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Original Investigation

Spinal Dysraphisms of the Cervicothoracic Region in Childhood

Çocukluk Çağı Servikodorsal Yerleşimli Spinal Disrafizm Olguları

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ABSTRACT

PURPOSE: Spinal dysraphisms are the most common congenital anomalies of the CNS. Spinal dysraphism (SD) of cervical and upper thoracic region are rare, demonstrating distinct clinical and structural configurations compared to lumbar counterparts.

METHODS: In Haydarpasa Numune Hospital, a total of 7 cases (3 male and 4 female) ranging between 20 days to 9 years of age with cervicothoracic SD were operated on between 2002 and 2008. The sacs were located in the cervical and thoracic region in 4 and 3 of the cases, respectively. The associated anomalies were diagnosed in 3 cases, including SCM type 2, Chiari type 2, hyrocephalus and hydromyelia. All cases underwent surgical treatment that involves excision of the sac and intradural exploration providing untethering of the spinal cord. Postoperative follow up of these patients was uneventful and neither neurological deficits nor complications were observed.

CONCLUSION: Cervicothoracic SD has more favorable outcome in respect to neurological, orthopaedic and urologic problems compared to lumbar counterparts. In order to prevent forthcoming neurological deterioration, surgical treatment consisting of intradural exploration of the lesion, untethering of the spinal cord and excision of potential adhesions should be performed in the early period.

KEYWORDS: Cervical, Childhood, Spinal dysraphism, Upper thoracic

ÖZ

AMAÇ: Spinal Disrafizmler santral sinir sisteminin en sık görülen doğumsal anomalileridir. Servikal ve dorsal bölgede yerleşimleri nadirdir. Klinik ve yapısal olarak lomber yerleşimlilerden farklı şekillerde görülebilirler.

METOD: Haydarpaşa Numune E.A. Hastanesi Beyin Cerrahisi Kliniği'nde 2002 ile 2008 yılları arasında, yaşları 20 gün ile 9 yaş arasında değişen 3 erkek, 4 kız olmak üzere toplam 7 servikodorsal spinal disrafizm (ASD) olgusu ameliyat edilmiştir. Keselerin yerleşimi 4 olguda servikal, 3 olguda ise dorsal bölgedir. Üç olguda tip 2 split kord malformasyonu, Chiari tip 1 malformasyonu, hidrosefali ve hidromyeli, miyelomeningosele eşlik etmiştir. Tüm olgular ameliyat edilerek kese eksizyonu ve intradural omurilik serbetleştirilmesi yapılmıştır. Postoperatif dönemde komplikasyon görülmemiştir.

SONUÇ: Servikodorsal spinal disrafizmler, lomber bölgede yerleşenlere göre nörolojik, ortopedik ve ürolojik açıdan daha iyi prognoza sahiptir. Olası nörolojik kötüleşmeyi önlemek amacıyla olgular erken dönemde ameliyat edilerek kese eksizyonu yapılmalı, intradural eksplorasyonla omurilik serbestleştirilmesi gerçekleştirilmelidir.

ANAHTAR SÖZCÜKLER: Çocukluk çağı, Servikal, Spinal disrafizm, Üst dorsal

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INTRODUCTION

Myelomeningoceles in the cervical and cervicothoracic region are rare entities accounting for approximately 1-8% of all neural tube defects [4,6,7,8,10,11,12,13,14). Cervicothoracic Myelomeningoceles (CTM) are different from thoracolombar and lumbosacral counterparts in some respects (5,8,10,11). These lesions are also commonly associated with Chiari malformations, syringomyelia and split cord malformations (2,6,8,13). The following report is to emphasize the distinct nature of the CTM compared to involvement of the lower levels based on clinical, radiological and surgical features with particular stress on the age group of the patients.

MATERIAL and METHODS

Seven cases with CTM were operated at the Haydarpasa Numune Hospital, Neurosurgery Clinic between 2002 and 2008. Four of the 7 cases were female and their ages ranged from 20 days to 9 years. All patients were referred to our clinic with sac lesions in the cervical and thoracal region. Neurological examinations revealed normal findings, moreover there were no orthopedic and urological finding. Myelomeningoceles located at the cervical and thoracic region were observed in 4 and 3 cases, respectively. CT and MRI evaluation was performed in all patients as a standard evaluation parameter. Cervical dysraphisms were of the fibrovascular type in 4 cases, myelocytocele type in 2 cases and meningocele type in one case. One of the cases was associated with SCM type 2, chiari type 1 and hydrocephalus whereas one other case was associated with hydromyelia besides chiari type 1 and hydrocephalus. (Table I). Resection of the sac and intradural exploration was performed as the surgical treatment in all cases and untethering of the spinal cord was provided. Two cases with hydrocephalus were managed with ventriculoperitoneal shunting. Postoperative examination revealed no neurological deficits and patients were discharged without any complication.

