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RESEARCH ARTICLE

CERVICO DORSAL SPINAL DYSRAPHISM - AN INSTITUTIONAL EXPERIENCE

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Key words:-
Spinal Dysraphism, Hydrocephalus, Chiari Malformation, Diastematomyelia

Abstract

Introduction: Spinal dysraphisms are most common congenital anomalies of central nervous system.Cervicodorsal spinal dysraphisms are rare compared to lumbosacral dysraphism, with incidence of 1-6%. Associated anomalies such as split cord malformation, hydrocephalus, Arnold chiari malformation, corpus callosum agenesis, are frequently seen in cervico dorsal compared to lumbosacral dysraphism. In our study, we highlighted the clinical spectrum, associated anomalies, surgical nuances and outcomes of cervicodorsal spinal dysraphism presented to department of neurosurgery, coimbatore medical college hospital, coimbatore.

Material and Methods: This study includes patients with cervico dorsal spinal dysraphism, presented to Department of neurosurgery, coimbatore medical college hospital,Coimbatore between 2015–2019. All patients underwent neurological and radiological examination and associated anomalies were addressed accordingly either prior to or along with definitive treatment. The last available follow up in hospital recordings was taken for outcome assessment.

Results: A total of 13 patients with cervico dorsal spinal dysraphism were operated between 2013 and 2019. Among them 5 [40%] were of cervical region and 8[60%] were of thoracic region.The age at time of admission was from 10days to 10years.There were 10 female and 3 male children. The most common presentation was isolated swelling.Mild lower limb weakness in 2 cases (1 cervical and 1 thoracic). There were also no orthopedic abnormalities.3 patients had hydrocephalus , of them chiari type 2 malformation was found in two cases(1 cervical and 1 thoracic).Diastematomyelia was seen in 5 patients. All cases were evaluated extensively with Magnetic Resonance imaging of Spine with Brain screening and the case associated with split cord malformation was further evaluated by CT scans. All patients underwent surgical excision of the sac and exploration of the indradural sac using the standard technique.Hydrocephalus was treated with ventriculo-peritoneal shunt, Diastematomyelia was treated with excision of bony spur and reconstruction of Dural tube.

Conclusion: Cervicothoracic spinal dysraphism has more favourable outcome in respect to Neurological, orthopaedic and urologic problems compared to lumbosacral Dysraphism.Surgical treatment consisting

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intradural exploration of Lesion,untethering of spinal cord and excision of potential adhesions should be Performed in early period to prevent neurologic deterioration. Patient outcome depends on presence of associated anomalies and whether complete resection is performed.

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Introduction:-

Spinal dysraphisms are the most common congenital anomalies of central nervous system. Cervicodorsal spinal dysraphisms are rare compared to lumbosacral dysraphism, with incidence of 1-6%. Associated anomalies such as split cord malformation, hydrocephalus, arnold chiari malformation , corpus callosum agenesis, are frequently seen in cervico dorsal compared to lumbosacral dysraphism. Cervico dorsal variety have better prognosis due to lack of functional neurological tissue in dysraphic sac. In our study, we highlighted the clinical spectrum, associated anomalies, surgical nuances and outcomes of cervicodorsal spinal dysraphism presented to Department of neurosurgery, Coimbatore medical college hospital, coimbatore.

Material and Methods:-

This study includes patients with cervico dorsal spinal dysraphism, presented to Department of neurosurgery, coimbatore medical college hospital,Coimbatore between 2015–2019. All patients underwent neurological and radiological examination and associated anomalies were addressed accordingly either prior to or along with definitive treatment. Surgical excision of sac and exploration of intra dural sac with detethering of spinal cord was done. Neurological , orthopedic and urologic abnormalities were analysed in our study. The last available follow up in hospital recordings was taken for outcome assessment.

