Cystic Cervical Dysraphism: Experience of 12 Cases

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**Abstract**

**Introduction:** Cystic spinal dysraphism of the cervical region is a relatively rare entity, which is more frequently associated with congenital anomalies such as split cord malformation, Chiari malformation, and corpus callosum agenesis, when compared to their lumbosacral counterpart. In our study, we have highlighted the clinical spectrum, associated anomalies (both neural and extra-neural), and surgical nuances of these. **Materials and Methods:** This study is a retrospective analysis of 225 patients from June 2010 to April 2017. Twelve patients who were between the age of 1 month and 16 years were included in our study. Average age was 32.6 months, and there were five female patients and seven male patients. All patients underwent neurological and radiological examinations followed by surgical excision of the sac and exploration of the intradural sac using the standard microsurgical technique. Neurological, Orthopedic and urological outcomes were studied in our description. **Results:** Of the 12 cases, 9 patients (75%) had some associated anomaly. Four of the 12 patients (25%) had split cord malformation, 3 had corpus callosum agenesis, and 5 had Chiari malformation. Patients with cervical spina bifida cystica (SBC) present with less neurologic deficits and greater association with CCA. **Conclusion:** The management strategy and association with other congenital anomalies separates cervical dysraphism as a different clinical entity rather than just group. These patients rather show favorable outcome with regard to neurologic, orthopedic, and urologic problems as compared to their caudal counterpart. Early surgical intervention even before the onset of symptoms is recommended. A proper radiological and urological evaluation is warranted.

**Keywords:** Associated anomaly, cervical dysraphism, double dysraphism, meningocele, myelomeningocele

**INTRODUCTION**

Cystic cervical dysraphism (CD) is a rare clinical entity accounting for 1%-5% of the total spina bifida cystica, with limited publications in literature describing clinical features, etiopathogenesis, and surgical approach.[1] CD is a distinct entity and is different from their more caudally placed counterparts. The patients with CD have higher rate of almost normal motor functions than the patients with lesions at other levels of the spine. CD is associated with higher incidence of associated anomalies and favorable clinical outcome. In this study, we present our experience with patients of CD as a different form of spinal dysraphism emphasizing their differences from the more common lower dorsal/lumbar spinal dysraphism.

**MATERIALS AND METHODS**

Our series consists of 12 patients with cystic CD, who were managed at our center from June 2010 to April 2017. A total of 225 patients were treated...
for spinal dysraphism during this period. All cases were retrospectively analyzed, and all available data, including demographic profile, level of lesion, type of lesion, associated anomalies, and so on, were recorded from hospital case records, outpatient files, and the hospital information system. Radiological assessment consisted of magnetic resonance imaging (MRI) of whole spine with computed-tomography (CT) scan in some patients.

All patients underwent surgical treatment with standard microsurgical techniques. Operative details were noted from the operation notes. In cases that required resurgery, details of the indication and findings of the resurgery were noted. Complications associated with surgery were also recorded. Neurological condition at the last follow-up was noted from the records, and the condition was ascertained over the phone.

RESULTS

Demography

There were 12 patients in our study. Seven patients were of less than 1 year of age, four patients were between 1 and 10 years, and one patient was of 16 years at the time of presentation. Average age was 32.6 months (median, 11.5 years; range, 4–192 months). There were five female and seven male patients.

Clinical features

The patients presented with asymptomatic cervical swelling (8/12), urinary difficulties (3/12) (retention with overflow incontinence in two cases and incontinence with loss of bladder sensation in one case), and difficulty in walking (2/12). None of the patient was referred with cerebrospinal fluid (CSF) leak or dimple.

One patient had ulcer over foot with clubfoot deformity. One patient had curvature anomaly in the form of segmental kyphoscoliosis. One patient was operated for cervical swelling at some other neurosurgical center and was referred to us for new onset of difficulty in walking and increased urinary frequency. On evaluation, it was found that there was persisted tethering. This highlights the need for detethering along with excision of sac in CD [Table 1].

Tethering of the cervical spinal cord

The CD swelling can be meningocele, swelling with stalk, and rarely myelocystocele. In our series, five (41.67%) had thick stalk with lipomatous nodular swelling [Figure 1]. The stalk tissue attached on dorsal surface of cord, coming out through defect in posterior bony elements and attaching to the swelling. One (1/12) patient had myelocystocele. Another patient (1/12) had sac (myelomeningocele). This patient with myelomeningocele had associated Chiari type II malformation and hydrocephalus. Five patients (41.67%) had meningocoele.