Case 1: A 9-year-old girl was admitted to our clinic with a mass lesion in the cervical region. Physical examination revealed normal findings except a myelomeningocele sac in the lower cervical region measuring 3.5 x 3 cm in dimension (Figure 1). Neurological, orthopedic and urological evaluations were normal. Cervical CT evaluation demonstrated spina bifida at the C6-7 level (Figure 2). MR showed the myelomeningocele sac and a type II split cord malformation (Figure 3A,B). She was operated on and the sac was excised with C6 and C7 laminectomy. The fibrous septum was removed with intradural exploration (Figure 4A,B). The spinal cord was released and arachnoidal adhesions were

Table I: Demographic and characteristic features of the 7 cervicothoracic myelomeningocele cases.

Patient no	Age/sex	Neurological examination	Level	Associated anomaly	Surgical treatment	Postoperative outcome	Type
1	9/F	Normal	C 6-7	SCM type II	Resection of sac and antethering of arachnoid bands	Uneventful	Fibrovascular stalk
2	9/M	Normal	C 3-4	None	Resection of sac and intradural exploration	Uneventful	Fibrovascular stalk
3	7/M	Normal	C3-4	None	Resection of sac and intradural exploration	Uneventful	Fibrovascular stalk
4	3/F	Normal	T3-4	None	Resection of sac and intradural exploration	Uneventful	Fibrovascular stalk
5	2 months/F	Normal	C 6-7	None	Resection of sac and intradural exploration	Uneventful	Myelocystocele
6	20 days/F	Normal	T 2-3	Chiari type 2, hydrocephalus, hydromyelia	Resection of sac and intradural exploration, VPS	Uneventful	Myelocystocele
7	7 months/M	Normal	T 3-4	Chiari type 2, hydrocephalus	Resection of sac and intradural exploration, VPS	Uneventful	Meningocele

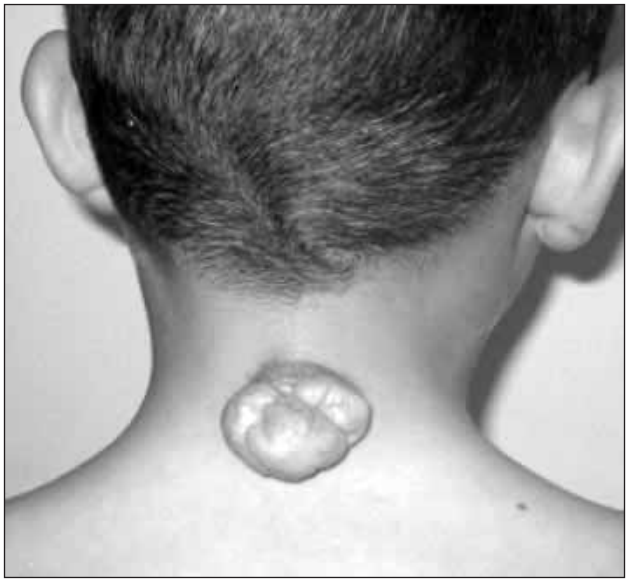

Figure 1: *Photograph showing myelomeningocele sac located in lower cervical region.*

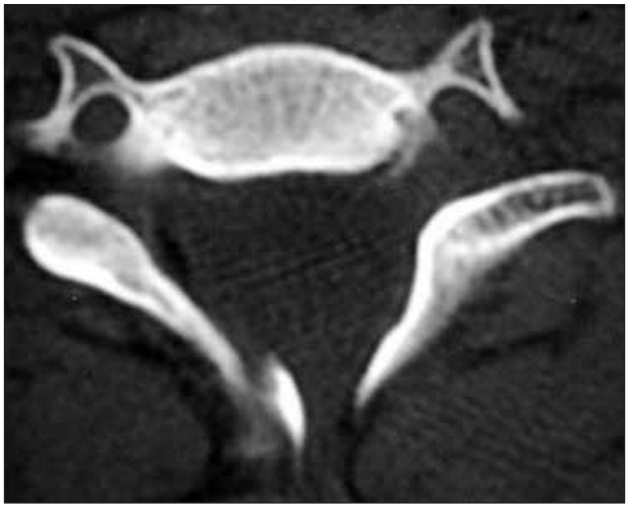

Figure 2: *CT scan of the Case 1 the spina bifida is obvious at the level of C6-7.*