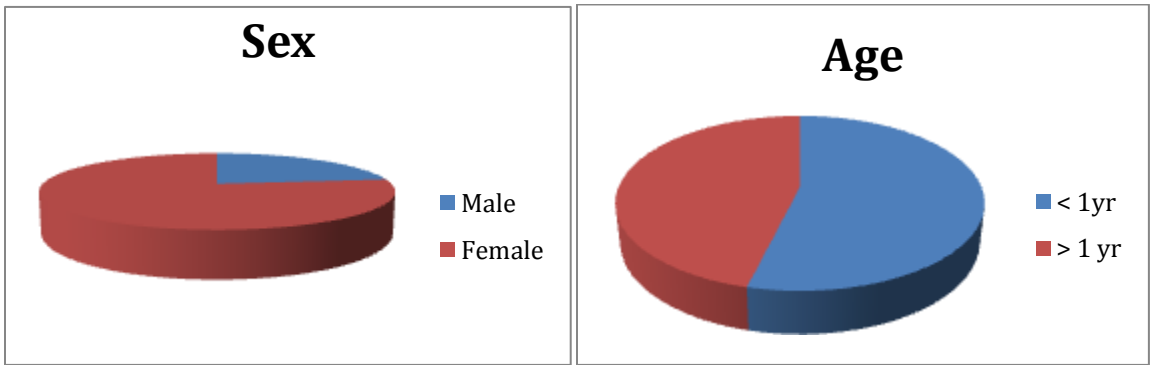
Results:-

Demographic profile:

A total of 13 patients with cervico dorsal spinal dysraphism were operated between January 2015 and December 2019.

Among them 5 [40%] were of cervical and 8[60%] were of thoracic.The age at time of admission was from 10days to 10years.In total, 55% (n = 7) of patients were under age of 1 year and 45% (n=6)were age of over 10 years.

There were 10 female and 3 male children.



Clinical features:

The most common presentation was isolated swelling . Mild lower limb weakness was seen in 2 cases (one cervical and one thoracic). There were also no orthopedic abnormalities.Of the 3 patients with hydrocephalus , chiari type 2 malformation was found in two cases(one cervical and one thoracic). All cases were evaluated extensively with craniospinal X rays and Magnetic Resonance imaging of Spine with Brain screening and the case associated with split cord malformation were further evaluated by CT scans.

Associated anomalies:
Chiari malformation (CM):
2 Chiari malformation was seen in two patients(one cervical and two thoracic)

Splitcord:
Out of 13 patients, 5 had associated SCM , All of these had the bony spur at the level of the dorsal region.

Hydrocephalus:
Out of 13 patients,3 had hydrocephalus (one in cervical group and two in dorsal group). Of these 3 patients, All presented with swelling. All were treated with Ventriculo-peritoneal shunts.

Surgical findings:
All patients underwent surgical excision of the sac and exploration of the indradural sac using the standard technique. Laminotomy was performed at least one level above and one level below the involved segments so as to observe the normal dural tube. Associated complications were addressed. Hydrocephalus was treated with ventriculo-peritoneal shunt, Diastematomyelia was treated with excision of bony spur and reconstruction of Dural tube.

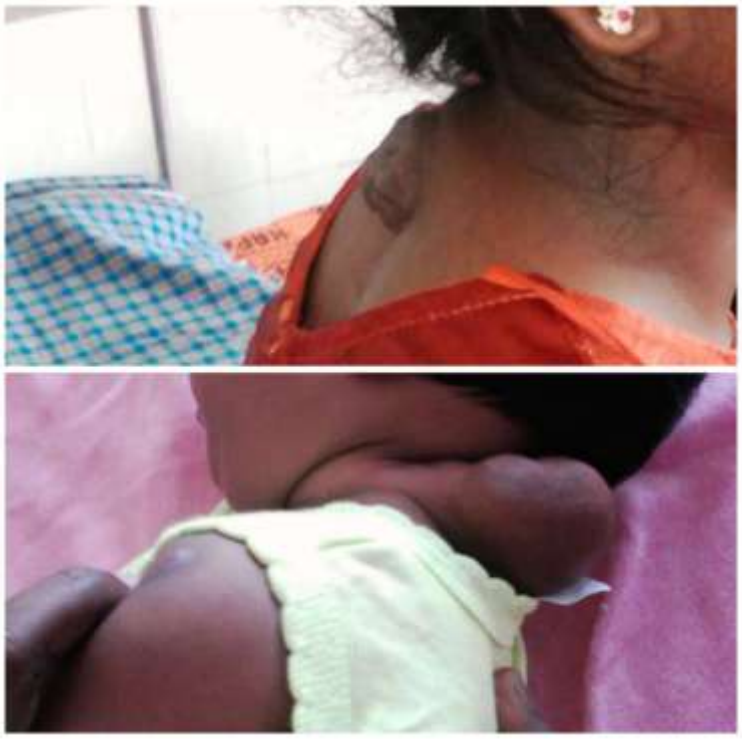
Postoperative course:
One patient had postoperative CSF leak (thoracic region lesion)and the CSF leak subsided on acetazolamide and wound aspiration with daily dressing.