Associations

Nine patients (75%) had some associated anomaly (AA). Four of the 12 patients (25%) had split cord malformation (SCM). Among the four patients, one had type I SCM at D7 level, whereas three had type II SCM at the same level of CD. All four patients required surgery for SCM; in three patients, intraoperative fibrous spur was excised, which was at the same level as the CD; and in one patient, a different level of laminectomy was required for the removal of bony spur. Three patients (75%) had associated syrinx.

Table 1: Clinical details of patients in our series with associated anomalies

| S. no | Age/gender | Level | SCM | CM | HCP | CCA | Syrinx | Other AA |
|-------|------------|-------|-----|----|-----|-----|--------|----------|
| 1     | 7 months/F | C4-C6  | Y (Type II) | Y  | Y   |     |      | Kyphoscoliosis |
| 2     | 5 months/F | C2     | Y (Type II) | Y  | Y   |     |      |          |
| 3     | 16 years/F | C2     | Y (Type II) | Y  | Y   | Y   |      |          |
| 4     | 6 years/M  | C4     |       |     |     |     | Y      |          |
| 5     | 12 months/F| C1-C2  |       |     |     |     | Y      | C1 occipitalized with block C2-C3 vertebrae, high-arched palate |
| 6     | 6 months/M | C3, D4-D5, and L4 | Y (Type I) | Y  |     |     |      | Tethering at L4 also with tuft of hair and foot deformity |
| 7     | 4 months/M | C1     |       | Y  | Y   |     | Y      | Developmental delay |
| 8     | 7 months/M | C5     |       |     | Y   | Y   |      |          |
| 9     | 36 months/M| C2     |       | Y  | Y   |     |      |          |
| 10    | 15 months/M| C2-C6  |       |     |     |     | Y      |          |
| 11    | 11 months/M| C2-C3  |       |     |     |     | Y      |          |
| 12    | 24 months/F| C3-C5  |       |     |     |     | Y      | Lumbar LPMC |

HCP: Hydrocephalus
Three of the 12 children (25%) had corpus callosum agenesis (CCA). Among the three patients of CCA, tethering was present at C2 level in all the three cases, and all three had syrinx limited to the cervical spine only. One patient of CD with associated CCA and ventriculomegaly required postoperative CSF diversion for wound leakage.

Five patients (41.67%) had Chiari malformation (CM), and of these five cases, one had type III CM. Three of the five patients of CD with CM had urinary difficulties, whereas one patient had difficulty in walking. This suggests increased incidence of urinary problems when CD is associated with CM. In patients associated with CM, one patient had developmental delay (verbal and social milestones were delayed). The patient with developmental delay had associated CCA. Two patients had hydrocephalus. One patient (20%) had associated SCM. Three patients had syrinx (two holocord and one focal syrinx).

Three (25%) patients had hydrocephalus. Two of these three cases had associated SCM. Two (66.67%) patients required CSF diversion. Between both the cases that required shunt surgery, one patient required diversion surgery before definitive surgery, whereas the other required it in the postoperative period for CSF leak from the wound site.

One patient had short neck, high-arched palate with C2-C3 block vertebrae, and partially occipitalized atlas. The patient was not symptomatic for bony pathology, and surgical excision of CD was performed. One patient of CD had associated lumbosacral lipomyelomeningocele (LPMC) [Figure 2]. This is a rare association with few cases reported so far. The cervical swelling was operated on, and the patient is under follow-up for second surgery.

**Investigation**

MRI is an investigation of choice in these patients. Screening for hydrocephalus (using cranial ultrasound or non-contrast CT scan) is warranted along with urological investigations. Apart from the presence or absence of stalk, these methods will also provide information about the associated anomalies such as CM, hydrocephalus, and hydromyelia. We also recommend a plain X-ray of the whole spine to look for curvature anomaly. We also screen these patients with echocardiography to look for associated congenital cardiac anomaly.
Surgery

All patients underwent surgical excision of the sac and exploration of the intradural sac using the standard microneurosurgical technique. Laminctomy was performed at least one level above and below the involved segments to observe the normal dural tube. Some authors propose two-level laminectomy for adequate exposure of tethering element.[2-3] In five patients, we found thick tubular stalk protruding from the posterior surface of the cord, which was contained in the sac. The stalk penetrated the meningeal covering through a defect in posterior midline structure and was adhered to the cyst either. The stalk was cut a few millimeters away from the spinal cord [Figure 3].

In one patient, there was a cystic mass inside the protruding dural sac. The outer cyst was communicating with the subarachnoid space, whereas the inner cyst was communicating with the hydromyelic spinal cord (myelocystocele); and in one patient, there was only herniation of the meninges filled with CSF without any neural tissue. None of the patients that had Chiari type II malformation required foramen magnum decompression until the last follow-up.