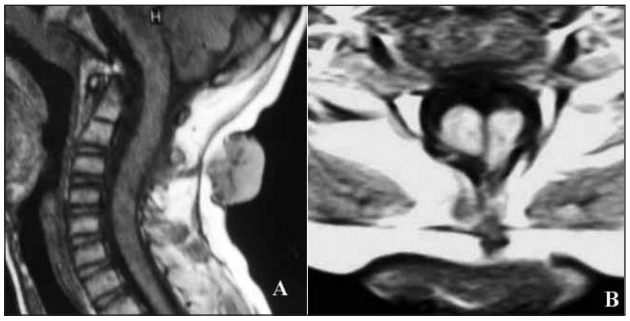

Figure 3: *A myelomeningocele sac with stalk extending into spinal canal associated with type II split cord malformation is demonstrated on T1 weighted sagittal (**A**) and axial (**B**) cervical MRI scans.*

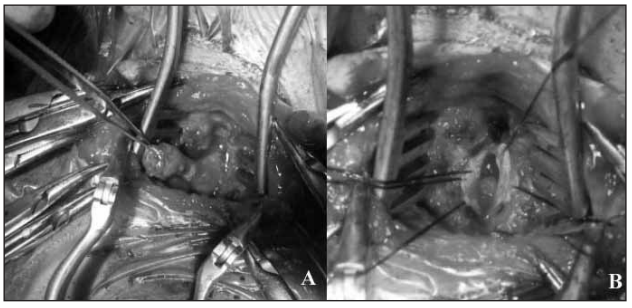

Figure 4: *Intraoperative view of sac excision performed through C6 and C7 laminectomy (**A**). Dural defect and intradural exploration is demonstrated (**B**).*

excised. Histopathological examination reported dispersed fibrosclerosis, meningothelial cellular proliferation and psammoma bodies in the stroma covered by skin tissue. Hyaline cartilage, bony trabeculations, fibrocollagenous connective tissue, focal skeletal muscles and mature fat tissue were also detected in the pathological specimen. The patient was discharged without neurological deficits.

Case 2: A 7-month-old baby was referred to our clinic with a mass in the thoracic region. Physical examination revealed a midline myelomeningocele sac in the posterior thoracal region, 9.5x8 cm in size. Neurological examination findings were normal for his age. Spinal CT and MRI evaluation revealed spina bifida and a myelomeningocele sac at T1-5 level. MRI also demonstrated a Chiari type 1 malformation (Figure 5A,B) and hydrocephalus (Figure 6). The patient was operated for spina bifida, sac excision and spinal cord releasing was performed by intradural exploration. Ventriculoperitoneal shunting was also performed for hydrocephalus. Histopathological evaluation demonstrated chronic nonspecific inflammation and granulation tissue together with recent and old connective tissue

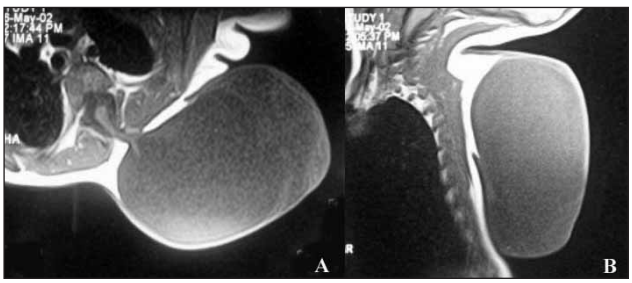

Figure 5: *Axial T1 weighted MRI scan shows huge meningocele sac at T1-5 level (**A**). Associated Chiari type 1 malformation demonstrated together with the huge sac on T1 weighted sagittal MRI scan (**B**).*

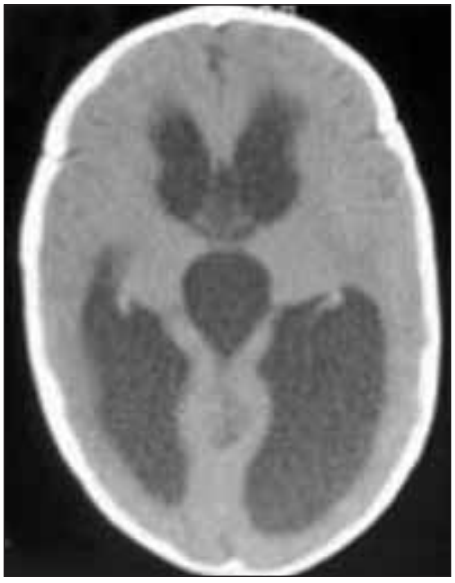

Figure 6: CT scan showing hydrocephalus in case 2.

proliferation was demonstrated. On the internal side, hyperkeratotic, atrophic squamous epithelial inclusion was detected.