Cervical group:

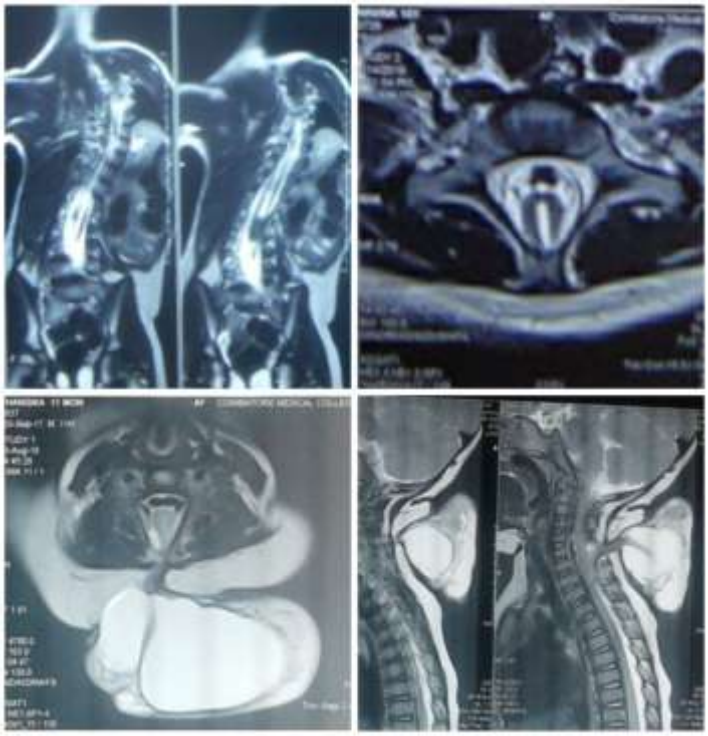
Case no	Age/ sex	Date of surgery	Imaging findings	Associated anomalies	Procedure
1	1y/m	28/2/18	C4-C5 Spinal defect /sac with Syrinx at that level	-	Excision of sac and repair
2	3m/m	21/9/18	C3-C4 Spinal defect /sac	-	Excision of sac and repair
3	10d/m	28/5/19 8/6/19	C6-C7 defect /sac	Chiari type2 with hydrocephalus	Ventriculoperitoneal shunt Excision of sac and repair
4	10y/f	21/6/19	C7-D1 defect with atresia sac	-	Excision of sac and repair
5	8m/m	30/11/19	C7 -D1 defect with sac	-	Excision of sac and repair

Thoracic group:

Case no	Age/ Sex	Date of surgery	Imaging Findings	Associated anomalies	Procedure
1	16d/f	16/6/15	D4-D6 spinal defect/ sac	Hydrocephalus	Ventriculo peritoneal shunt Excision of sac and repair
2	2y/f	15/3/16	D12 bony spur with split cord	Diastematomyelia	Excision of bony spur and repair
3	6y/f	5/8/16	D11.-L1 bony spur with splitcord	Diastematomyelia	Excision of bony spur and repair
4	17d/f	30/8/16	D1-D3 spinal defect /sac with C7 -D1 syrinx	-	Excision of sac and repair
5	2y/f	2/9/16	D10-L1 bony spur with splitcord	Diastematomyelia	Excision of bony spur and repair
6	23d/f	28/9/18 5/10/18	Mid dorsal spinal defect with sac	Chianti type 2 with hydrocephalus	Ventriculo peritoneal shunt Excision of sac and repair
7	7y/f	12/10/18	D8 bony spur with split cord	Diastematomyelia	Excision of bony spur and repair
8	1y/f	9/7/19	D9 bony spur with splitcord	Diastematomyelia	Excision of bony spur and repair



Representative Clinical images



Representative Radiographic images.

Discussion:-

Embryological basis:

The embryological development of CTM is obscure. Spinal dysraphism results from an abnormality in developmental steps including neurectodermal, mesodermal and cutaneous (somatic) ectodermal components .

Abnormal neurulation in embryological period is proposed in distal Myelomeningocoele development. In case of CTM, the neurulation process is uneventful except for fusion of the two sides of the neural fold . Imperfect closure of the neural tube and deficient separation of the cutaneous ectoderm from neural ectoderm results in dorsal myeloschisis. Pang and Dias were the first to suggest the failure of cutaneous ectoderm to cause dorsal myeloschisis . Another theory regarding failure of closure is fusion of the cutaneous ectoderm properly while attachment of neural ectoderm to cutaneous ectoderm incurs maldevelopment of the skin .

Classification:
According to the anatomical structure of the lesions; Cervical dysraphism can be classified to 3 types

Pedunculated:
Protrusion of vascularized tissue from posterior surface of spinal cord and passing through the defect in posterior midline structures to attach to the sac. This may be of neuroglial or fibrovascular in origin.