One patient with low-lying cord tethered at L4 required two-level laminectomy at different levels. One patient with bony spur at D7 level required laminectomy and drilling of spur separately.

Management and follow-up

None of our patients showed evidence of neurological deterioration after surgery. Two patients required additional surgery; one required CSF diversion in the form of ventriculo peritoneal shunt before surgery, and second required shunt procedure after surgery for CSF leakage from wound. Intraoperative period was uneventful in all the cases. The follow-up period ranged from 1 to 7.4 years.

Discussion

There are few published case series on CD as a different clinical entity.[4-6,12,13,15] Herein, we present our experience of 12 cases. Incidence of CD in our study was 5.3% (12/225), which almost matches with the incidence quoted in other series (1%–6%).[4-6,12,13,15] Our hospital is a referral center; hence, most of these cases of CD represent almost true incidence of pathology in northern India. Another study from northern India by Kasliwal et al.[4] quoted almost similar incidence in their series. Although the series from Habibi et al.[5] also showed male preponderance of 56.2%, majority of studies showed female preponderance. Our study also showed male preponderance of 58.33%.

Kiyamaz et al.[6] highlighted the etiological factors of CD. The most frequent etiologies of spinal dysraphism include low socioeconomic level, inadequate folic acid intake, diabetes mellitus, obesity, vitamin B12 deficiency, high body temperature, valproic acid, metabolic teratogenic agents, and environmental contamination. In their series of seven patients, mothers of all seven had not taken folic acid supplementation during their pregnancy,[6] although 2/10 patients gave similar history in our series.

The classification of cystic dysraphism changed with the introduction of the term “limited dorsal myeloschisis” in the early 90s. The terminology included spectrum of dysraphism including meningoceles and meningoystoceles. Pang and Dias[7] further subcategorized them according to their internal structures such as limited dorsal myeloschisis containing a fibrovascular stalk of neurons, glia, and peripheral nerves inside a dural sac; and SCM consisting of two hemicords in a dural sac separated by a fibrous septum that attaches the hemicords to the dura at the site near the limited dorsal myeloschisis stalk. Salomão et al.[8] and Habibi et al.[5] also divided them according to the presence or absence of fibrovascular stalk.

Most of the authors believe that embryologically CD occurs as a part of incomplete neurulation. It is believed that most of the neurulation occurred...
normally except the final fusion of the apposed neural fold. A thin area in dorsal midline remains open. To be more specific, separation between the cutaneous ectoderm and neural ectoderm never occurred at this point, although neurulation occurred normally. [7] The myofascial tissues develop a dorsal median stalk of central nervous system tissue; it remains as the original attachment between the nearly closed neural tube and the gaping cutaneous ectoderm. The higher incidence of normal motor function in CD is in agreement with the limited and delayed neurulation abnormality hypothesis. Kasliwal et al.[4] highlighted the fact that CD differs from patients with dysraphism at other places, as content is not a misplaced neural placode but a neuroglial stalk that arises from the dorsal surface of the cervical cord, which causes the tethering. Theory of multiple-site closure by Van Allen et al.[9] and the role of specific gene controlling separate site explain

| S. No | Study                | CM  | SCM | HCP | Syrinx | Curvature anomaly | CCA | Others                                      |
|-------|----------------------|-----|-----|-----|--------|-------------------|-----|---------------------------------------------|
| 1     | Konya et al.[12]     | 0/1 | 0/1 | 0/1 | 0/1    | 0/1               | 0/1 | --                                         |
| 2     | Kasliwal et al.[4]   | 2/10| 1/10| 5/10| 3/10   | --                | --  | Subluxation of C1 over C2 in 1/10, thoracicolumbar MMC in 2/10 Developmental delay in 1/6 Sacral MMC in 1, Lipo-MMC in 1, mature teratoma in 1 Klippel–Feil syndrome in 1/5 |
| 3     | Andronikou et al.[3] | 4/6 | 3/6 | 2/6 | 2/6    | --                | 2/6 | --                                         |
| 4     | Habibi et al.[5]     | 4/16| 1/16| 8/16| 2/16   | --                | --  | --                                         |
| 5     | Duz et al.[4]        | --  | 1/5 | --  | --     | --                | --  | --                                         |
| 6     | Kymaz et al.[6]      | 3/7 | --  | 3/7 | --     | --                | --  | --                                         |
| 7     | Delashaw et al.[13]  | 2/4 | 1/4 | 2/4 | 3/4    | 1/4               | --  | --                                         |
| 8     | Huang et al.[1]      | 1/10| 1/10| 3/10| --     | --                | --  | Sacral spina bifida occulta in 1 Pes equinus in 2/5, mental retardation in 2/5, periventricular heterotopias in 1, ADHD in 1/5, FSH muscular dystrophy in 1, thickened filum with lipoma in 1 LDM in 5/9 |
| 9     | Meyer-Heim et al.[9] | 3/5 | --  | 2/5 | --     | 1/5               | --  | --                                         |
| 10    | Pang and Dias[7]     | 4/9 | 4/9 | 6/9 | --     | --                | --  | --                                         |
| 11    | Steinbok and Cochrane[10] | 5/8 | --  | 5/8 | 3/8    | --                | --  | --                                         |
| 12    | Sun et al.[14]       | 5/8 | --  | 5/8 | 3/8    | 1/8               | --  | Hemivertebrae in 1/8, block vertebra in 1, thoracicolumbar MMC in 1 1 patient had block C2-C3 vertebrae with high-arched palate, 1 had developmental delay, 1 had clubfoot, and 1 lumbosacral LPMC |
| 13    | Present study        | 5/12| 4/12| 3/12| 6/12   | 1/12              | 3/12| --                                         |
|       | Total                | 38/96| 16/73| 44/96| 22/65 | 4/30 (13.33%) | 5/19 (26.3%) | -- |