DISCUSSION

Classification

Cervical dysraphism can be classified to 3 types according to the anatomical structure of the lesions;

Type 1 CTM (stalks): a vascularized tissue protruding from posterior surface of spinal cord and passing through the defect in posterior midline structures to attach to the sac wall. This tissue may be in neuroglial or fibrovascular in origin.

Type 2 (Myelocytoceles): a second cystic tissue 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.

Type 3 (Meningocele type): meningeal tissue herniates through the defect and the sac contains CSF. Neural elements do not exist in the sac and arachnoidal band may tether the spinal cord. Some nerve roots may be present in the CSF-filled sac (11).

Differences

Cervical dysraphism lesions are structurally distinct lesions than myelomeningoceles of the thoracal and lumbar regions (3,5,9). 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 of the CTM, which is lined by squamous epithelium or with scar tissue (11). The context of the CTM is quite strong. Neural structures are therefore not exposed through the defect and CSF leak is not usual (10). However, tethering of the neural structures to nearby dural or intrasaccular structures may ensue (8).

Neurological Examination

Initially neurological findings in patients with CTM are not distinctive and usually normal in newborns (3,7,9,11,13). They present with more subtle neurological findings compared to lower levels, urological disorders and long tractus findings are encountered less in CTM (4). Although CTMs causes tethering of the spinal cord, generally neurological functions of the patients are preserved below the level of lesions (10). Should no tonsillar ectopia and hydranancephaly exists, normal cognitive function may be observed Posterior fossa distortions and hindbrain herniations will be correlated with intellectual dysfunctions (8), IQ levels being in normal ranges (14).

Embryological Basis

The embryological development of CTMs is obscure. Spinal dysraphism results from an abnormality in developmental steps including neurectodermal, mesodermal and cutaneous (somatic) ectodermal components (14). Abnormal neurulation in embryological period is proposed as in distal MM development. In case of CTM, the neurulation process is uneventful except for fusion of the two sides of the neural fold (4). 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 (10). 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 (13).

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 abnormalities. Other anomalies associated with cervical dysraphisms include hydromyelia, hydrocephalus, Chiari malformations, diastema-

tomyelia, lipomyelomeningoceles, thickened filum terminale, Klippel-Feil syndrome and thoracic hemivertebra etc (2,3,6,8,9,13).

Diagnostic work up

Neural structures cannot be evaluated thoroughly just with plain X-rays. A detailed examination should include magnetic resonance and 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 crucial information about associated Chiari malformation, hydrocephalus and hydromyelia. 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 colon, and the postoperative follow up and prognosis (1,2,4,5,6,10,14).

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 (3,4,6,8,10,11,12,14).

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 (4,5,7,14).

In the presented study, the neurological examination and urodynamic studies were normal in all cases. There were also no orthopedic abnormalities. The associated conditions were hydrocephalus, chiari type 1 malformation and SCM type 2 malformation seen in two, one and one cases, respectively. All cases were evaluated extensively with craniospinal X rays and Magnetic Resonance imaging and the case associated with split cord malformation was further evaluated by cervical CT scans. Besides these radiological evaluations, all cases were also examined by orthopedic and urodynamic means.

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 two cases with hydrocephalus were further operated for VPS in addition to the basic surgery for the sac.

The cases were classified into groups as 4 cases in type 1, 2 cases in type 2 and one case in type 3 groups. It is intriguing that 4 cases were diagnosed in preschool and school age children, both of which belonged to the type 1- fibrovascular group. These four cases were also distinct from the other cases in literature, to the best of our knowledge, as there is just one symptomatic case with cervical meningocele reported in adulthood (6). All four cases presented with cosmetic expectations in the school age period, without any symptoms, neurological deficits or deterioration in the neurological status. All other cases are diagnosed at newborn period and infancy. All these cases, whether symptomatic or not, were operated on based on the fact that normal cervical motions lead to microtrauma to the spinal cord, causing tethering of spinal cord and forthcoming neurological deterioration. Surgical treatment was aimed at untethering the spinal cord and relieving risk of neurological deterioration.

CONCLUSION

The presented study, in accordance with the reviewed literature, indicated that CTM has a more favourable outcome than lower myelomeningoceles

in respect to neurological, orthopedical and urological dysfunctions. Thorough evaluation of the patient including both the lesion itself and the associated anomalies will provide a better perspective of the CTM. Future neurological deterioration will be prevented if extensive surgical treatment with untethering of neural structures in and around the defect together with management of the associated anomalies is provided.

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