Myelocytoceles:
A second cystic cavity lined by ependyma herniates through defect into sac. The initial outer cyst is associated with subarachnoid space but an internal second cyst has connection to the hydromyelic canal.

Meningocele:
Meningeal tissue herniates through the defect and the sac contains CSF. There are no neural elements in the sac and arachnoidal band may be present that tethers spinal cord. At times few nerve roots may be present in the CSF-filled sac .

Differences from lumbosacral lesions:
Cervical dysraphism lesions are structurally distinct lesions than myelomeningoceles of the lower thoracic and lumbar regions. The neural placode is absent in CTM. They are more limited and more protuberant and are usually covered by normal skin tissue to a certain extent of the defect,excluding the dome, which is lined by squamous epithelium or with scar tissue . The wall of the CTM is quite strong. Neural structures are therefore not exposed through the defect and CSF leak is not usual . However, tethering of the neural structures to nearby dural or intrasaccular structures may occur. Weakness of lower limbs and bowel/bladder involvement are rare.

Neurological examination:
Initially neurological findings in patients with CTM are not distinctive and usually normal in newborns. They present with more subtle neurological findings compared to lower level lesions, urological disorders and long tract findings are rarely seen in CTM . Although CTM causes tethering of the spinal cord, generally neurological functions of the patients are preserved below the level of lesions . Unless there is tonsillar ectopia and hydranancephaly , cognitive function may be normal. Posterior fossa distortions and hindbrain herniations is correlated with intellectual dysfunctions , IQ levels being in normal ranges .

Associated lesions:
Cervical dysraphisms are usually associated with other developmental abnormalities of spine and central nervous system. A Chiari type 2 malformation is the leading congenital lesion among these associated abnormalities. Other anomalies associated with cervical dysraphisms include hydromyelia, hydrocephalus , diastematomyelia, lipomyelomeningoceles, thickened filum terminale, Klippel-Feil syndrome and thoracic hemivertebra etc.

Diagnostic work up:
Neural structures cannot be evaluated thoroughly just with plain X-rays. A detailed examination should include magnetic resonance with/out computerized tomography studies to delineate cervical lesions, the position of neural structures and associated anomalies, the CTM and its contents. These methods will also provide critical information about associated Chiari malformation, hydrocephalus and syringomyelia. CT myelography also provides some useful information in particular cases. Besides these imaging techniques, urodynamic studies should be performed as a routine evaluation. These diagnostic tests provide preoperative information about the current condition of the lesion and spinal cord, and the postoperative follow up and prognosis .

Surgical treatment:
Surgical treatment of cervical myelomeningoceles aims basically at cosmesis, untethering of the neural structures, and prevention of infections. Surgical treatment should always involve intradural exploration to untether the neural structures, and excise arachnoid band and septations. Some authors recommend at least a two-level laminectomy to expose the lesion properly. This provides detailed anatomical orientation and stalks, bands, and roots can be identified. Limited surgery focused on removal of the sac and cosmetic correction of the lesion is not advantageous from the prognostic point of view and carries high risk of tethering of the important neural structures and therefore late neurological deterioration. Even if intraoperative and radiological findings preclude tethering, untethering should be performed prophylactically. Inadequate treatment may cause postoperative neurological deterioration. Adequate treatment should include the following; preoperative diagnostic work up to identify tethering of neural structures, intradural exploration of the CTM, and excision of fibrotic and other aberrant tissues adhering to the spinal cord .

In addition to surgical treatment of CTM, there may be associated anomalies that can lead neurological deterioration and tethering of the spinal cord. Split cord malformations, thickened filum terminale, lipomyelomeningocele, etc. Add a risk of traction or compression to the spinal cord. They should be treated accordingly if diagnosed by a thorough diagnostic workup .

Resection of the sac and intradural exploration was performed in every single case. Arachnoidal adhesion, if present, leading to tethering of the spinal cord was excised and untethering achieved. The three cases with hydrocephalus underwent VP shunt in addition to the basic surgery for the sac.

Conclusion:-
Cervicothoracic spinal dysraphism has more favourable outcome in respect to Neurological, orthopaedic and urologic problems compared to lumbosacral Dysraphism. Imaging plays a critical role in surgical planning and screening the central nervous system for additional anomalies. Surgical treatment consisting intradural exploration of Lesion, detethering of spinal cord and excision of potential adhesions should be performed in early period to prevent neurologic deterioration. Patient outcome depends on presence of associated anomalies and whether proper detethering is performed.

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