ADHD: Attention deficit hyperkinetic disorder, FSH: Facioscapulohumeral muscular dystrophy, LDM: Limited dorsal myeloschisis, MMC: Myelomeningocele
the occurrence of swelling at cervical level and its rare associations.

In our series, one case had double LPMC. The concept of multiple-site closure of neural tube as proposed by Van Allen et al.,[9] suggests embryogenesis of these swellings. According to him, the neural tube closes at five to six different sites, so defect in closure at different sites can lead to multiple swellings. Kasliwal et al.[4] had reported 2 cases of thoracicolumbar myelomeningocele (MMC) of the 10 cases in their series. There were two cases (2/16) of lumbosacral LPMC in series from the study by Habibi et al.[5]

The incidence of associated anomalies has been quoted differently in other studies,[4,5,6,12,13,15] A total of 3/10 patients had hydrocephalus. Meyer-Heim et al.[10] reported 2/5, Salomão et al.[8] reported 7/18, Steinbok and Cochrane[11] reported 5/8, and Habibi et al.[5] found 50% association. The incidence in our study matches with that from the other series in literature.[4,5,6,12,13,15] The high incidence of hydrocephalus demands screening in all patients of CD. Mere presence of hydrocephalus should not be considered as an indication of CSF diversion in preoperative period, but these patients should be followed up closely in postoperative period with low threshold of diversion [Table 2].

It seems that the universal law of association between CM and spinal dysraphism does not apply to CD. A total of 5/12 (41.6%) patients in our study had CM. Habibi et al.[5] found only 25% association in their series.[10] One of the patients in our series had type III CM, and to the best of our knowledge, we believe this co-occurrence has never been reported before. Previous series reported higher correlation among Chiari type II malformations in myelocystocele subgroup of CD.[16] In our series, one patient of myelocystocele was not associated with CM.

Contrary to thoracicolumbar variety, surgical indication for CD remains mostly cosmetic or for the prevention of motor deficits. The surgical management of CD requires excision of sac with detethering. All of our patients underwent intradural exploration and detethering in addition to sac resection. One patient required CSF diversion in postoperative period. In the series by Kasliwal et al.,[4] 3/10 patients had CSF leak, of which one required theco-peritoneal shunt and two required dural repair.[15]

These patients require meticulous dissection of sac with identification of cause of tethering. The characteristic lack of placode in these cases obviates one to identify attachment of stalk. The surgical exposure including two vertebral levels above and two vertebral levels below is advocated for proper access of tubular stalk. It is said that even if intraoperative and radiological findings do not show tethering, dura should be opened and the cause of tethering should be searched. In our series, we could find either adhesion or stalk placed dorsally in all patients. The stalk should be cut at the base, and intradural exploration should be carried out. Sometimes posterior surface of the cervical spinal cord is attached with filamentous attachments to the wall of the meningocele in these lesions.

The exact mechanism of tethering is yet to be proved; however, in their study, Duz et al.[14] described how band could lead to tethering. They believed that the mechanism is same as that of compressive cervical spondylotic myelopathy. The dorsal traction force of the tethering stalk or adhesion may be the cause of the possible neurological deterioration because of the stretch injury to the cervical spinal cord.[12,13]

**CONCLUSION**

Cervical spine dysraphism is a rare entity with different clinical spectrum from their lumbosacral counterpart. The patients of CD are safe to operate with fewer incidences of CSF leak and motor weakness. The two important indications to operate are early detethering and cosmetic. The patients of CD should be screened for associated anomalies and urological workup for prognostication and better follow-up.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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