of the sinus was cervical region in all the three studies.

Table 3: Neurological status

Group	Outcome
Neurologically intact pre- and post-operative	28 (56)
Neurologically improved	7 (12)
Neurologically unchanged	8 (16)
Neurologically worsened	7 (14)

Table 4: Comparison with other studies

Type	Menezes	This study	Mahapatra
Bifid laminae	17 (61)	24 (96)	21 (91)
Block vertebra	3 (11)	-	-
Scoliosis	4 (14)	4 (16)	4 (17)
Costal dysplasia with hemi vertebrae	-	2 (8)	-

One of the patients in this study presented with dermal sinus at two different spinal levels (cervical and dorsal). Development of two DSTs at different spinal levels could be explained by multisite closure theory of Van Allen.[3]

In all patients, DST was seen in midline except one where it was slightly off the midline on the right side in the lower lumbar region. DST with dual ostia was not seen in this study which was seen in one patient by Menezes. Development of dual ostia can be explained by recently proposed zipping error hypothesis[4]

Female patients were more common in other studies while male patients were more common in this study.[5,6] Although most of spinal dysraphism conditions were commonly seen in females, the number of male patients with spinal dermal sinus was slightly higher in this study.

Cutaneous findings (48%) were the most common reason to seek medical attention which was also seen by Menezes (54%). However, the presence of neurological deficit was the common reason to come for clinic in Mahapatra's series. Neurological deficits (32%) and local infection and discharge from the sinus (16%) were other frequent reasons in this study.

Incidence of neurological deficit was high in Mahapatra *et al.* (87%) as against 61% in Menezes *et al.* and 32% in this study. One of four adult patients (25%) and seven of 21 pediatric patients (33%) presented with deficits in this study. Menezes *et al.* and Mahapatra *et al.* also observed that deficits were common in pediatric age group. Although only 8 patients (28.5%) were referred for neurological deficits initially, Menezes subsequently found out neurological deficits in 17 (61%) patients. 8 patients (32%) in this study found to have neurological deficits. All of them had autonomic deficits. Regarding the type of deficits, autonomic deficits were common in our study in contrast to motor deficits observed in other series. Gait disturbances were seen in two patients.

When comparing age and sex with the presence of neurological deficit, there was no significant association. (Group A: Patients presenting with neurological deficits, Group B: Patients without neurological deficits) (Tables 5 and 6).

Bifid laminae were seen in all patients except one. Scoliosis was seen in four, another patient who had worsening of deficits developed scoliosis later. Dysplastic development of ribs with hemi vertebrae was seen in two patients who were not reported in other two series.[7] Foot abnormalities were noticed in four patients by Menezes *et al.*, three

patients by Mahapatra *et al.* In our study, no patients had foot abnormality (Table 4).

All patients were evaluated with MRI in this study and by Mahapatra *et al.* Only 24 patients out of 28 had MRI evaluation in Menezes *et al.*

All patients were found to have tethered cord in this study as against 22/28 (78.5%) by Menezes *et al.*[1,8,9] One patient with terminal lipoma was seen.[4,8] Dermoid was seen in 4 (16%) of patients. Three patients had intramedullary dermoid at dorsal level inclusive of the patient with double dermal sinus who also had extramedullary dermoid in cervical region. Another patient had extramedullary dermoid in the lumbar region. Abscess formation in intramedullary dermoid was seen in two patients.[10] One patient with neuroenteric cyst was also seen.[11,12] Split cord malformation was seen in one patient. Epidermoids and hydrocephalus were not encountered in any of the patients. Terminal syrinx was seen in three patients in this study (Table 7).[13]

When applying a statistical test to good (intact and improved) and poor (not improved and worsened) outcome groups, the *P* value is 0.0004. Since, the *P* value is significant (<0.05), it is evident that patients without deficit at presentation have a better prognosis than the patients with deficit.

When the outcome was compared between this study and Menezes *et al.* ($P = 0.5586$), it yielded insignificant value. Hence, there is no difference in outcome between Menezes *et al* and our study. When outcome was compared with Mahapatra *et al.* ($P = 0.0427$), it yielded significant value. Hence, the outcome in this study is better than in Mahapatra *et al.* This may be due to a large number of patients presented with neurological deficits in Mahapatra *et al.* series. Hence, for a good outcome, surgery is warranted before the onset of neurological deficit (Tables 8 and 9).

CONCLUSION

The presence of cutaneous stigmata over the midline neural axis should not be considered benign and must initiate prompt radiologic evaluation and neurosurgical referral. All patients with OSD should be offered aggressive surgical treatment in the form of total excision of cutaneous lesions like dermal sinuses, detethering of tethering elements and correction of associated abnormalities as soon as diagnosed. Chance of preserving and/or improving the neural function is high when surgical intervention is done before the onset of gross neurological deficits.

Table 5: Age difference between groups

Age	Group A	Group B
Range	1.5-27 years	1 month - 24 years
Mean	6.5 years	5.9 years
SD	8.4 years	8.1 years
P	0.1988	
	Not significant	

Table 6: Sex difference between groups

Sex	*n* (%)	
	Group A (8)	Group B (17)
Male	6 (75)	8 (47.1)
Female	2 (25)	9 (52.9)
Total	8 (100)	17 (100)
P	0.2337 Not significant	

Table 7: Associated abnormalities

Associated abnormalities	Menezes	Mahapatra	This study
Dermoid	3	4	4
Epidermoid	-	5	-
Epidermal and endodermal cyst	2	-	-
SCM	3	6	1
Terminal syrinx	-	-	3
Neuroenteric cyst	-	-	1

Table 8: Outcome comparison

Outcome	*n* (%)	
	Group A (8)	Group B (17)
Improved	3 (37.5)	-
Not improved	3 (37.5)	-
Worsened	2 (25)	-
Intact	-	17 (100)
Total	8 (100)	17 (100)

Table 9: Outcome comparison with other studies

Outcome	Menezes	Mahapatra	This study
Neurologically intact pre- and post-operative	11 (39)	3 (13)	17 (68)
Neurologically improved Post-operative	12 (43)	8 (35)	3 (12)
Neurologically worsened	3 (11)	1 (4)	2 (8)
Neurologically unchanged	2 (7)	11 (47)	3 (12)

This is important for clinicians to be able to distinguish between the benign coccygeal pits and the potentially more ominous DSTs. Timely, definitive operative intervention with intradural exploration can preserve or improve neurologic function for many in this patient population. The risks associated with surgical exploration are very low and the benefit is high. There

is no justification for a conservative approach as such a therapy entails the risk of the development of progressive or sometimes acute neurological deficit. Skeletal abnormalities may also occur in the form of neurogenic scoliosis.

A high index of suspicion is required for diagnosing OSDs. MRI is the investigation of choice. Intradural exploration is the most important part of surgical management. Associated pathologies are common and should be dealt appropriately. Outcome is directly related to the pre-operative neurological status which further reiterates the importance of early diagnosis.

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REFERENCES

1. Ackerman LL, Menezes AH. Spinal congenital dermal sinuses: A 30-year experience. Pediatrics 2003;112:641-7.
2. Ackerman LL, Menezes AH, Follett KA. Cervical and thoracic dermal sinus tracts. A case series and review of the literature. Pediatr Neurosurg 2002;37:137-47.
3. Van Allen MI. Multisite neural tube closure in humans. Birth Defects Orig Artic Ser 1996;30:203-25.
4. Lee CS, Phi JH, Kim SK, Cho BK, Wang KC. Spinal congenital dermal sinus with dual ostia. J Neurosurg Pediatr 2009;3:407-11.
5. French BN. Midline fusion defects and defects of formation. In: Youmans JR, editor. Neurological Surgery. Philadelphia, PA: WB Saunders Company; 1990. p. 1081-235.
6. Gupta DK, Shastank RR, Mahapatra AK. An unusual presentation of lumbosacral dermal sinus with CSF leak and meningitis. A case report and review of the literature. Pediatr Neurosurg 2005;41:98-101.
7. Tortori-Donati P, Fondelli MP, Rossi A, Raybaud CA, Cama A, Capra V. Segmental spinal dysgenesis: Neuroradiologic findings with clinical and embryologic correlation. AJNR Am J Neuroradiol 1999;20:445-56.
8. Warder DE, Oakes WJ. Tethered cord syndrome: The low-lying and normally positioned conus. Neurosurgery 1994;34:597-600.
9. Warder DE. Tethered cord syndrome and occult spinal dysraphism. Neurosurg Focus 2001;10:e1.
10. Morimoto K, Takemoto O, Nakamura H, Takeuchi M. Spinal dermal sinus associated with intramedullary abscess and dermoid. Pediatr Neurosurg 2003;39:225-6.
11. Rauzzino MJ, Tubbs RS, Alexander E 3rd, Grabb PA, Oakes WJ. Spinal neurenteric cysts and their relation to more common aspects of occult spinal dysraphism. Neurosurg Focus 2001;10:e2.
12. Muthukumar N, Arunthathi J, Sundar V. Split cord malformation and neurenteric cyst - Case report and a theory of embryogenesis. Br J Neurosurg 2000;14:488-92.
13. Muthukumar N. Terminal syringomyelia communicating with a spinal dermal sinus. J Clin Neurosci 2007;14:688-90.

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